Parent and Caregiver Handbook for Sickle Cell

A guide for birth to age 18

California Department of PublicHealth

go.cdph.ca.gov/SCParents

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Written by physicians and other medical providers

at UCSF Benioff Children's Hospital Oakland Sickle Cell Center

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SICKLE CELL DISEASE FOUNDATION

Dear Parent or Caregiver:

Someone may have recently told you that tests show that your baby or young child has sickle cell disease. You may be hearing new information from medical professionals about your child's future. It may seem like there is so much to learn. You may have a lot of questions.

Because you need a reliable source with the latest information on sickle cell disease, we have put together this handbook and a web page for you.

Please use this handbook and the web page to get involved with your child's care. Learn about sickle cell disease and how to handle the most common symptoms of sickle cell disease so that you can help your child lead a full, successful, happy life.

By working together with your child's doctor and other medical providers, you can make sure that your child gets the best care. In addition to medical care, this handbook addresses other parts of your child's life that can be affected by sickle cell disease, such as their well-being at home, school, and in the larger community.

This handbook was created by families and staff at the University of California Benioff Children's Hospital Sickle Cell Center of Excellence in Oakland and San Francisco, in Northern California. The Sickle Cell Disease Foundation and the State of California Department of Public Health provided information, services, and support. We have worked to put this book together to support you and your family.

Please note that in the past this handbook had Part I for birth up to age 6 years and Part II for ages 6 to 18 years. We have moved the important sections of Part II into this handbook. It now covers birth to age 18 years.

This handbook and related information are online (go.cdph.ca.gov/SCParents).

We hope this handbook is useful to you.

Sincerely,

Elliott Vichinsky, MD, and Beth Apsel Winger, MD, PhD,

UCSF Benioff Children's Hospital Oakland Sickle Cell Center

Note to Medical Providers

This handbook was developed for parents of children with sickle cell disease. They requested a detailed handbook they could refer to as needed to guide them through the journey of caring for a child with sickle cell disease.

The role that parents play in early identification of infection, pain, and other problems is challenging and critical. A major goal of this handbook, and the associated web page, is to empower parents to assume the role of partner in the delivery of sickle cell disease care to their child.

We hope that the handbook is also useful for parents to share with primary care providers and Emergency Room (ER) staff who are unfamiliar with the treatment of sickle cell disease.

We do not recommend that you as a medical provider give this handbook to all parents of infants with sickle cell disease at their first visit. Many will be overwhelmed with the amount of information as they try to understand their child's new diagnosis. Parents should receive the handbook only once they seem ready for the information in this resource.

In addition, the intent is not for parents to sit down at one session and read the whole handbook, but rather for it to be a teaching tool read at home and discussed at visits as needed. Parents should be encouraged to keep the handbook, write down their notes and questions, and bring those questions to visits. Parents can use the handbook to look up terms and concepts they do not understand or need to review.

Parents of children with sickle cell disease were involved in the development of this handbook from the very early stages. In the formal field tests with consumers, we asked for feedback on the readers' understanding of the material, usefulness and quantity of information, usability as a reference tool, completeness, message of graphics, cultural sensitivity, and overall appeal. Field testing was done with individuals and focus groups. More than 30 families throughout California participated in the field testing.

Reading level was an important consideration. The reading level of this book aims to: 1) not oversimplify the material, and, 2) be as accessible as possible. In trying to attain these goals, we included many technical terms and concepts. We tried to explain them clearly, however. There is also a glossary at the end of the handbook.

We recognize that some adults still may have trouble with the technical terms. However, we intend for the handbook to be used in conjunction with clinic visits. Specific information can be explained by medical staff, and we expect it to be useful to people with varied reading levels. Furthermore, the format of the handbook has been designed with many images to make it easier to read. Feedback from parents on earlier editions has been very positive.

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Chapter 1:
Basic Questions

Introduction

This chapter provides answers to many of the first questions asked by parents of children with sickle cell disease. The answers to these questions and the suggestions in this handbook will help you give your child the best start.

These questions include:

- What is sickle cell disease?
- What problems are caused by sickle cell disease?
- How serious is sickle cell disease?
- What care and treatment can make the disease less severe?
- What should you expect in the newborn period?
- What steps should you take when planning pregnancies?
- What are the most important ways to help your child each day?
- What are danger signs and when should you call a medical provider?

This chapter will teach you that with good medical care and good care at home, most children with sickle cell disease can grow up to lead full and productive lives.

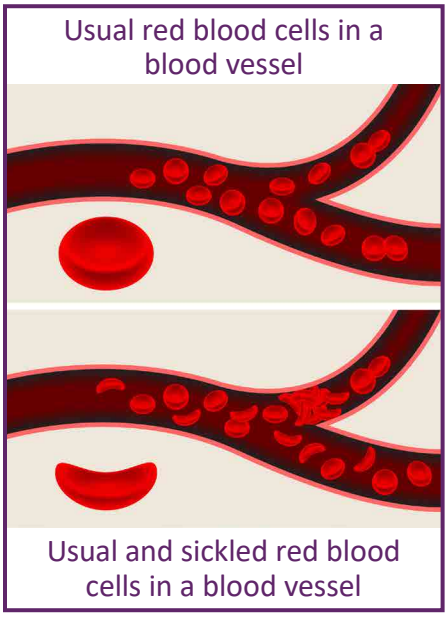
What is sickle cell disease?

Sickle cell disease is a genetic condition that is present at birth. A genetic condition cannot be "caught," like the flu or a cold.

Sickle cell disease affects a special protein in our blood called hemoglobin. You can think of a protein as a tiny machine that has a specific job. The very important job of the protein hemoglobin is to pick up oxygen from the lungs. Then it takes the oxygen to every part of the body where it is made into energy.

Hemoglobin travels through the body packaged in a red blood cell. Red blood cells, with

hemoglobin inside, are smooth, round, and very flexible (like floppy frisbees). They can fit through the smallest parts of our body to deliver oxygen and provide energy.



A person with sickle cell disease makes a different kind of hemoglobin than most people. This hemoglobin causes the red blood cells to change their shape. Instead of being smooth, round, and flexible, the cells become hard and sticky. Their shape looks like a banana or a "sickle." A sickle is an old-fashioned hand tool used to cut tall grass. We call these blood cells "sickled" or "sickle cells." The sickled shape of the red blood cells gives "sickle cell" disease its name.

The hard, sticky sickled red blood cells have trouble moving through the small tubes that carry blood to all parts of the body. These tubes are called blood vessels. Sometimes the sickled cells temporarily clog up blood vessels. This results in blood not being able to pass through and bring oxygen to all parts of the body. Too little oxygen can cause pain and damage to these areas.

See Chapter 13, How Sickle Cell Disease Is Inherited, for information about how sickle cell disease is passed down from parents to children.

What problems are caused by sickle cell disease?

Sickle cell disease can cause many kinds of problems. Some of the most common problems are:

- Infections
- Pain
- Anemia (low red blood cell count)
- Damage to body organs (such as the brain, lungs, kidneys, and eyes)

Not everyone who has sickle cell disease will have all of these problems. In fact, many people with sickle cell disease will feel well most of the time. However, most people with sickle cell disease will have to deal with one or more of these problems at some time during their lives.

Infections

Infections are one of the most serious problems that people with sickle cell disease have. Pneumonia and other infections pose a special problem for people with sickle cell disease. They can get very sick or even die if they don't get treatment quickly.

These infections are caused by problems with the spleen. The spleen is a large organ in the body that removes damaged red blood cells and helps fight infections. One of the spleen's jobs is to create proteins called "antibodies" that fight bacteria. These antibodies are released into the bloodstream to stop infections anywhere in the body.

Another of the spleen's jobs is to quickly filter and kill bacteria from the blood. In sickle cell disease, the sticky sickle cells can clog the spleen so it can't do its jobs. This leaves the body open to infection.

Parents can learn to check their child's spleen

See Chapter 7, Common Medical Problems, for more information on learning to check your child's spleen.

Thankfully, we can prevent many infections in people with sickle cell disease by giving babies and young children penicillin every day. This happens until they are at least 5 years old. Penicillin has been used since 1986 and is very effective. It has dramatically decreased infections in people with sickle cell disease without significant side effects.

Two other actions have dramatically reduced serious infections in early childhood. The first is giving children with sickle cell disease vaccines against dangerous infections, including the flu. The second is the practice of responding quickly to a fever of 101°F and greater. There are many effective ways to treat infections once they happen, especially if they are found quickly.

Pain

As mentioned earlier, the hard, sticky, sickled red blood cells can cause pain by blocking blood flow and preventing oxygen from being delivered to all parts of the body. This is the source of the pain that comes from sickle cell disease. Later sections of this book describe ways to prevent cells from being sickled (sickling), which cuts down on pain.

Some sickle cell disease pain can be very severe and needs to be treated in the hospital. Most pain is milder and can be handled at home. Later sections of this book describe ways to help your

child deal with pain, including medications that prevent pain or make it less likely. Although there are many ways to treat the pain to make your child feel better, having the pain can be upsetting.

Sickle cell disease pain is sometimes called a "crisis." That term should not be used anymore because it could be taken to mean that someone with sickle cell disease is at fault for the pain, which, of course, they are not. A better term is "vaso-occlusive episode," abbreviated "VOE," to describe these pain events.

Anemia (low red blood cell count)

Sickled red blood cells do not live as long in the body as normal red blood cells. The sickled cells may break down when they're traveling in the blood stream or are pulled out of the blood by the spleen.

The spleen and liver break down these cells, and the hemoglobin inside them, faster than normal red blood cells. When the hemoglobin is broken down by the spleen, it forms a chemical called "bilirubin." Bilirubin can cause jaundice, a yellowing of the eyes or skin.

In a person with sickle cell disease, the body tries to produce more red blood cells to make up for the short lifespan of the sickled cells. However, the body cannot make enough new red blood cells to replace all of the cells that have been broken down and removed. This results in anemia (low red blood cell count).

When blood from a person with sickle cell disease is tested, results from a laboratory (lab) test show a low hemoglobin value and a low red blood cell count. A sickle cell disease complication or other illness can make the usual lab results for a person with sickle cell disease drop quite a bit more. This low red blood cell count can become hard for the body to handle. If this happens, your child may get extremely tired and need a blood transfusion to prevent heart failure and other problems.

Organ damage

While children can develop organ damage, usually it happens later in life. Over many years, lack of oxygen from clogged blood vessels can damage organs, including the brain, lung, kidney, heart, or bones.

Not all organ damage can be prevented, but most of it can. With regular screening, early treatment, and good self-care, organ damage can be minimized. It is important to have regular screening tests and check-ups to find and prevent organ injury even if your child seems very healthy. Regular appointments will have a major impact on your child's health and quality of life as an adult.

Sometimes the sickled red blood cells clog small blood vessels and damage parts of the body without causing pain or symptoms. This is called "silent injury." This damage can happen even when your child seems fine.

Watching out for this silent injury is important. It is the main reason that your child needs to have regular visits with medical providers for screening and monitoring even when they are feeling healthy. The main goals of these visits are to find, prevent, and treat early signs of damage that your child can't feel.

Later sections of this book describe preventing and treating these problems in more detail. It is worth repeating that not everyone with sickle cell disease will have even the most common problems. Many people living with sickle cell disease have very few problems. They may go years without pain or without needing to stay in the hospital overnight. Only a few people with sickle cell disease have a lot of pain or need to go to the hospital often.

For the most part, children and adults with sickle cell disease can lead normal, full lives. Their lives can include careers, marriage, and having children if they wish. While they may miss school or work or go to the doctor or hospital more often than others, their lives can still be happy.

How serious is sickle cell disease?

Sickle cell disease is a long-term (chronic) disease that can be treated and possibly cured. Children living with sickle cell disease are normal children. They and you (the parent and caregiver) just need to pay close attention to their health. They live with a chronic disease, similar to children with asthma, cystic fibrosis, diabetes, and many other diseases.

Sickle cell disease affects different people in different ways. Out of 100 children with sickle cell disease, somewhere between 10 to 30 will have health problems that interfere with their daily lives. No one can predict if your child will be someone who has these health issues or how serious the disease will be for your child. But we do know that three things can make a difference:

1. The type of sickle cell disease (see next section)
2. The care a person gets
3. How the person and the people around them deal with the disease

Types and severity of sickle cell disease

There are many different types of sickle cell disease. The table below shows the three most common types and their severity.

Table: Three most common types of sickle cell disease

Type	Known as	How severe
Sickle cell anemia	**SS disease**	Is the most severe type, with lower hemoglobin values and more pain
Sickle "C" disease	**SC disease**	Is often less serious than sickle cell anemia; patients with this type of disease are usually, but not always, only mildly anemic
Sickle beta thalassemia disease	Either **S beta zero thalassemia (SB^0) disease** or **S beta plus thalassemia (SB^+)**	S beta zero is like SS disease but S beta plus has generally milder symptoms

The names of common types of sickle cell disease can be shortened to "SS," "SC," "SB^0," "SB^+". Other types of sickle cell disease are less common.

The different types of sickle cell disease can cause more or less severe illness. However, it's important to remember that there are some people with types that are usually less severe that end up having a lot of sickle cell disease problems. We can't completely predict how many symptoms or complications a person will have just based on their type of disease. Within each type of sickle cell disease each person can have a different experience. There are other genetic factors that affect how severe the disease is too.

It's important to understand that hemoglobin type is not the same as blood type (such as type A, B, or O). People with any sickle cell disease type can have any blood type.

Care and treatment that can make disease less severe

Nearly all newborns with sickle cell disease who receive the best medical and home care will survive to adulthood (95 out of 100 people with sickle cell disease or 95%). With comprehensive care, people with sickle cell disease can live to old age.

For children with sickle cell disease to stay healthy, they need to do the following:

1. Receive comprehensive care
2. Take certain medications and consider other treatment options to make it easier for their body to handle the disease

Comprehensive care. Comprehensive care means a person with sickle cell disease has a team of people who monitor and support their mental, physical, and social health and well-being.

The medical team should include your child's regular doctor and a doctor who is an expert in sickle cell disease (a hematologist). Check-ups with a regular doctor or pediatrician are needed for preventive care, like getting penicillin and shots for your child. Regular visits to the hematologist are needed to pick up early signs of damage.

The hematologist may work at a sickle cell disease care center. This sickle cell disease expert regularly checks for sickle cell disease complications in order to prevent them. Monitoring signs and providing early treatment can really make a difference in your child's life. The medical team at the care center may also include other medical specialists, like cardiologists (heart doctors).

Your child's comprehensive care team should also include social workers, teachers, or guidance counselors, and nurses, genetic counselors, and specialists like "neuropsychologists" to provide mental health and extra school support.

Other critical members of the team include parents, grandparents, friends, relatives, or any other caregiver who is supporting the child at home. Your child's caregivers, medical team, and other professionals should work together to maximize your child's health.

See Chapter 2, Routine Medical Care, for more on comprehensive care.

Treatment options. All people with sickle cell disease should consider therapies that prevent the red cells from sickling or help decrease symptoms from the sickling. Talk to your child's doctor about treatments. Treatment options include:

- Hydroxyurea: this is a safe medication taken by mouth that helps lower red blood cell sickling. Hydroxyurea helps most people who take it have fewer disease symptoms and stay healthy. In studies, children taking hydroxyurea have half as many pain events, hospitalizations, and transfusions, as well as better growth than those not taking it.
- Other treatments: L-Glutamine (Endari), Voxelotor (Oxbryta) and Crizanlizumab (Adakveo) are the names of additional drugs that are beneficial. These drugs may decrease complications from sickle cell disease alone or with hydroxyurea.

▶ Stem cell therapies:

- Stem cell (bone marrow) transplant: Healthy stem cells, often from a relative, are given to the child. This is for children with sickle cell disease who meet certain medical requirements.
- Gene therapy: The child's own stem cells are modified to make healthy red blood cells, rather than getting stem cells from another person. As of 2024, there are now two FDA-approved gene therapy treatments called Casgevy and Lyfgenia.

See Chapter 10, Research and Treatment, to find out more about approved treatments and potential cures for sickle cell disease.

What should you expect in the newborn period?

The newborn screening process

All newborns in California are screened at birth for sickle cell disease and other diseases. Screening is a tool that identifies newborns with an increased chance of having a disease. During screening, a small amount of blood is taken from the baby's heel a day or two after they are born. The blood is then sent to a laboratory (lab).

Lab results may strongly suggest that the baby has sickle cell disease. This result is shared with the doctor and family. Another blood test is needed to confirm the results.

The baby's medical provider will share test results and tell parents what type of sickle cell disease their child has. The goal of knowing about sickle cell disease while your baby is so young is to get your family counseling and comprehensive care immediately. A hematologist, or sickle cell disease expert, and public health nurse probably will be involved as well.

Why your newborn seems healthy if they have a disease

While babies are developing in the mother's womb, they are called fetuses. The fetus makes a special type of hemoglobin, called "hemoglobin F," or "fetal hemoglobin." Fetal hemoglobin does not sickle, even in a fetus with sickle cell disease.

At birth, the amount of fetal hemoglobin is still high. Since the amount of sickle hemoglobin is low, newborns will have no signs of sickle cell disease.

It often surprises parents and relatives that their newborn with sickle cell disease looks so healthy.

However, after birth the amount of fetal hemoglobin decreases, and the sickle hemoglobin increases. By 10 to 12 weeks of age, babies have enough of the sickle hemoglobin to develop sickling problems.

At this young age, most babies have few signs or symptoms of sickle cell disease. However, the hemoglobin level is dropping, leading to anemia. The sickle cells have already begun to clog the spleen, placing the baby at risk of infection. By three months of age, the spleen can't fully filter and clean bacteria from the blood. Starting penicillin before this period reduces the risk of infection.

What steps should you take when planning pregnancies?

Having a child with sickle cell disease means that your family will have some special things to consider around having future children. This applies to children when they become sexually active and parents. Getting information can be easy and very helpful.

We recommend speaking with a genetic counselor and getting hemoglobin genetic testing before pregnancy. These actions can help you or your child understand the chances of having a baby with sickle cell disease.

Tell your medical provider when you want to speak with a genetic counselor. It is your right. You can ask your medical provider how to get tested. People with sickle cell trait or another hemoglobin trait can also call the Newborn Screening Hemoglobin Trait Follow-up Program at 1 (866) 954-2229. They provide information and counseling.

Taking these steps will allow your family to plan pregnancies and have options to decrease the chance of having a baby with sickle cell disease if you choose.

See Chapter 12, Sex and Reproduction, and Chapter 13, How Sickle Cell Disease Is Inherited.

A summary of the most important ways you can help your child every day follows this section. It is a preview of what is included in the rest of the handbook in more detail.

We hope that you will use this chapter and the rest of the handbook to learn how to best help your child live with the sickle cell disease. Together with your child's medical team, you can help your child live the fullest life.

What are the most important ways to help your child every day?

A summary of what you can do to help your child stay healthier and happier

This section includes the most important information you should know about supporting your child with sickle cell disease at all ages. Many of these recommendations are discussed in more detail later in this handbook. Your child's needs will change as they grow.

This section covers these topics:

- Medical care
- Care for your child at home
- Well-being and mental health
- Danger signs: When to contact a medical provider

Medical care

Proper medical care can keep your child healthier and help prevent frequent illnesses. With support from medical providers, you and your child can learn to help your child feel better when they have signs or symptoms of their sickle cell disease.

▶ **Medical providers:**

- Find a regular medical provider for your child for routine care. Try to find a provider you can talk to easily.
- Find a hematologist for your child. They may work at a sickle cell disease care center. This sickle cell disease expert will watch for warning signs. They work to prevent and manage sickle cell disease complications.
- Ideally your child will have a medical team that includes people like medical specialists, social workers, nurses, genetic counselors, and psychologists for comprehensive care.

▶ **Routine care.** Get your child routine health screening and preventive care (examples are shots and penicillin).

▶ **Early treatment.** Get problems treated before they become serious. Not all problems can be prevented. But most problems can be handled if they are found and treated early.

▶ **Danger signs.** See the "danger signs" page at the end of this chapter. Take a photo or copy the page. Keep it in your phone and on your wall or refrigerator. Also keep the phone numbers of your child's regular and sickle cell disease doctors in your phone and at home.

▶ **Know when to contact your medical provider:**

- If your child has a fever of 101°F or higher or if something else is wrong with your child that is on the "danger sign" page, contact a medical provider.
- You know your child better than anyone. Reach out to a medical provider if you think something might be wrong.

▶ **Online patient portal.** Ask your medical provider to help you get on to their online patient portal. Reaching the portal on your phone could be helpful. You may need to show a new provider your child's treatment information in the emergency room, for example.

Care of your child at home

A healthy lifestyle is important for all children, but even more important for children with sickle cell disease. These recommendations will support your child's best health.

▶ **Home care:**

- Make sure your child drinks lots of water. They need more fluids than other children.
- Give your child a balanced diet. Your child may sometimes need more calories than other children. Those calories should come from good food sources of protein, vitamins, and minerals, including fresh fruits and vegetables.
- Make sure your child gets plenty of rest and sleep.
- Have your child dress warmly when it is cold.

▶ **Pain.** Help your child handle any pain they have from the disease. Give your child fluids, medications, and home remedies like hot packs. If these do not ease the pain, contact the medical provider.

▶ **Stress.** Reduce physical and emotional stress at home as much as possible.

▶ **School.** Send your child to childcare or school unless they are too sick to go or you home-school all of your children.

▶ **Activities.** Your child can participate in sports and any other activities they have the energy to do. They need to take rests when they are tired.

▶ **Sickle cell education.** Learn as much as you can about sickle cell disease. Use this handbook as a tool for education. Support your child and those who are close to your child to learn about the disease. The more you all know, the more you can get support when you ask for the care and services you need.

Well-being and mental health

"Well-being" can mean many things. It is about mental health and feelings. We use "well-being" to talk about feeling happy, healthy, socially connected, and purposeful. You can help your child and family learn how to live with the disease for better health and well-being.

- **Family.** Caring for a child with sickle cell disease is not easy. The disease affects the whole family. All family members may need support.
- **Get support:**
  - Ask family and friends for help.
  - If things get to be too much for you, talk to your medical provider, school counselor, spiritual leaders, a social worker, or a therapist. You are not alone.
  - Find out if there is a parent support group near you and join it. Other parents who have children with sickle cell disease can give you a special kind of support.
- **Ask for care and services.** Learn to ask for needs to be met at school and in medical care.
- **Stay future focused.** Children with sickle cell disease can lead full, productive, and happy lives. There are new treatments for sickle cell disease and even cures. (See Chapter 10)
- **Help your child cope with the experience of sickle cell disease:**
  - Treat your child with sickle cell disease as you treat your other children.
  - Help your child learn to take care of themselves. Children with sickle cell disease need to learn to do things for themselves and build self-esteem.
  - Teach your child to ask for the care and services they need at school and in medical care.

Danger signs: When to contact a medical provider

Call to be seen right away for the danger signs below

Call your medical provider immediately to find out where you should bring your child to be seen if your child has any one of the danger signs below. **If you can't reach your medical provider quickly, take your child to the Emergency Room or call 911.**

Fever	101°F (38.3°C) or higher
Head or neck	Severe headache or dizziness. Stiff neck.
Chest	Pain or trouble breathing
Stomach	Severe pain and swelling
Color	Loss of normal skin color, very pale or gray
Penis	Painful erection
Behavior	Seizures. Weakness or paralysis (can't move arm or leg). Can't wake up. Limping without pain.

Call for advice for the less serious signs and symptoms below

Stomach	Vomits more than once. Has diarrhea more than once.
Higher temperature	100°F that lasts more than 24 hours
Color	Jaundice (eyes or skin look yellow)
Arms, legs, or back	Pain with no other symptoms
Chest	Coughs without fever or chest pain
Nose	Runny or stuffy nose
Behavior	Acts strangely. Refuses to take medicine. Refuses to eat or drink. Less active than usual.

Medical provider's contact information: _____

Sickle cell disease provider's contact information: _____

Chapter 2:
Routine Medical Care

Introduction

Regular check-ups are a must for all children with sickle cell disease. At these visits, the doctor or other medical provider will check your child's growth and development. Your medical provider will also check to see if your child is having any health problems. A sickle cell disease specialist should also check your child regularly for early damage from sickling.

This chapter covers early childhood, primary school years, and teen years. Some of the information on routine medical care does not change as your child gets older. Some information applies to children of certain ages.

In this chapter, you will learn about the following areas:

- Where to go for care
- Well-child exams
- Using other doctors (specialists)
- Questions asked during a check-up
- Information to share
- Taking medication at home
- Special considerations for teen years

Different centers and doctors may use other approaches to treating sickle cell disease than what is presented in this handbook. Your medical provider knows your individual needs best. We recommend that you follow your medical provider's advice.

Where to go for care

Your child can get basic medical care from a pediatrician or a family medicine doctor. Or they can get basic care from a nurse practitioner or a physician's assistant who work under the

supervision of a medical doctor. A combination of these may work out best. Your baby's regular medical provider is called their "primary care provider."

Medical provider

Your child's regular doctor or medical provider can provide most of the care your child will need. This includes well-baby care, routine shots, and treatment for some illnesses. Your child's medical provider will refer you to a sickle cell disease care center or a hematologist for special care. A hematologist is a blood specialist. Any specialists your child visits will work together with the medical provider to make sure your child gets the care they need.

Sickle cell care center

In some places, there are special sickle cell disease care centers that have a team of experts trained in sickle cell disease. These centers do most of the research on sickle cell disease. They also train medical providers.

The medical team at a sickle cell disease care center can provide both routine medical care and special care for sickle cell disease. Besides doctors, nurses, and nurse practitioners, this team may also include genetic counselors, social workers, psychologists, and others who can help you and your child. This team approach is called providing "comprehensive care."

Getting a complete evaluation

To help your regular medical provider give your child the best care, we suggest that your child start with a complete evaluation at the nearest sickle cell disease care center. This should happen as soon as your child is confirmed to have sickle cell disease.

The medical team at the sickle cell disease care center will share the results of this evaluation with you and your child's regular medical provider. The sickle cell disease care center will also let you know how often they think your child should see them either in person or using a video chat or phone call.

Learning from your medical team

Medical visits are a time for you and your child to learn more about sickle cell disease. Get to know the medical team and learn from them at each visit. Learning as much as possible will help you to care for your child to the very best of your ability. It will also allow your child to play a major role in their own care as they get older.

An important part of every visit is to get answers to questions you or your child have. Be sure to ask your medical provider about anything that you or your child don't understand or find concerning. No questions are silly or stupid. All of your questions are important. The staff will be happy to answer them.

It may help to write down questions or concerns before your visit. Then you can check your notes to make sure that you remembered everything. When your teen or young adult is old enough to go to their visits alone, you can teach them to write down their own questions and to review them before the visit.

Well-child exams

During childhood, your child will have many check-ups. From birth to 6 months old, your child should be checked once a month. You and your family should be taught and should learn a lot about sickle cell disease during those early visits.

Between 6 months and 1 year of age, your baby should go to their medical provider every 2 months. After 1 year of age, visits should be set up for every 3 to 4 months until your child is 8 years old. If your child is sick, they will need to be seen more often.

The below table is just a guide. Your child's medical provider may have a different schedule depending on your child's specific form of sickle cell disease.

Table: How often your child should see a medical provider for a regular check-up

Age	How often
Birth - 6 months	Every month
6 months - 1 year	Every 2 months
1 year - 8 years	Every 3-4 months
9 years and older	Every 4-6 months

Sometimes these visits might be done by phone or video online. However, most of them should be in person. At these doctor visits, the medical staff will check your child's:

- Temperature
- Heart rate and breathing rate
- Height and weight
- Blood pressure (when they are old enough)
- Head size (for babies)

The medical provider will also check your child's:

- Eyes, ears, mouth, and throat
- Soft spot on the head (until one year of age)
- Neck
- Heart and lungs
- Abdomen
- Spleen size
- Skin
- Penis and scrotum or vaginal area (pelvic and breast exams are needed at the start of puberty)
- Joints
- Back
- Nervous system

These scheduled visits are a very important way to keep track of the spleen size and hemoglobin level that your child has when they are well. When a child with sickle cell disease is sick, it is important to know if their spleen has gotten much bigger or if the spleen size is the same as when they are well.

Similarly, each child has a usual hemoglobin level. Some children may have a usual hemoglobin level of 6 grams per deciliter (g/dL) and others may have a usual hemoglobin of 9 grams per deciliter (g/dL). When your child is sick, it's important to compare the hemoglobin level during their sickness with the hemoglobin level they have when they're well. This is to find out if there is a big change.

Do not be afraid to ask the medical provider any questions. Your medical providers are there to help you understand your child's health.


See the online Appendix for sample Comprehensive Sickle Cell Disease Care Plans. They describe what should be checked at different ages.

Shots (vaccinations)

Shots (vaccinations) are a very important way to protect your child's health. Vaccines work in people with sickle cell disease. Those with sickle cell disease do not have an increased chance of side effects from vaccines.

Because children with sickle cell disease get more infections than other children, they need these shots even more than other children. Your child will get the same shots that other children get. They will also get additional shots to help them fight the type of infections that are particularly dangerous to children with sickle cell disease.

Your medical provider will go over the shots needed and the timetable for those shots. They are necessary to keep your child safe and healthy.

Many medical providers have shot records kept on the computer. These make it easier to track what shots have been given and which ones still need to be given. However, you should also keep a record of your child's shots. Bring this record with you whenever you take your child in for medical care.

When it comes to COVID-19, children with sickle cell disease are a high-risk group. It is important to get all COVID-19 shots and boosters as soon as they are offered. Talk to your child's medical provider about this.


See the online Appendix for links to charts on what shots to get and when.

Common tests and evaluations

When your child goes to the medical provider's office, they may be sent to the lab for blood and urine tests. Or they may have these tests right there in the office. For a blood test, providers will take some blood from the finger or arm with a small needle.

This may hurt like a little pinch. If a urine sample is needed, the nurse will explain how to get it. The following are some of the most common tests that are done on the blood sample:

Hemoglobin electrophoresis. This is the test that is used to find out a person's hemoglobin type – A, S, C, beta thalassemia or another type. It tells you what type of sickle cell disease your child has.

This test is also done when it is important to know how much sickle hemoglobin (hemoglobin S) is in your child's blood. It may be done before your child receives a blood transfusion. A transfusion is when your child receives donated blood. The blood is put into the body through an intravenous line (IV).

Before a blood transfusion, medical providers use the test to help decide how much donated blood should be given. After a transfusion, the test is used to see if enough blood was given to lower the amount of sickle hemoglobin and prevent sickling-related complications.

Complete blood count (CBC). The complete blood count (CBC) is the most common blood test. It is done to find out the number, shape, and size of the red blood cells and the hemoglobin level. It also finds out the number of other blood cells (such as white blood cells and platelets). This information is used to tell if any treatment is needed.

The usual hemoglobin level in children without sickle cell disease is 11 to 14 g/dL. Children with sickle cell disease usually have a lower hemoglobin level of 6 to 10 g/dL. This varies with the type of sickle cell disease. If your child's hemoglobin level is less than 5 or 6 g/dL, they may need to be given blood or may need to go to the hospital to be observed.

Reticulocyte (Retic) count. Reticulocytes are young red blood cells. The number of these young red blood cells shows whether the bone marrow is doing its job of making and releasing young red blood cells into the blood. Bone marrow is the soft part inside bones where blood is made.

Kidney and liver function tests. These tests show if the liver or kidney have been damaged by sickle cell disease. In young children, this type of damage is rare. However, over time, the sickle cells can plug up the small blood vessels damaging the liver, the kidneys, or both.

Blood chemistry tests. These tests measure substances in the blood that are important for health and growth. These substances include iron, glucose (sugar), and minerals.

Urine sample tests. Urine sample tests can show if there are certain problems. The urine sample is analyzed in the following way:

1. To see if it has a lot of protein or blood. These can be a sign of kidney damage.
2. To see if there is an infection. With a bacterial infection, urine is cloudy, smells bad, and can have tiny bacteria and white blood cells.

Common medical evaluations that are not blood or urine tests

Many of the common medical tests ordered on babies and young children with sickle cell disease are also ordered on older children. However, there are a few tests that are mainly done in older children. The tests and the age when they are done are described below.

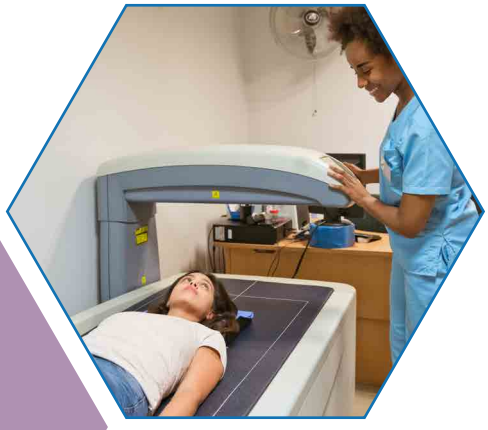
Pulse Oximetry. This test is done on children of all ages to find out how much oxygen is getting from the lungs to the blood. In the hospital or clinic, a plastic band is put on a finger or a toe. A machine then measures how much oxygen is in the blood. If your child is in the hospital with pneumonia or acute chest syndrome or other lung problems, they will be attached to this machine so the test can be done continuously. The same type of testing can be performed with a small device at home.

Transcranial Doppler (TCD). This is a painless test in which a plastic probe is placed on the head. It records changes in sounds waves that measure blood flow in the brain. It shows if the important blood vessels are narrowed or clogged. This is a screen for stroke. All children with sickle cell anemia (SS disease) or sickle beta zero thalassemia (SB^0) disease should have exams every year beginning at 2 years old. They should continue until they are 16 years old.

Pulmonary Function Tests (PFT). These tests check how well your child's lungs are working. Your child puts a plastic mouthpiece in their mouth and takes deep breaths in. They blow out into a machine that measures how the lungs expand and take in oxygen. Starting at around 6 years old, these tests should be done regularly if your child has a history of lung problems. These problems include asthma, pneumonia, or acute chest syndrome. It is very important for your child to have this testing done if they have had to take medicine for any breathing problems, including asthma. The results of these tests will help the medical providers figure out the best medicines and treatment to help your child breathe more easily.

Magnetic Resonance Imaging (MRI). MRIs take a picture of the inside of the body using a computer. No X-rays are involved. MRIs require a person to lie still in a big plastic tube for up to an hour (there can be short breaks if a person needs it). As the picture is being taken in the tube it is very loud, with big clunking sounds.

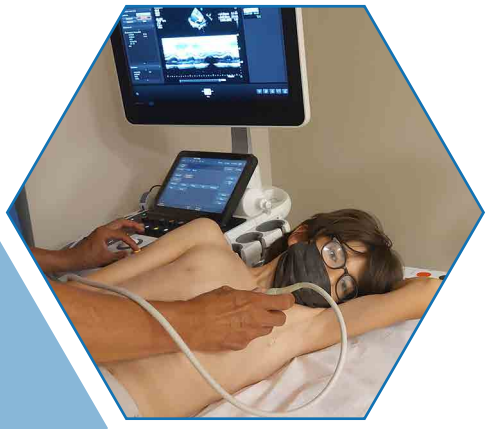
Once your child is old enough to lie still in the MRI machine, this MRI test will be done at least once to look at the brain for any damage from sickle cells. MRIs can also look at the hips or other bones or joints. Sometimes children can listen to music or watch a movie during the test. If this test needs to be done in younger children, they need to be placed under anesthesia so that they sleep through the test since they need to be still for it. In addition, the MRI is used to measure how much iron is in the liver and organs.



Bone Density. This test is sometimes called a DEXA Scan. It measures bone density by finding out the amount of calcium in the bones. For the test, your child will lie on a comfortable padded table. The arm of the instrument moves above to scan the spine and hip.

Many people with sickle cell disease have a low bone density. They need to take calcium tablets. We recommend doing this test in all children around the age of 10 years old to make sure their bones are strong. The test finds out if they need to take calcium. Providers check vitamin D levels regularly in the blood tests. They recommend vitamin D for anyone whose levels are low. Taking vitamin D has been associated with improved bone health and density. This reduces long-term pain and improves quality of life.

Electrocardiogram (EKG). This is a test that measures the electrical activity of the heart. The test is performed by placing several stickers on the chest. Wires are attached to the stickers. There are usually 12 stickers, but the number can vary. The wires are then attached to a big machine that reads out the electrical activity of the heart. It is usually done on people lying on their back. It gives the medical provider information on how the heart is functioning.



Ultrasound. Ultrasound can be done on many different body parts to find out if something is wrong. An ultrasound uses sound waves to look at structures in the body. A technician places warm gel on the skin above the area being pictured. They use a plastic probe to take the picture with sound waves. The images are not as clear as with an MRI.

Echocardiogram (Cardiac Echo). This is a test for how well the heart works. It is an ultrasound of the heart. Some warm gel is placed on the chest. The technician uses a plastic probe to take a picture of the heart with sound waves. It is used to find out if the heart is working too hard to pump blood through the lungs. This could happen because of lung damage. Lung damage can happen from sickle cell disease.

X-rays. X-rays are used to see if there is an infection in the child's lungs. They are also used to look for bone damage caused by sickle cell disease. Getting an X-ray is similar to having your picture taken with a camera. The X-ray machine is bigger than a regular camera though. It uses a much bigger machine. The technician will position your child in whatever way is needed to get the best picture.

Using other doctors (specialists)

Hematologists are blood specialists who are experts in sickle cell disease. A hematologist may be the specialist your child sees most. Your child will also be sent to other doctors for special exams. Two of the most important other "specialists" your child may see are the eye doctor and the heart doctor.

Ophthalmologist (eye doctor). Starting at 10 years of age, your child should see an ophthalmologist who knows about sickle cell disease once a year. The eye doctor dilates the pupil to check for damage from sickle cell disease in the back of the eye. This is not the same as a vision screening.

Cardiologist (heart doctor). If your medical provider is concerned about heart problems, the heart doctor checks to make sure your child's heart is healthy. Sickle cell disease may cause one side of the heart to become large. The heart may need to work harder than normal and that can cause problems over time. The earlier this is found, the earlier it can be treated.

See the online Appendix for descriptions of other types of medical providers.

Questions asked during a check-up

During medical exams, the medical providers will usually ask you for information about your child. Your answers will help them learn more about your child's health, development, and habits. Try to answer the questions fully so that they can give your child the best care. You may want to write down your answers to the questions here before you see your medical provider.

Here are some examples of the kinds of questions you may be asked:

Question: What does your baby do? Smile? Roll over? Talk?

Reason for question: It is important to know if your baby is developing normally.

Question: Has your child had any fevers, colds, or jaundice (yellow eyes or skin)?

Reason for question: Your medical provider needs to know about any illnesses you may have treated at home or had treated elsewhere. They are trying to learn about the general health of your child. It is very important to always contact or see your medical provider right away if your child has a fever, jaundice, or seems sick. This question is also a way for your medical provider to make sure you understand to contact them when your child is having symptoms that could be dangerous.

Question: What is your child eating?

Reason for question: This helps the medical staff know if your child is eating right. Sometimes children eat too much, too little, or the wrong foods. If your child is having problems with weight or food, the staff can help you plan a healthy diet.

Question: What medicine is your child taking at home?

Reason for question: Your medical provider needs to know the kinds and amounts of medicines you give your child. This includes both prescribed medicines and those you buy over the counter at the store. Try to bring photos of all your child's medications to every clinic visit. Be prepared to discuss your child's dosage if you can. It may help to take photos or write down a list. Bring them to clinic visits.

Question: Is your child having any problems taking prescribed medicine, like penicillin, at home?

Reason for question: Your medical provider needs to know if you or your child is having any problems with medicine. For example, you may have trouble getting the prescription filled or getting the child to swallow it. If there is a problem, tell the staff so that they can help you figure out a way to make sure your child gets the medicine they need.

Involving your child in their own health needs as they age

As they grow, children must learn to play an active role in their own medical care. Even young children can answer some questions about themselves. Your medical provider should ask your child some questions. If not, you should direct the provider's questions to your child.

As your child gets older, your child will be able to do more and more things on their own, including some things on this list.

Younger children

- Drink water and other fluids regularly
- Take breaks and rest when feeling tired
- Learn the danger signs for sickle cell disease complications (such as fever or pain)
- Know what medicine they take and be able to remind someone when they need it
- Speak up at medical visits by asking questions and giving answers

Talk to younger children about sickle cell disease so that they can tell others about it.

Older children

- Contact their medical team by themselves about concerns
- Take their medicine (until they are old enough, you still need to track how much they take)
- See the medical team alone, either with an adult in the waiting room or not, depending on the child's age
- Learn to manage mild pain by taking acetaminophen (Tylenol) or ibuprofen (Advil or Motrin), drinking extra fluids, using using heating pads and other home remedies, staying busy or relaxing
- Keep a diary about pain at home to share with the medical provider so that they can see how well treatment addresses pain and quality of life
- Speak to a genetic counselor and other specialists about reproductive choices and their health

Older children can write a report for school, read books, and ask their medical providers for more information.

Information to share

Many things can affect your child's health. The more your medical provider knows about your child's life, the better care they can provide. Be sure to tell your medical team at the sickle cell care center if any of the following things apply.

Your child has been treated by another doctor or clinic. Your child's medical provider needs to know about what problems your child was having. They need to know what treatment or tests were done. If any medicine, shots, or tests were given, your child's medical provider should know so they do not repeat them unless needed. They can give you an Authorization for Release of Medical Information Form. This form says you are okay with getting a copy of the records from other places added to your child's medical record to make it complete.

See the online Appendix for a sample of an Authorization for Release of Medical Information Form.

Your child looks or acts in a way that concerns or upsets you. You know your child better than anyone. Your medical provider may not see things during the visit that you may see at home. Help your medical provider by sharing things that upset or concern you or your child. If your child keeps a pain diary, they can share that.

Your child has had a major loss. This could include divorce or the death or illness of a family member or friend. Emotional upsets can affect the body. If your child is going through a stressful time, your medical provider may want to watch them more closely.

You or your child are upset by something that happened at the clinic or hospital. Talking about problems can help lead to solutions. Your medical provider may be able to help you make sure something doesn't happen again. You may find a better way to handle a problem by talking about it.

Your child is having trouble in daycare or school. Here, too, talking about a problem may help you solve it. Teachers or daycare staff may need to learn more about sickle cell disease so that they can better understand your child. Your medical provider will be able to help you get the right information for them.

You are planning to take a trip. Ask your medical provider to give you a letter stating:

- What kind of sickle cell disease your child has
- Your child's normal hemoglobin level
- The medication your child takes
- Other special problems
- Who to contact in case of an emergency

Take the letter with you when you travel with your child. Also, ask your medical team about where you should take your child if they need medical care on your trip.

See the online Appendix for a travel letter template you can use to ask your doctor for a letter.

You are planning to move. Ask the medical staff for information. They can give you the name of a sickle cell disease care center or another doctor near where you will be moving. Your medical team can send a copy of your child's medical records to your new medical team.

You may be staying within the same medical system when you move. Your medical system may have an online patient portal that you can use to look at medical records and contact the provider. Sign up for it. It is a very useful tool, especially when you change doctors within the same system. Usually, you can look at the portal on a smartphone as well as a computer.

Special considerations for teen years

The teenage years are both a rich and challenging time as children become more independent. With puberty, medical needs change. Pain, fatigue, and other sickle cell disease symptoms can worsen. Teens can also start taking more responsibility for choices that affect their own health.

See Chapter 12, Sex and Reproduction.

Taking care of their own health needs

Your child may have been involved in their own medical care from a younger age. They may have asked and answered questions with a medical provider, for example. As your child grows, their involvement should also grow.

Your teen should know the name of their condition (for example, sickle cell anemia or SS disease). They should know what kinds of medicines they are supposed to take every day, at what doses. They should know the steps to take when they are having pain, what triggers their pain, and how to prevent pain when they can. They should also know what steps to take when they have a fever.

You should support your teen to keep track of medical appointments and to start to make their own appointments. You should still make sure they stay on schedule with appointments, however.

You can go to appointments, but your teen should see the medical provider alone for part of the appointment. You can still fill prescriptions and monitor that your child is taking their medicines regularly.

By the time most teens with sickle cell disease are seniors in high school, they can fill prescriptions, make medical appointments, and get to the appointments on their own. However, many teens still need their parent's support with these activities.

Every person is different. Teens are different from one another regarding when they will begin taking care of their own medical care. Some people with sickle cell disease will continue to need support with medical needs into adulthood.

Most people with sickle cell disease should be able to take care of their health and home care needs once they are adults.

Even if your teen is taking care of his or her own health and medical care, they may still want you to be involved. Check and see how things are going from time to time. Once you've stopped taking care of all your teen's medical needs, it can still be nice to help them out sometimes.

Finding out more about adult medical settings

Talk with your teen about adult medical care options. Talk about what things will be the same or different in the adult medical care setting. One difference is that pediatric hematologists (blood specialists) sometimes, but not always, can be both the child's primary doctor and also provide specialized care. Adult hematologists often will not provide primary care. Your teen or young adult will need to have a primary care doctor along with a hematologist.

A key issue is that the primary care provider should be in close communication with the specialists, including the hematologist. The providers should work together as a team.

Moving from a pediatrician who sees children to a medical provider who treats adults can be hard. One hard part can be finding a doctor for adults who has knowledge and experience helping people with sickle cell disease. If you or your child have this problem, it is best to just find a doctor your teen likes. Your teen should be comfortable talking to them. The medical provider should listen without judgment. They should be willing to learn about the disease.

Moving to an adult medical setting

Helping your teen settle into an adult medical setting is important. This process generally happens between 18 and 26 years of age. It is a gradual process. As discussed above, the preparation for this transition is throughout the teenage years. Your teen should visit the doctor in the adult medical setting ideally within 2 to 6 months after the last visit with their regular pediatrician or medical provider. The visit can happen by phone, video, or in person.

It is best when the pediatrician and the new adult medical provider have a "warm handoff." This means they either speak to each other on the phone, communicate by email, or, even better, have a clinic visit together with your teen and both medical providers to make the transition smooth. Both medical providers should confirm with each other when your teen has formally transferred from one care setting to the other. Your pediatrician should also continue to be available to answer questions for the adult doctor.

Your teen and your family should be able to tell the doctors whether you are satisfied with the process of moving from one care setting to the other. The doctors should learn whether you are satisfied with the adult medical setting itself.

Medical care privacy and consent

Before your child is 18 years old, they require a parent or guardian to be with them for major medical decisions or to consent to surgery or transfusions. Once your child is 18 years old, medical providers cannot talk with you about your child without your child's permission. So, by 18 years old, it is important that your child can communicate independently.

Saving medical documents

Your child will have important medical records. These can include their shots and medical history. Keep these records together in a folder, either a paper folder or a folder on your computer. Both you and your teen should sign up for the medical provider's online patient portal if they have one. Make sure your teen knows about the records folder or has access to the portal. Your teen should learn to manage and update the records. They should put documents from visits into the folder.

Preparing to leave home

Give your child a copy of everything in their medical records folder when they leave home or go away to college. Make sure they have signed up for their medical provider's online patient portal before leaving.

Many college-bound teens can benefit from asking for special accommodations through student services at the campus. Their sickle cell disease team can help them with these requests.

Talk to your child about financial resources. Discuss health insurance, medical care related expenses, and expenses related to daily living away from home. Most employers allow children to stay on their parents' health insurance until the age of 26. Your teen should start thinking about how and when they might be able to support themself or contribute to your household.

See Chapter 4, Well-being and Mental Health, for more information on managing the teen years.

See the online Appendix for information on what health issues your child may expect as an adult.

Chapter 3:
Care at Home

Introduction

Children with sickle cell disease need some extra care at home to help them stay well. Giving your child penicillin twice a day and hydroxyurea every day as prescribed are both important parts of good home care for children with sickle cell disease.

Extra water and other fluids and a good diet also help to keep your child well. With just a little extra care, your child can be active, go on trips, and do almost all the things that children without sickle cell disease do.

This chapter covers home care, including taking medicine, fluids, nutrition, staying active, and traveling. It does not cover easing pain at home.

See Chapter 5, Basics on Fever, Pain, and When to Get Help, on easing pain at home.

Taking medications at home

Give your child medicines as your medical team tells you. Involve your child in taking medicine as they get older. Make it a game or put your child in charge of how they take it. Even a 4-year-old can help remind you that they need to take their medicine.

Penicillin

Your provider will start penicillin for your child as a very young baby. Make sure that your child keeps taking penicillin at home twice a day until your medical provider says to stop. All children continue penicillin until age 5.

Whether your child will take penicillin after age 5 depends on your medical team's evaluation of how much your child is at risk for infections. If your child has had a lot of infections or had their spleen taken out, your child may need to continue penicillin for longer. Some people continue penicillin their whole life.

Your child can take either liquid penicillin or a penicillin pill. It is better to give penicillin in pills because they don't need refrigeration and don't expire as quickly as liquid penicillin. Liquid penicillin must be

discarded after 2 weeks. Either way, penicillin needs to be taken twice a day, morning and night.

Pills can be crushed and mixed with a teaspoon of applesauce, ice cream, flavored yogurt, or other soft food. It is not good to put medicine in formula or juice because the baby may not drink all of it. Liquid can be given by spoon or dropper.

The amount of penicillin that your child should take depends on your child's age, as shown below:

Table: How often your child should take penicillin and the dose

Age	Penicillin dose	How often
2 months old to 3 years old	**125 mg**	Twice a day
3 years old to 5 years old	**250 mg**	Twice a day

As children get older, it is harder to make them take something they don't want to take. If this is a problem with your child, explain why this medicine is so important. If your child still won't take it, tell your medical team. As a last resort, they can arrange for shots.

Hydroxyurea

Once your child reaches 9 months old, you should discuss starting hydroxyurea with your medical provider. Hydroxyurea is a prescribed medicine that helps lower red blood cell sickling.

Hydroxyurea helps most people who take it have fewer disease symptoms and stay healthy.

See Chapter 10, Research and Treatment, for more information about hydroxyurea.

Other common medicines

Antibiotics

If your child has an infection, a medical provider will probably say they need antibiotics. The provider will write a prescription to be filled at a pharmacy. The antibiotics will come with instructions.

The antibiotics must be taken until there is no medicine left. Even if your child feels fine, they need to take the medicine until it is gone. Stopping early can cause the infection to come back. It could increase the chances that the infection may get resistant to the antibiotic.

Most of the time, antibiotics are taken in pill form. If it is in liquid form, check the bottle to see if it needs to be kept in the refrigerator.

Pain medicine

Common pain medicines to have at home include the following:

- Acetaminophen (Tylenol) for fever, mild pain, or both
- Ibuprofen (Advil or Motrin) for fever, mild pain, or both
- Hydrocodone for severe pain (by prescription only)
- Oxycodone for severe pain (by prescription only)

Some pain medicine can be bought in a store. Getting some pain medicine requires you to fill a prescription at a pharmacy.

Antibiotics and pain pills are not the same. Antibiotics fight infections and pain medicine treats pain only. As stated earlier, antibiotics must be taken for as long as they are prescribed.

Pain medicine should only be taken as long as it is needed to relieve pain. As soon as the pain is gone, your child should stop taking it. Pain medicine should be taken exactly as prescribed by your child's doctor.

Make sure you have received and understand the directions for the pain medicine and what to do if the pain gets worse. It's helpful to have this information written out by your doctor. It may be helpful to keep a limited amount of prescription pain pills (such as oxycodone or hydrocodone) at home. All medicine should be secured in a safe place. Medicine should be used only according to your doctor's directions.

Iron chelators

Medications called "iron chelators" are given when children with sickle cell disease need frequent blood transfusions. Chronic transfusions prevent sickling, but they can cause too much iron to build up in the body. This can damage the heart, liver, kidneys, or other organs. Iron chelators prevent and treat iron overload. Medicines called Jadenu, Exjade, Ferriprox, and Desferal (Deferoxamine) are all iron chelators.

See Chapter 9, Blood Transfusions, for more information on iron chelators.

Folate

Folate is a vitamin, not a medicine. Medical providers often prescribe folate for children with sickle cell disease. See the section on vitamins later in this chapter.

Water and fluids

Children with sickle cell disease need more water and fluids than other children because they continue to urinate even when dehydrated. They usually get thirsty more often than other children. Give your child fluids whenever they are thirsty. It is best to drink water. Try to replace fluids that have sugar in them (like soda and juice) with water.

Keep enough fluids on hand so that your child can have as much as they want. Breastfed babies should be allowed to nurse often. Bottle-fed babies should have formula or breastmilk in the bottle whenever they want it. Once your pediatrician says that your baby is old enough to have water, your baby should be allowed to drink water whenever they want it.

Special times when your child needs to drink more

Your child needs more fluids with:

- a fever
- pain
- hot weather
- activities that are very active, like sports
- travel

Your child may not want to drink a lot of fluids at these times, but they still need them. You may have to push your child to drink more fluids. Try ice chips, popsicles, jello, milk, or soup as well as water. Use the chart below to figure out how much fluid your child needs during these special times.

A baby who is breast feeding or on infant formula only needs extra fluids during special times. At these times, you should encourage them to take breast milk or formula, unless they are vomiting.

Table: Amount of clear fluids your child needs each day during special times

Child's weight	Number of 8-ounce cups per day
10 pounds	2 cups
15 pounds	3 cups
20 pounds	4 cups
25 pounds	5 cups
30 pounds	5-6 cups
35 pounds	6-7 cups
40 pounds	7 cups
50 pounds	8 cups
60 pounds	9 cups
Over 60 pounds	10 or more cups

Nutrition

Good nutrition is especially important for people with sickle cell disease. It helps promote health and prevent complications.

Children with sickle cell disease sometimes need to have more calories in their diet to make new red blood cells. They still need to eat a well-balanced diet like everyone else. This means having good food sources of protein, vitamins, and minerals every day. Eating more fruits and vegetables is helpful. It is better not to skip meals. Having more meals with smaller portions is good for developing good eating habits.

Some children with sickle cell disease, especially those with a severe type, may have difficulty gaining weight in childhood. On the other hand, obesity is a growing problem in children with sickle cell disease.

Doing something about excess weight is important because that excess weight can contribute to your child having health problems. If your child is very overweight, talk to a nutritionist who knows about sickle cell disease. They can review what your child is eating and suggest changes if needed. Speak with your social worker about resources available for children with sickle cell disease for a heathy meal if needed.

There are three common reasons why children with sickle cell disease can become overweight:

- The fear of exercise and physical activity causing pain is an important factor. People with sickle cell disease should participate in exercise. They should not be excluded from physical activities in school.
- Diet in children with sickle cell disease is often high in calories, but low in good food sources of protein, vitamins, and minerals.
- Like all children, children with sickle cell disease need extra calories during a growth spurt. After their puberty-related growth spurt, the amount of calories they need decreases. Children may gain too much weight if they continue to eat like they did when they needed more calories.

Vitamins

When your child is about a year old, they may be given a folate vitamin supplement to take daily. Folate is also called folic acid. It helps the body make new red blood cells. The vitamin can be crushed and mixed with milk, juice, or food. Some children don't need extra folate. Check with the medical provider to see whether your child needs more folate.

In addition to folate, some children with sickle cell disease may need to take other vitamins and minerals such as zinc, iron, and vitamins D and E. Your doctor will prescribe these when needed.

You can also give your child a multi-vitamin **without iron**. The multi-vitamin isn't necessary but can be helpful.

Height and weight

Your child may be smaller or thinner than their siblings or cousins, even with a good diet. This is because they use more energy to make new red blood cells. As your child grows up, their height and weight should catch up.

If you are concerned about your child's weight or eating habits, talk to a nutritionist who knows about sickle cell disease. They can evaluate what your child is eating. They can suggest changes if needed. These changes may include giving your child extra snacks or more calorie-rich foods.

Fasting

People with sickle cell disease who go on fasts should talk to their medical provider about an individual plan. Religious groups allow people with health needs to change fasting plans to fit their needs.

Activity

Sickle cell disease won't keep your child from doing most activities they enjoy, as long as they take care of their health. That means:

- Resting when they feel tired
- Drinking extra fluids when they are active
- Dressing for the weather

Tell your child to rest as often as they need to. Then let them find their own level of activity and enjoy it. A few activities can sometimes cause problems related to sickle cell disease. These activities should be avoided. If your child wants to do any of these, talk to your medical provider first. These activities are the following:

- Activities that expose them to cold temperatures, such as swimming in cold water
- Activities at high altitudes, such as backpacking, hiking, or skiing

All preschool and day care playground activities and most elementary physical education class activities are safe and healthy for your child. If they are in a program where there is a leader or a teacher, tell that person about your child's need to drink extra fluids and to rest when tired. The activity leader can help your child take good care of themself.

Children with sickle cell disease may participate in sports for fun. They should avoid overexertion, overheating, dehydration, or chilling, though.

Traveling

Most travel is fine for children with sickle cell disease. There are a few rules your child should follow when you take a trip.

- Fly only in a pressurized plane. This should not pose a problem for most trips because almost all commercial planes are pressurized.
- Drink plenty of fluids when traveling. This is important when your child is:
    1. Flying in an airplane
    2. Riding in a car
    3. Visiting an area that is very dry or very hot
    4. Riding or walking at high altitudes
- At elevations over 5,000 feet, your child should drink extra fluids and rest often. If your child starts to feel sick, take them to a lower elevation.
- Dress for the weather and stay warm.

Let your doctor know if you plan to take your child on a trip and ask the clinic staff for a "travel letter."

See the online Appendix for a sample travel letter.

Make sure that you take along the penicillin your child needs. Make sure you also bring hydroxyurea and any other medicines they are taking. Talk about your plans with your doctor to see if any other special care needs to be taken.

Travel Checklist:

☐	Talked to doctor
☐	Have a travel letter filled out by doctor
☐	Filled prescription for penicillin
☐	Made arrangements for hydroxyurea and other medicines
☐	Packed extra fluids
☐	Have names and addresses of doctor or center to contact if needed
☐	Included thermometer

Chapter 4:
Well-being and Mental Health

Introduction

"Well-being" can mean many things. It can mean feeling happy, healthy, socially connected, and purposeful. Well-being includes mental health and feelings.

Taking charge can increase feelings of well-being. Now that you know more about sickle cell disease, you can take charge to help your child live a healthy life with sickle cell disease. Taking charge involves learning more about yourself and your child. It also means dealing effectively with those who are close to your child.

It's hard to be a parent, whether your child has a chronic disease or not. Always remember that there will be good times. Try to focus on the positive even if the steps forward are small.

One way to help your child is to prepare and empower them to manage their own personal and health needs. You can prepare them by always asking them to play an active role in their own care. How you can do this will be based on their age and abilities.

This chapter describes what you can do to support your child and your family's mental, emotional and social needs. It includes:

- Learning about sickle cell disease
- Acknowledging and accepting your feelings
- Helping your young child at different ages
- Supporting brothers and sisters
- Getting the best childcare
- Asking for help and support
- Managing the primary and secondary school years
- Managing the teen years

Use this chapter to help you learn better ways to help your child and your family.

Learning about sickle cell disease

For most people, knowing more about the disease helps them feel less scared and less helpless. You may find out that some of your beliefs about the disease aren't true. The disease may not be as you first thought.

You may also find out that there are many things you can do to help your child that you didn't know about. Learning about these things can make you feel more in control.

As you know more, you will make better choices for your child. You'll be better able to plan for their future. You can also teach other people how to help your child. Reading this handbook and sharing it with others will help you and your community learn about the disease.

Acknowledging and accepting your feelings

You will have feelings after learning your child has sickle cell disease. Sometimes these feelings are negative. If you have these negative feelings, that is okay and common. You may feel sad about the loss of the wished-for "perfect baby." You may feel guilty about the fact that the disease is caused by the genes you and your partner have.

You may be angry that you or your partner did not know that you both carry sickle cell trait. You may feel angry that your baby's and family's future will be changed in an unknown way. You may also feel afraid and helpless in facing your baby's future. No matter how you feel, it is okay to feel that way. Feelings aren't right or wrong. What matters is how you deal with your feelings.

The first step is to become aware of how you feel. Many people aren't aware of their feelings. But unless you know what your feelings are, you can't manage them. Instead, your feelings will control you. To better understand your feelings about your child's sickle cell disease, think about your answers to the following questions:

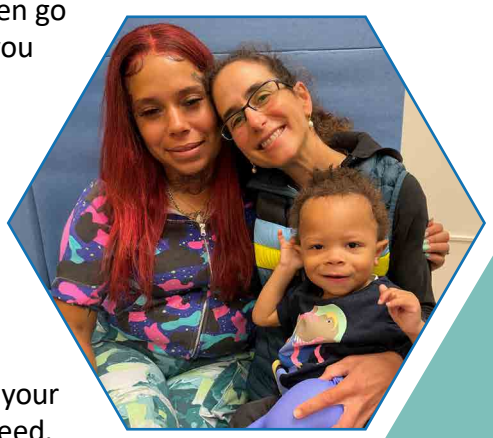
- Do you feel mad, even when there isn't a clear reason?
- Do you feel like crying more often than you used to?
- Do you feel like it's your fault that your child has sickle cell disease?
- Do you worry all the time?
- Do you want to see your friends and family less?
- Are you alone more than you used to be?

You might ask a close friend or your partner if they notice any changes in you, positive or negative. They may see things that you can't see. Also, just talking with others will help you learn more about how you feel.

Just being aware of your feelings can help you handle them. Sometimes, though, you may need to do more than just know how you feel. You may need to express your feelings, so you don't take them out on your child, yourself, or other people you love. To express your feelings, you can do the following:

- Share your feelings with your partner, family members, a close friend, or your spiritual leader
- Write about your feelings in a journal, poem, or a letter
- Find a place where you can be alone and say whatever is on your mind out loud
- Get help from a social worker, therapist, or psychologist

Sometimes feelings will change when you express them. But even if they don't change right away, most feelings do change with time. They become less strong or even go away. Learning more about your feelings and expressing them will help you feel better and be a better parent.



Even when you do all you can to help your child stay well, they may still have problems. These problems can affect your whole family. As your child grows up, your child may face new problems, like learning problems or pain. You may feel scared and angry again, even though you thought those feelings had left for good.

It is important for you to take care of yourself, as well as your child. Learn about your limits and needs. Sometimes you may need time for yourself, your other children, or your work. Ask for help so you can have the time you need.

Helping your young child at different ages

Your infant

During their first year, babies grow and change rapidly. Many babies are able to smile by 2 months. At 3 months you may notice your baby making happy sounds. By 3 months they begin to recognize their mothers and close family members. By 4 months babies may begin to be shy with strangers until they get to know them. At 7 months they can respond playfully to other persons and by 10 months they can wave bye-bye.

For the first 2 to 3 months of your baby's life, they are protected from sickle cell disease. It is very unlikely that they will have sickle cell disease complications during this time period. This will give you some time to get to know your baby and deal with your own feelings.

After the first few months, your baby may begin to have problems from sickle cell disease. The first signs of the disease may be hard for you. You may realize there is little you can do to prevent fevers, infections, or pain.

A baby has few ways to let you know if something doesn't feel right. They may cry more than usual, be fussier than usual, eat less, or be less active. Give them as much comfort as you can. When your baby isn't feeling well, your touch and soothing voice can make a big difference. If your baby is in the hospital, your presence can help make them feel better.

Your toddler

Toddlers learn fast. Because they are curious and active, they can get into dangerous places. They need opportunities for walking, running, and climbing. This is a time to make sure your home is safe to prevent accidents. Children at this age must be watched at all times.

Like other parents, you may be amazed at your child's energy and development. You will also at times become tired of caring for the active toddler.

Most toddlers do not have words to express how they are feeling. Because of this they may be happy one moment and grumpy the next. Try to be patient with this behavior. Providing the same time daily for eating, playing, and sleeping can be helpful.

At this age, your child may be very afraid of being away from you. Most toddlers feel this way, but it is important for them to spend time with other people. As your child becomes more comfortable with others, it will be easier for you to leave them with someone else.

Your toddler needs to feel secure that you love them always. Play with them, comfort them, include them in family activities and let them learn things on their own. Your child needs and thrives off of your comfort.

Your 2- to 4-year-old

Children from the ages of 2 to 4 years want to be independent. They say "no" and want to do things themselves. Let your child begin to make their own decisions. Even in the hospital, they can choose their own meals or TV programs. They can walk to the playroom with a staff member without you when they are feeling better.

At home, they can help you remember when to take their penicillin and remind you to give them drinks. Use your judgment about things they can try for themselves. Avoid doing things for them that they can do themself.

Even though your child is more independent, they still need you. Comfort them, respond to their fears, answer questions, and help them learn more about their world.

Your 4- to 6-year-old

Between the ages of 4 and 6 years, children use imagination to understand their world, including

their disease. For example, a child may believe that the pain is a punishment for something they did. Or they may believe they caught sickle cell disease from something they ate.

Pay attention to what your child may be saying about the illness. Clear up any incorrect ideas. Don't worry if you have to go over the same ideas more than once.

Let your child ask any questions they may have about sickle cell disease. Answer these questions clearly, in words that your child can understand. You may want to use stories, stuffed animals, or puppets to help you explain what will happen to them.

Even though your child is talking, they may not be able to tell you what they are feeling. Watch your child's play to get helpful information. Also, look for patterns in how your child acts when they feel sick or in pain so you can know how to help them. For example, they may want to stay in bed in the morning because they say they "feel funny" when they in fact have pneumonia. They may act listless and want to be left alone when in pain.

This is also the age to start teaching your child how to take care of themself. For example, you can explain, "You need to drink 8 cups of water today." Or you can let them do things for themself. Or you can let them pour their own drinks or get their own cup.

Because your child has sickle cell disease, you may feel they should be treated differently than other children (special treats, toys, attention, less discipline). This is not really best for the child, however.

Children with sickle cell disease need to feel that they are the same as other children. If your child has brothers and sisters, they should all have the same rules to follow. What you expect for their behavior should be the same.

Brothers and sisters

Whenever a new child comes into a family, each family member may have different reactions and feelings. For some brothers and sisters, they share their parents' joy. Others may be afraid the new baby is competition for the love and affection they usually get. This feeling can be even stronger if the new child has special needs.

Siblings who don't have sickle cell disease may feel afraid of becoming sick themselves. They may be afraid they caused their brother's or sister's illness or feel angry about the extra attention the child with sickle cell disease is getting. This is very normal and can be true even if the attention your child with sickle cell disease is getting is not fun, like having IVs, shots, and seeing medical providers. Assure your healthy children that these feelings are natural. Encourage them to come to you to ask any questions or share any feelings that they have.

Share your time and love with all your children. It is best for all family members if you make sure to spend time with each of them. Try not to miss school plays or sports events that your healthy children are involved in because your total focus is on your child with sickle cell disease.

Let your other children help your new child, or if your child with sickle cell disease is older, let that child help with younger siblings. Many parents have found that sharing care even in small ways helps with sibling relationships. Use your knowledge of your children to help you figure out how they can best be included in their brother's or sister's care.

As you learn more about sickle cell disease, teach what you learn to your other children. You can also let them ask the doctor or the medical staff questions, especially ones that you don't know how to answer. This will help them feel included.

Avoid "special treatment." Brothers and sisters of children with sickle cell disease know that these children are often treated in a special way by relatives, neighbors, and parents. Some are able to understand why this happens, but others can't. This is normal and not their fault. Try to use the same system of discipline and rewards with all of your children. This can relieve much of the resentment over one child getting special treatment.

Childcare

Childcare is another common area of concern during the early years. Like many parents, you may be worried about what the caregiver will do if your child becomes sick. It is often helpful to tell your child's childcare provider the following about your child:

- Treat your child like other children. Except for having sickle cell disease, they are the same as other children.
- Watch for signs of infections and respond. Go over a list of signs of infection, including a fever, and other problems. Make sure your caregiver knows that any temperature 101°F (38.3°C) or above is a medical emergency for a child with sickle cell disease. Make sure your childcare provider has access to a thermometer.

Make sure caregivers know how to contact you or another responsible adult who can pick up your child and take them to the clinic or emergency room if they have a fever. The childcare provider should also have your medical provider's name and phone number so they can contact the medical provider if they can't reach you. Be sure your caregiver has an Authorization for Medical Treatment Form from you. This will give them permission to get emergency medical care for your child.

- Take care of your child's special needs. Your child has a few extra needs that are very important. These needs include drinking more fluids, resting when tired, and needing more frequent trips to the bathroom or diaper changes.

Many times, caregivers or teachers will want to learn more about sickle cell disease. You can bring them this handbook. It will provide them with more information. If they have more questions, you can refer them to your child's medical provider.

Sharing all of this information with these care providers can relieve much of the stress and worry that they may have in caring for your child. It will also relieve many of your own concerns because you will know that they are prepared.

See the online Appendix for a sample Authorization for Medical Treatment Form to give to all caregivers, including babysitters.

Asking for help and support

The demands of parenting can often seem endless. When a child has special medical needs, even more involvement is needed from parents. In most cases, family, friends, neighbors, people from your place of worship, and community groups are a great source of love and support.

Help others give you the support that you need so that you don't have to do it all yourself. Ask them to watch your other children, make a meal for your family, or give you a ride. Many people will be happy to know how they can help.

It can be very helpful to talk to other parents of children with sickle cell disease or to join a parent support group. These groups have helped many parents learn ways of dealing with problems that only those who have experienced the same problems could know. Ask your medical staff about sickle cell parent groups in your area. Ask them to introduce you to another family with a child with sickle cell disease.

See the online Appendix for a list of community-based organizations that may host or know about parent support groups.

Part of learning to live with sickle cell disease is telling others about the disease and handling their questions and opinions. Some people that you have contact with may have fears and incorrect ideas about sickle cell disease. With the help of your medical staff and your own knowledge of the disease, you can teach others the truth about sickle cell disease. You can show them how they can provide support to both you and your child.

The medical team

Because you see and deal with your child's needs every day, you know how your child usually functions. You and your child's other caregivers will know before anyone else if your child is acting differently than usual. It is very important that you let the medical team know about your concerns. Teach others who care for your child to do the same.

Sharing information goes both ways. The medical team has a lot of knowledge and experience with sickle cell disease. They want to answer your questions and provide you with the support that you need.

You may find that medical providers sometimes disagree on how to handle a problem your child is having. Differing opinions are common in any type of medical care. Therapy in sickle cell disease is changing as more information becomes available. Talk to the team if this happens. You should understand the different opinions.

If differing opinions are about a major treatment, it is reasonable to request a second opinion from an expert in sickle cell disease. You can ask your doctor to recommend a doctor to give a second opinion. You can also get the names of sickle cell disease experts from the Sickle Cell Disease Association of America (SCDAA) and other sources. You can do an online search for this organization.

Sometimes you may have a different opinion than your medical provider. If this happens, share your thoughts and reasons. Share what you plan to do. Being honest about your plans will help your child and also your relationship with your medical provider. If a conflict arises between you and your medical team, talk about it so that your child can receive the best care.

Many times, problems happen when parents don't understand completely what the medical provider is saying. Or the medical provider may not understand the parents. It may help to have a social worker, community health worker, or another third person help you and the medical provider understand each other better.

To make sure that you have enough time to talk with your medical provider, you can:

- Make an appointment to talk to the medical provider. You can discuss your concerns or get more information about your child's health.
- Write down your concerns as they come up. Bring these notes with you to the appointment.
- Get to know the medical team and the roles that each of the members of the team play. Think about who could be most helpful in dealing with each problem or question.
- Get tips from other parents about ways to express your concerns to the medical team. New families can learn a lot from other families who have worked with the same team.

Take care of yourself

It can be challenging to deal with a serious illness that affects someone you love. You may find that your life is more stressful in other areas, such as your marriage, your family, your finances, and your own personal life. It is important to take good care of yourself so you can take care of your child.

Although you may want to devote your whole life to your child, you can't. Trying to do this won't help your child, and it won't help you. Your needs are also important. Throughout the ups and downs of your parenting journey, you should find time for yourself, your other children, your partner, and your friends. Make time to do things that you enjoy. A short break, even when your child is sick, can give you a lift.

Money can be a major concern for parents of children with long-term diseases. Most states have special programs to help pay medical costs, such as Medi-Cal. In California, California Children's Services (CCS) pays many of the costs of care for children with sickle cell disease. Talk to your provider or social worker about your concerns. Ask them how to get more information about medical care and insurance coverage for your child.

Be sure to get help if you need it. Reach out to medical staff, friends, family, spiritual leaders, support groups, or other parents to help you get through hard times. Sometimes you need more than support. You may need to talk to a social worker or therapist. Don't wait too long before you ask for help.

Managing the primary and secondary school years

While you need to care for your child's health, you also need to focus on more than their medical care needs. Their social and academic needs and growth are also important.

This section will give you information about how to help your child during the primary and secondary school years. You can help them do more things for themself, build self-esteem, and achieve school success. It will also help you look at the needs of your whole family and how you can take care of yourself during these years.

Primary school years

When your child starts kindergarten, they begin a new stage in life. They will spend a lot of time away from you, in the care of other adults. They will also spend more time with other children their own age. Their social life and their school life will lead to an exciting time of growth and independence, but can also raise new questions and challenges.

Transitions involve change, including adding new expectations, responsibilities, or resources, and letting go of others. There are transitions that are normal for development, from infancy, to childhood, then adolescence and adulthood. These developmental transitions occur for children with and without long-term illness.

Other transitions that all children face include changing from preschool to grade school, then middle school, high school, and college or work. Still other transitions include working with new agencies or providers.

You are the most important person in your child's life. You are the key to their success and self-confidence. The primary school years are a time to support your child to be more independent and do more things on their own.

Let your child do more

Your child can learn to take care of themself, even though they have sickle cell disease. They need to do things on their own. You need to support them to do things on their own. At the same time, you still have to see that your child gets what they need. But your job as parent is changing.

By age 6, most children want to start doing things themselves, such as:

- Help make their own breakfast or lunch
- Dress themselves and get ready for school
- Take a class or learn a skill, like playing the piano

Besides the things they want to do, they can also help out in the home. They can help with household chores like setting and clearing the table and doing dishes or cleaning up their room.

Let them do things themself unless the activities are not safe. Even if it takes longer, it is good for your child to be independent.

Help your child plan what they need to do. Young children often need more help than older children. For example, when you ask a 6-year-old to clean up their room, explain what you mean. Tell them to do the following tasks:

- Put toys in the chest
- Put clothes in the hamper
- Pull the covers up over the bed

By the time they are older, they should know what it means to "clean up their room." If your child doesn't ask to do things by themself, you may need

to push a little. Parents often find it hard to push a child who has an illness. They may feel guilty or afraid. Don't let your child's illness get in the way of helping them to grow up. It's okay to let them fail sometimes. That is part of how we all learn what we can do. No matter what, don't do everything for your child. When they do things for themself, they will feel good about it.

You are not the only one who may try to protect your child too much. Other family members may feel that they need special care. Let them know how you want them to treat your child. Ask them to focus on what your child can do, not what they can't do. The child doesn't need their "help." They need support for growing up.

Teach your child age-appropriate ways to take care of themselves with your close guidance. Encourage hobbies and social activities, including music lessons, Boys and Girls Club, sports, art, and other activities.

You can consider giving your child a small allowance. You can help your child learn about budgeting and saving. You can encourage them to think about their future by asking, "What will you do when you grow up?"

Sickle cell summer camp

Summer camp is always filled with outdoor fun, new friends, singing, swimming, story times, and games. At sickle cell summer camps, counselors know how to help children with the disease do as much as they can, without hurting themselves. The staff is aware of the special needs of children with sickle cell disease and can handle any problems that may arise.

Sickle cell summer camp provides a safe and fun setting for your child to gain confidence and self-esteem. Spending time away from parents with other children their own age will help your child be more independent. Ask your medical provider or local sickle cell disease group about where you can get information about camp.

Involving your child in their own care

In the primary school age years, encourage your child to begin interacting directly with the medical provider. Make sure that your child can say what special things they need to do, like drink more water, take certain daily medicines, not get overly tired, and visit the medical provider regularly. They also should know when they need to tell you, or a medical provider or teacher, that they are not feeling well.

By the time they are ages 12 to 14 years old, you should start talking to your child about what changes to expect in medical care when they are older. The big change will be moving to an adult medical care setting after turning 18 years old. Your child will also be more in charge of taking care of their own health and medical care decisions as they grow up.

Build self-esteem

Help your child feel good about themself. Pay attention to more than their problems. Notice their skills, strengths, interests, and style.

- Praise your child when they do well. You can't give too much praise. Children thrive when they are told that they are special. They feel good when someone sees something they've done well.
- Listen to what they have to say. Ask questions. Show them that you care about what they think and feel.
- Help them get involved in things besides illness.

They can try some of these things to see if they want to do them:

- Playing music
- Being with friends
- Learning to use the computer
- Drawing or working with clay
- Playing chess or other board games
- Acting in a play
- Reading books

Be careful not to push your child to compete where they can't succeed. If they can't run very long without getting sore or tired, don't force them to play fast sports. Help them find at least one thing to do well.

Self-acceptance

When your child starts school, they may begin to notice that they are different from other children. When they become aware of having "a disease," they may feel afraid or angry. They may think that they got sick because they did something wrong.

Make sure your child knows that they didn't get the disease because they were "bad." Nothing they did gave them the disease. Your child needs to accept the fact that they have sickle cell disease and make the most of their life. Tell them that except for their disease, they are just like other children. Learning to cope with the disease gives them special strength and resilience. Help them learn what they can and cannot do. This will give them a sense of control.

Fitting in

We all like to feel that we belong. Your child may be afraid that they won't fit in because sickle cell disease makes them different. They may think that others will make fun of them or treat them in "special" ways.

Some children feel good about telling their friends about sickle cell disease. Others don't want anyone to know. It is good for children to tell at least one close friend about their disease. If no one knows, it is more likely that they will feel ashamed and think of it as a secret. When friends know, they can provide support with better understanding and context.

If your child doesn't know what to say to friends, let them practice with you first. If your child explains to friends what the disease is – a blood disease – and that it is not contagious, it could help everyone feel better. Once they have told others, they will probably feel better about it.

School success in primary and secondary school

Most children want to do well in school. School is their work. Doing well in school builds a strong future.

Expect the best

Like other children, children with sickle cell disease can excel in school. They can be at the top of their class. Both you and the teachers need to expect the best from your child. When you expect more of your child, they will do better.

Some teachers may not expect enough from your child. Don't let a teacher underestimate them. Work with your child's teacher to get your child to have a positive outlook with high hopes and dreams. Help your child want to succeed in school and reach their full potential.

Be a partner with teachers

At the start of a new school year, go to school and meet your child's teacher and school nurse. Bring your child with you. Let your child ask questions. Tell the teacher and nurse about sickle cell disease. Give them things to read to learn about the disease.

The teacher needs to know that your child will come to school with minor aches and pains. Your child should be sent home only if they have a fever or severe pain or if they need to see a medical provider.

Explain your child's special needs. They need to do these things:

- Get water when thirsty
- Go to the bathroom as soon as they feel the need

- Make up schoolwork if they have to miss school
- Rest or slow down if they are tired or sore. For example, during gym class, they may not be able to do as much as other children
- Rejoin the class as soon as they are ready
- Get medicine if they need it
- Dress for the weather and stay warm

Check to see that the teacher gives your child what they need. Some teachers may protect your child too much while others may ignore them. Talk to the teacher about these things if you are concerned. If you need help or support, talk to your school principal, medical provider, school nurse, or social worker.

Stand up for your child's rights

Children with sickle cell disease can sometimes have learning differences and need some extra support. Learning difficulties can include taking longer to process questions and complete assignments. An intelligent child with great potential may have a specific learning difficulty that may impair their school performance. Identifying the problem allows it to be addressed so that your child can reach their full potential.

Your child has the right to get an education that meets their needs – this is the law. The Education for All Handicapped Children Act of 1975 (Public Law 94-142) means that the school must provide help if your child needs it.

If your child is not doing well in school, talk with the teachers. You may want to ask that your child be tested for learning differences. If they have a learning difficulty, they should get special help so they can learn better. Ask the school counselor for an "individualized educational plan" ("IEP").

This IEP plan could include any of these:

- Regular school classes
- Home instruction
- Time with a resource specialist
- Special classes

A "504 plan" is a different way to get support for your child. A 504 plan is a legal, binding document. It is designed to help any student attending public school who has special physical or psychological needs. The goal is to help them feel comfortable in the regular classroom. A 504 plan calls for unrestricted bathroom and water fountain breaks or other necessary accommodations, so that a child does not have to explain their needs to their teachers on a daily basis.

If a teacher suggests that your child be held back, get an opinion from another learning expert. Often being held back is not helpful to children with learning difficulties. It can actually make things worse and make your child feel bad about themself. Instead, make sure that your child is tested for learning problems and gets any extra help they need.

Tell your medical provider or a social worker if you don't think your child is getting enough help. Your medical team can work with the teacher and school so your child gets the help they need.

See the online Appendix for more information on an "IEP" and "504 plan."

Plan ahead for illnesses that take your child out of the normal school day

Set up a plan with your child's teacher for your child to do homework or make up missed work. Talking about the plan will give you a sense of what the teacher thinks about your child's disease. It is a good time to give the teacher more facts about sickle cell disease. Most teachers are happy to do what they can to help.

Find out what resources your school has for children with long-term illnesses. Ask your child's teacher, school counselor, social worker and medical provider if there is a hospital tutor program or other community programs that could be helpful if needed.

If you don't think that there is enough help, speak up before your child gets too far behind. Ask the social worker who works with your sickle cell disease program, a family member, or friend to help you speak up about your child's needs.

School sports

One of the ways that many children build self-esteem is through sports. If your child wants to play school sports, help them to be realistic about what they can do. Talk to your medical provider about sports.

Look at your child's strengths and find a sport that matches these strengths.

If the sports program has a leader or coach, tell them about your child's special needs. Remind your child that they need to take good care of themself. They need to do the following:

- Drink when thirsty
- Rest when feeling tired
- Dress for the weather and stay warm

Keep your child in school

Send your child to school unless they are sick enough to see a medical provider. Don't keep your child home from school if they just have something like a runny nose. They also don't need to stay home because of bad weather. Just make sure they wear the right clothes.

Children benefit from going to school and being with other students. Keeping your child home from school when they don't need special care will cause problems. They may be left out of friendships and have trouble learning social skills. They can also fall behind in schoolwork and may find it harder to do well in school.

If your child does have to spend time in the hospital, have them try to do homework while they are there. You can also encourage them to talk or write about what has happened to them in the hospital. If they miss a lot of school, they may need outside help or a tutor so they can keep up.

Most of the time, it's best to send your child back to school as soon as they come home from the hospital. The more they are in school, the better off they'll be.

Unless your doctor says it is needed, don't agree to home instruction for your child. Home instruction can't replace the school setting. In the classroom, children learn from each other as well as from books and teachers. This recommendation may not apply if you home-school all your children.

Managing the teen years

The teen years are a time of major changes for you and your child. There is a lot that you can do to help your child during these years. There is also a lot that is out of your control. Your support and help still matters, but your teen will be making their own life choices.

The teen years are exciting because your child is becoming a young adult and gaining independence. These years build an important foundation for adulthood. As your child learns to take care of themself more and more during these years, they will need to learn about the sickle cell disease complications that can happen during the teenage years too.

Your teen is getting ready for the time when they will be on their own. They are growing up and learning to live their own life. Your teen needs limits, but they also need freedom. You and your teen will keep trying to find a balance that works.

As a parent, there are many things you can do to help your teen during these years. Each of these guidelines will be described in this section:

- Let your teen do more
- Set limits for your teen
- Build self-esteem
- Consider the present and future
- Plan the transition to adult medical care

Let your teen do more

Your teen will soon be an adult living on their own. Now is the time for you to help them take control of their life.

It can be hard for a teen with sickle cell disease to feel in control. Many teens feel chained to the disease and the special care it requires. They feel angry that they have to listen to lots of adults: you, teachers, and medical providers. While this is true, there are ways that they can have more control over their lives even though they have this disease.

Start to let go

Your teen may not take care of themself as well as you took care of them. Try to let them do it anyway. If you are afraid that they are hurting themself, talk to them about it. Explain what might happen if your child doesn't take care of their health. Try to find out why they don't want to take the steps you recommend. Help them find a better way if needed.

Now is the time to shift control from you to your teen. You can start slowly, but keep moving towards giving them more control. You need to trust them to ask for help when they need it. If they make a mistake, help them learn from it. If they learn now, they can take charge of their own life as an adult.

Let them do things their own way

There are many ways for your teen to be independent besides managing their sickle cell disease. The way they talk, the clothes they wear, the music they like, and the way they do their hair are all ways to express themself.

You may not like the way they look or act, but try to accept it. Unless they are hurting themself or others, try to leave them alone. They want to be different from you. They want to be their own person.

Remember that growing up may be scary

Many teens with and without sickle cell disease are afraid to grow up. They don't want to take care of themselves and face their lives. They may try to act younger than their age. Sickle cell disease can make things more complicated because of the effect that the disease has on growth. A 13- or 14-year-old with sickle cell disease may look 10 years old.

Since they may look younger, you and others may be tempted to treat them that way. Don't let this happen. Expect your teen to act their age, not their size. Don't do everything for them. If you don't do it all, your teen will find out that they need to take care of themself.

It can also be helpful for your teen to get more involved in things other than the disease. Help them find a part-time job, a hobby, or new friends. This will give them a chance to be with others and prepare for their future.

Give them more independence

People with sickle cell disease should look forward to growing up to become independent adults. As a parent, it is your role to prepare your teen for adulthood and living on their own.

As your teen gets older, you should encourage their independence, no matter what your child's capabilities are. Even if your child has severe learning disabilities, or is in full-time special education, they should be encouraged to do some things for themself.

Help your teen identify and build on their strengths. Discuss issues related to reproductive health, risky behaviors that teens can get into, and substance use. Talking about it won't push your teen to do these things. They need your guidance in making the best judgments.

Groups who may offer help on how to parent a teen:

- Your local YMCA or YWCA
- Community groups
- Schools
- Support groups for parents of children with sickle cell disease

Set limits for your teen

While independence is important for your teenager, it is also important to be the parent. It is your job to decide how much freedom to allow. You have a right to know what your teen is doing and who your teen is with.

Limits are important and provide rules, boundaries, and guidelines to protect your teen and support their growth. Limits can cover many issues like curfew,

homework, chores, and use of the car. Communicate with your teen and tell them how you expect them to act. Make sure that the limits are clear to both of you.

If the rules are broken, you should discuss the action and consequences with your teen. Sickle cell disease is not an excuse for breaking rules. Your teen needs to be held responsible for their choices.

Teens with sickle cell disease may have a stronger need to prove that they fit in. They may also be depressed and want to escape from their pain. This can lead them to take risks that can be hurtful to themselves and others.

Some of these risks may carry extra danger for your teen with sickle cell disease:

- Sex without condoms can bring a greater risk of getting a sexually transmitted infection (STI)
- Unplanned pregnancies can create some health issues and concerns
- Alcohol can increase sickling because it dehydrates the body
- Cigarettes can also increase sickling because smoke lowers the oxygen level in the blood

All of these are risks for any teen, but they can cause more harm to teens with sickle cell disease.

Know when to step in

If your teen is taking risks that could be harmful, you need to get involved. Your teen needs proper and fair discipline if they are hurting themselves or hurting others.

They also need your guidance and attention. Listen to your teen. Find out what is behind their actions. Ask why they are taking risks and listen to their answers. Tell them how they could hurt themself and ask them what they think.

If your teen doesn't stop or can't stop taking these risks, they need help. Taking risks that put them and others in danger can be a sign of a deeper problem. You may need outside support and help.

Most people are not taught how to be parents, let alone parents of teens. It is okay for you to be confused about how to handle your teen. It is okay to get help.

Build self-esteem

Like all teens, teens with sickle cell disease need support to build good self-esteem.

These are things you can do as a parent to help your teen feel positive about themself:

- Listen to what your teen says
- Encourage your teen
- Do things with your teen
- Help your teen feel positive about their body
- Let them know how good you feel about them
- Don't put your teen down

Listen to what your teen says. Your teen needs to feel that you listen to them when they speak to you. Even if you don't like what you hear, you can still try to understand what they are saying. A good way to let them know that you have heard them is to repeat what they have said. Once they know they've been heard, you can then tell them how you think and feel.

Ask questions if you aren't sure what your teen means or how they feel. Questions that take more than a "yes" or "no" response are the most helpful.

Don't put your teen down. It can be easy to put down teens and say hurtful things to them. Put-downs are harmful, not helpful. They won't make your teen want to change or feel good enough about themself to change. Instead, they just make it harder for your teen to relate to you.

If you don't like something that your teen did, sit down with them and talk about how it affects you. Your teen needs your respect and praise.

Doing things with your teen is important. Being with you may not be your teen's favorite thing to do. But they still want to know that you want to be with them, at least sometimes. Think about something that you would both enjoy. It could be a movie, a ball game, a concert, or anything that you both like to do. Spending time with your teen can tell them that you think they are worth being with.

Help your teen feel positive about their body. Teens with sickle cell disease often feel ashamed of their bodies. Some look younger than their friends because they mature late. Some feel embarrassed by jaundiced eyes. Some have scars from surgeries and IVs.

Besides the things that can be seen, some teens may also have limits on what they can do, like sports or dancing. Some teens want to prove to themselves they are like other teenagers and engage in behavior that may not be appropriate.

There are some things you can do to help your teen feel positive about their body. If your teen is small for their age, help them find clothes they like that fit right. If they have scars, help them find clothes that cover them up. You can also help them find easy ways to talk about their differences to others. If they can talk about the differences, they may not feel as ashamed.

Remind your teen that their body will mature, and they will get bigger. It will just take them a few more years than their friends. Also let them know that it is okay to be their current size. Focus on their strengths and help them feel good about themself.

Encourage and praise your teen. We all need praise. Providing your teen with regular praise encourages success.

Keep an eye out for the times your teen does kind things for themself and for others. Don't be afraid to tell them how good it makes you feel when you see them doing things well. The more good things you notice, the more good things there will be.

Don't assume your teen knows that you love and care about them. Teens need to be told that they are loved and that they count.

Treat your teen like they have something to offer, and they will. Help them get involved in things that they do well. Ask them about school, work, or any special projects. Ask them to show you what they are working on. Your attention will show that you believe that what they do has value. Help with school and future plans.

Consider the present and the future

Staying in school

Many teens with sickle cell disease have missed a lot of school because of their illness. Some teens manage to still do well in school while others have major problems. They may be behind their classmates, have poor study habits or overlooked learning problems.

If your child is having problems with school, these problems may get worse during the teen years. They may feel depressed about their future. These hopeless feelings can make it hard to succeed. If your teen feels like they can't keep up, they may just want to give up and drop out.

Urge your teen to stay in school. Help them keep up their drive to do their best. Encourage them to focus on what they want to do in the future and to work towards it.

Your teen can arrange to make up schoolwork they miss if they have to be out of school. Also, find out if the school has help for students with special needs. Help your teen get extra help if they need it.

Planning for the future

High school is the time for your teen to focus on their goals for the future. Help them focus on what they can do, not what they can't do. No matter what their limits, there is a place for them. As they start to plan their future, help them look at their strengths and skills and see what type of career might be good for them.

It may help them to have a model of success. If you know an adult with sickle cell disease whose life they might admire, tell them about what that person is doing. They might even want to meet that other person.

See the online Appendix for a list of sickle cell disease "greats" (people who have led inspiring lives).

A part-time job can be a good way for your teen to explore interests. A job can also help your teen gain confidence in their skills and earn some money. During the school year, make sure that the job doesn't get in the way of school. During summers, a job can be a great way to help teens mature.

Your teen will need to decide whether they want to go on to college. Many people with sickle cell disease go to college. Some colleges have programs to help students with special needs.

Encourage your teen to plan for a full life. When they take some control of their life, their image of themself will improve, and they will have a better chance for success.

Discuss job worries

Many teens with sickle cell disease worry about whether they will be able to find a job. They are afraid that they won't be able to support themselves. They may feel scared that their limits will prevent them from being able to work or keep a job.

Listen to their fears and give them support. Tell them that some people with sickle cell disease have become doctors, lawyers, social workers, teachers, and business owners. Help them focus their mind on what they can do to get ready for their future.

- When you allow them to take care of themself, you are preparing them for the future
- When you help them feel positive about themself, you are raising their chances of success
- When you help them do well in school as well as get involved in other interests, you are giving them the best training for future careers

Financial resources

Talk about financial resources, both medical care related (insurance, for example) and related to daily living. Your teen should have a good plan about how and when they might be able to support themself or contribute to your household. Encourage work and volunteer activities. Talk about and help your teen explore possible career interests.

Help for teens who are struggling

If your teen is having problems with school or doesn't have plans for their future, they need help. A counselor from the school or social worker from the clinic can help them take a look at their feelings and their life. The counselor or social worker can work with them and help them find better ways to live with their illness.

There are times when your teen may need more help than you or their medical team can provide. These are some signs that your teen needs help:

- Your teen is angry a lot or taking dangerous risks
- Your teen seems very depressed or anxious
- Your teen won't take care of their health
- Your teen is doing poorly in school
- Your teen has more frequent visits to the emergency department, the hospital, or both

Reach out for help when there are any changes your child seems to have trouble adjusting to, such as when you and your teen feel like you can't talk to each other, or your other children are very upset about the way your teen is acting, or someone close to your teen gets divorced or dies.

Get help for your teen and your family

Individuals with sickle cell disease and their families have greater access to counseling and support for mental health, even delivered virtually by telehealth. Do an online search for the Sickle Cell Disease Association of America (SCDAA) website to learn more about working with a counselor who knows about the unique challenges that may come with living with sickle cell disease.

Most sickle cell disease care centers have social workers and counselors on their staff who know about sickle cell disease or chronic illness in teens. Ask your medical provider who they think would be most helpful to your teen or your family.

Many cities have support groups for parents of children with sickle cell disease. Some large cities also have support groups for teens with sickle cell disease. Ask your doctor if there is a program like these that you or your teen can join. Parents and teens can gain a lot from talking with other people in similar situations. You may also be able to get help through your place of worship. Be

sure to call 988 if you or someone in your family is in an immediate mental health crisis.

Additional places for teens to get help with school or work:

- School counselor
- Vocational counseling
- Educational consultants
- Tutoring programs
- Nonprofit organizations like the YMCA or YWCA

Get help for yourself

When teens have problems, parents have problems. Getting help for yourself is often the best way to help your teen. With support and guidance, you will be better able to give your teen what they need. Ask for help from the people and groups listed above if you need it.

Get help before problems get worse. Getting help early is a sign of strength, not weakness. It is an important way for you to help your teen and your family make the most of your lives.

Plan for the transition to adult medical care

Your child should be involved in their own medical care, with their involvement growing as they grow.

When your child reaches 14 years of age, the pediatric sickle cell disease team should start helping to measure your teen's readiness to take care of their own medical care. The transition to adulthood and adult sickle cell disease care can be difficult. The more independence your child shows when taking care of their own medical needs while they are still in pediatric care, the easier it can be.

During the late teenage years, it is important that the medical team helps assess your child's ability to take care of their own medical needs. This should happen **before** your family expects them to do things completely on their own.

There are resources that provide information on preparing for and moving to adult medical settings. There are online tools available for you and your teen to measure their readiness to take on more responsibility themself. Online tools to help with the transition are available at the Got Transition website (www.gottransition.org).

Every person is different. Teens will begin taking care of their own medical care at different times. Some people with sickle cell disease will continue to need support with medical needs into adulthood.

Helping your teen settle into an adult medical setting is important. This transition generally happens between 18 and 26 years of age.

See Chapter 2, “Special considerations for teen years” section, for more information on involving your teen in their medical care and transitioning to adult medical care.

See Chapter 8, “Teens and self-advocacy” section, for information on how your teen can deal with bias in the hospital or ER.

See the online Appendix for more resources on “transition.”

Chapter 5:
Basics on Fever, Pain, and When to Get Help

Introduction

With sickle cell disease, there will be times when your child doesn't feel well. You are a key part of your child's medical team. You need to know when to get your child help and when you can help them feel better at home.

This chapter will teach you how to recognize problems early and when to get help. It will also share ways to help your child deal with the pain that sometimes is a part of sickle cell disease.

The topics in this chapter are fever, easing pain at home, and when to contact a medical provider. Information on treating severe pain is in the next chapter.

See Chapter 6, Pain and Pain Medication.

Fevers

It is important to know when your child has a fever and what to do about it. When your child has a fever, it is a sign that their body is fighting an infection. Infections can be very serious and life-threatening in children with sickle cell disease. Noticing symptoms of an infection early allows you take actions to prevent the infection from getting worse.

If you think your child might have a fever, take their temperature. Using a digital, forehead, or ear scanning thermometer is easiest for your child, but there are many types of thermometers available. Thermometers usually show temperature in Fahrenheit (°F), but they might also have markings in Celsius (°C).

See the online Appendix for web pages that convert Celsius (°C) to Fahrenheit (°F) and Fahrenheit (°F) to Celsius (°C).

- The normal temperature by mouth is 98.6°F (37°C)
- If your child's temperature is 101°F (38.3°C) or higher, contact your medical provider right away

A child can have a fever of less than 101°F with a cold. However, a fever of 101°F or more may mean a serious illness that the doctor needs to know about.

Tell your doctor or nurse what kind of thermometer you used to measure your child's temperature, and where you used it (forehead, mouth, ear, armpit or rectum). It is important to know where you took the temperature because the body's temperature is different in different parts of the body. Before you give your child any medicine to bring down a fever (including cold medicines), contact and speak with your medical provider.

If the temperature is less than 101°F, the medical team will probably tell you to give your child acetaminophen (Tylenol) or ibuprofen (Advil or Motrin). They will also tell you how much of this medicine to give, depending on your child's weight. Aspirin should not be given to children because it is associated with a serious disease called Reye's Syndrome.

Take note of these tips on taking your child's temperature.

- If your child is sick, take their temperature early in the morning and late in the afternoon. If your child seems very sick, check their temperature more often.
- Don't give your child anything hot or cold to drink or eat for a half hour before taking an oral (in the mouth) temperature. Food or drink can change the reading by warming or cooling the child's mouth.

You don't need to take your child's temperature every day if your child is well. It is not needed and can be upsetting to your child.

Thermometers

There are many types of thermometers now available. There are thermometers that can be used on the forehead or in the mouth, ear, armpit, or rectum. You should have your own thermometer at home. You can buy one at a drugstore or ask your medical provider if they can give you a thermometer to use. Read the instructions for the thermometer so that you use it correctly and on the correct body part.

When to take a rectal temperature

If you can't get a temperature by the forehead, ear, armpit, or mouth, you can take a rectal temperature under these conditions:

- with a baby or a young child who can't hold a thermometer in their mouth for 2 to 3 minutes; or
- with a very young child who is congested and can't breathe through their nose.

Use K-Y jelly, Aquaphor, Vaseline, or some other lubricant on the thermometer to make it go in smoothly.

When to take an oral temperature

When your child is old enough to hold a thermometer in their mouth and keep their mouth tightly closed, you can take an oral temperature. Place the thermometer under the tongue.

Reading a digital thermometer

A digital thermometer shows you the exact temperature. Read it and then wash the tip with warm water. Wipe it with alcohol and put it back into its case. Make sure that the thermometer is working normally and does not need a new battery.

Remember: if your child's temperature is 101°F or higher, contact your medical provider right away. Make sure to say where you took the temperature (forehead, mouth, ear, armpit or rectum).

Easing pain at home

Pain is common in children and adults with sickle cell disease. Most pain can be handled at home. If the pain doesn't go away or gets too strong, your child may need to go to the clinic or hospital.

As soon as the pain starts, your child should drink lots of water and fluids and take acetaminophen (Tylenol) or ibuprofen (Advil or Motrin). Oral opioids are sometimes needed to manage pain at home. Your child's medical provider should oversee this.

Your child can also try other home remedies for pain, such as using heat packs or listening to relaxing music. Distracting your child with activities will lessen pain.

If the pain gets worse or doesn't get better, contact your medical provider. Your medical provider may ask you to find out if your child has a fever. The medical provider may also tell you to give your child more to drink or to try some of the other home remedies listed below.

If the pain is too severe, your child may need to go to a medical office or the emergency room.

Contact the office before you take your child to either of these places so your child will be seen as soon as you get there.

Infants and toddlers may show pain by crying, refusing to walk or use the body part that hurts, or pointing to the areas that hurt. Pay attention to these signs. Try different ways to ease pain to see which ones help the most.

Home remedies

More fluids. Extra fluids can help keep the sickled cells from clogging up small blood vessels. This is a major cause of pain. Encourage your child to drink more fluid than they usually drink.

See Chapter 3, Care at Home, for the table on the amount of clear fluids your child needs each day during pain and other special times.

Quiet activities. Cutting back on physical activity can be helpful. Complete bedrest may not be needed, just less active play and activities. Find things for your child to do quietly inside for a while. See if quiet activities will help them feel better. However, leaving a child alone in pain without ongoing monitoring and helpful quiet activities can make their pain worse.

Warm baths. Let your child soak in a warm bath for a while. When the bath cools off, you can add more hot water to keep your child comfortable. Sometimes it feels good to do mild exercises in the warm water.

Heating pad or warm, moist towels, or heat packs. Apply one of these to the painful area. If you use a heating pad, set the dial to medium heat. If you use warm towels, change them when they cool off. Wet them with warm water, and then wring them out. You can apply these as often as it helps.

Massage. Gently massage the painful area with warm baby oil or lotion to relax tense muscles and increase blood flow. You can massage the arms, legs, back and neck areas easily.

Acetaminophen (Tylenol) or ibuprofen (Advil, Motrin). These medicines can provide relief for pain caused by sickle cell disease. Make sure to give your child the right dose for their weight. As mentioned above, never give your child aspirin, unless ordered by your medical provider, because it is associated with a serious disease.

Pain medication. Contact the medical provider if acetaminophen and ibuprofen are not helping the pain. Do not give your child pain medication that has not been prescribed for your child. Your child should never be given pain medication that was prescribed to someone else.

Your child will feel less pain if they are involved in something they enjoy. Young children like to be busy. Talk to them, offer games, stories, and other fun things. Children of all ages can talk to people, play games, watch shows or movies, or listen to recordings or music. Let your child pick what they want to do to keep their mind off the pain.

Some children like to be alone when they feel pain, but it is best not to leave your child alone when they are hurting. Learn what works best for your child but try to have someone with them to provide distractions and oversight.

Teaching your child to relax

Your child may hurt less if they are able to relax. You can help your child learn to relax when in pain. With practice, your child may be able to stay calm and relaxed.

Relaxation is a skill that can be learned. Like any other skill, it takes practice to learn to relax. Pick times that are quiet and free of distractions to help your child practice. Bedtime is often a good, quiet time. It is also a time when you may have a few extra minutes to help lead your child through these exercises.

Deep breathing

Deep breathing is one of the simplest and best ways to relax. Try to find fun ways to help your child breathe deeply and evenly. Have your child take a few deep breaths and let them out to the slow count of 1…2…3… Or have your child pretend to be a bicycle tire that is slowly losing air.

Try one of these ways to keep the deep breathing going for a while. Tell your child to pretend that they can breathe under water if they breathe in this special way. Or they can pretend to be an astronaut in a space suit who has to breathe deeply and evenly.

Exercises for younger children

Exercise #1: The Rag Doll. “Pretend that you are a robot (or wooden doll), all stiff and straight. Your arms and legs don’t bend at all. They just stay straight.”

“Now you are a rag doll, all floppy, with no bones.” (Lift your child’s arm up, shake it a little to make sure it’s really loose.) “All loose and floppy.”

“When you need to relax, pretend to be the wooden doll first, then change to the rag doll.”

Exercise #2: Spaghetti. “Pretend that you are spaghetti in a package that has not yet been opened. You are all stiff and straight.” (Have your child hold this for a few moments.)

“Now you’re cooked spaghetti, all over the plate. Are you covered with sauce or meatballs?”

“When you need to relax, pretend to be spaghetti in the package, then the cooked spaghetti.”

Exercise #3: My Special Place. Begin with deep breathing practice. "Each time you breathe out, you get more and more comfortable. Now you're loose and comfortable. No tightness anywhere, breathing deeply and evenly. All your muscles are smooth and warm. Just an easy feeling. As I slowly count backwards, you are going to feel even more relaxed: 5 … 4 … 3 … 2 … 1 … "

"Now imagine that you're in your favorite place (maybe floating underwater or in a made-up land), feeling free and easy. What do you see? Hear? Taste? Feel? Smell?" (It's best if their eyes are closed, but they don't have to be.)

Exercises for older children

Begin with deep breathing practice. "Start at the top of your head. As you breathe out, your scalp feels loose and comfortable. Now go down through your face and feel all the little muscles smooth out. Now feel any tightness flow out of your shoulders, down through your arms and hands, out of your body."

"Now feel any tightness flow out of your chest as you breathe out. Your stomach muscles feel loose. You feel any tightness, any tension, flowing out from your hips down through your legs, out the bottoms of your feet. You feel loose and comfortable, more and more relaxed each time you breathe out."

"Now I'm going to count backwards from 5 to 1, and you will feel even more relaxed. 5…breathing deeply and evenly 4…more and more relaxed 3…further and further 2…loose and comfortable 1." (If your child wants, they can imagine their favorite place at the end of the count of 1.)

Teaching tips

- Keep your voice low and soothing. Speak slowly, but not in a monotone.
- Keep it natural. These are just outlines of what to say. Say what feels good to you. Soon, you will find your own words for the same things.
- Work with a psychologist. It can be helpful to learn these exercises from a psychologist. They will pick the ones that will work best for your child and teach them to both of you.
- Find or make recordings of the exercises. Relaxation or meditation recordings for children are available. A psychologist can also make a recording of these exercises for your child to use as a guide. If you and your child make your own recording, add music or a story if it helps.
- Keep it positive. Your child does not have to try to relax all of the time. Make it enjoyable so that they will be willing to try.

Self-hypnosis

A psychologist can teach your child self-hypnosis. This is a more detailed relaxation technique that your child can use on their own.

What to expect

It is important for your child to practice these techniques when they do not have pain, so that it comes easily to them during times of pain.

Relaxation techniques can help your child manage pain. If your child feels pain coming on, they should drink extra fluids, take acetaminophen or ibuprofen, and relax in whatever way works best. Relaxation exercises and other home remedies alone will often be enough. Other times, they may need medicine.

If your child is admitted (checked in) to the hospital because of pain, the physical therapist and other staff may be able to teach you other ways to ease pain. Ask them for suggestions so that you can learn more ways to help your child feel better.

Additional reasons to contact the doctor

In addition to fever, there are other situations when you should get in touch with your medical provider quickly. Contact your medical provider right away are if your child has any of the following signs:

- Chest pain or shortness of breath
- Abdominal pain
- Pain along with fever or swelling and redness
- Pain that isn't relieved by home remedies
- Severe headache
- Inability to walk or speak normally

Go over these with your child so that they know what signs are important to share with adults, such as teachers, parents and medical providers.

The sooner you find out about health problems, the better. Sometimes early treatment can save your child's life. Other times, it keeps a problem from causing damage. The longer your child waits to be treated, the worse a problem can get.

A more detailed list of when you should seek advice and the danger signs that need emergency attention are at the end of Chapter 1. Take a photo of that page and keep it in your phone. If you can, print it and put it up on your refrigerator or wall.

See Chapter 1, Basic Questions, Danger signs: When to contact a medical provider.

Chapter 6:
Pain and Pain Medication

Introduction

This chapter is about pain and pain medications. Information on how to ease pain at home is in the previous chapter.

Pain from sickle cell disease is the main reason that children with sickle cell disease need to go to the hospital. When people with sickle cell disease have pain, it is called a vaso-occlusive episode or a VOE. Sickle cell disease pain is less common in very young children and more common as children get older. However, sickle cell disease pain can begin as young as a few months of age. Sickle cell disease pain in all age groups can be mild to severe and last hours or days.

The first presentation of pain in babies and toddlers may be hand-foot syndrome (dactylitis). This is disease pain and sometimes swelling of the fingers or toes. Dactylitis results from sickling in the bones of the hands and feet. In older children, common places where sickle cell disease pain starts are the back, chest, and leg bone.

Over time, children and parents may recognize a common pattern of sickle cell disease pain (for example, one person may know that they develop sickle cell disease pain in the lower back and legs after being active). In assessing your child's pain, it is helpful to be aware of the number of sites that hurt and how much those sites hurt. There are simple tools that will help you and your child to report the pain, such as a face scale. These tools are explained more in this chapter.

See Chapter 5, Basics on Fever, Pain, and When to Get Help.

Pain that is too severe to be treated at home

If your child has pain that is too severe to be treated at home, they may need to go to the clinic, emergency room (ER), or hospital.

At the clinic or hospital, your child may get fluids through an intravenous catheter (also called an IV). The IV is placed by putting a needle into a vein, similar to when blood tests are done. A small plastic tube is left in the vein. It is connected to a larger tube that can be used to put fluid for hydration and pain medication and other medicine directly into the blood.

Your child may get stronger pain medicine through an IV, or if the IV is too hard to get or will take too long, they may get the medicine through a nose spray or a shot instead of an IV. The stronger pain medicine helps most children feel better quickly so they can go home.

If your child needs stronger pain medication than acetaminophen (Tylenol) or ibuprofen (Advil or Motrin) to take at home, you will get a prescription from the doctor. The prescription will be for a small amount of oxycodone or other prescription-strength pain medication (usually an opiate). Do not collect many of these pills at home. These drugs (like all medicines) should not be shared with other family members.

At home, the pain may last a few more days, but it should keep getting better. Use fluids, acetaminophen, relaxation, and other home remedies to help ease the pain.

If the pain does not get better in the clinic or ER, your child may need to be hospitalized. In this case, your child will be given more fluids by IV and more pain medicine. Hospital staff may offer physical therapy to help with pain. This can include whirlpools, massage, exercise, and heat packs. After 3 to 5 days in the hospital, your child should feel well enough to go home.

Patient Controlled Analgesia (PCA) Pump

At many hospitals, your child will get a Patient Controlled Analgesia (or PCA) pump so that they can control their own pain medicine. With a PCA pump, your child can press a button to pump the medicine into their veins. The pump is set up so that the right amount, but not too much, medicine is released. This gives people with sickle cell disease better control of the pain in a safe way.

Pain medications

If your child goes into the clinic, day hospital, or ER for the treatment of severe pain, they will most likely get strong pain medication.

Mild pain

Mild pain at home is managed with non-opioid medications. The most commonly used medicines are acetaminophen (Tylenol) or ibuprofen (Motrin or Advil).

Moderate to severe pain

Moderate to severe pain at home may need a class of medications called “opioids” or “opiates” for better pain control. You have probably heard of these in the news because of people using them in unsafe ways as street drugs. When people with sickle cell disease have severe pain, opioids are essential to their pain control and should be used. This is what they are meant for. They should not be withheld if your medical provider

recommends them. At home or in the ER, taking opioids according to your medical provider's direction is more likely to control pain and avoid hospitalization.

The most commonly used opioids are hydrocodone and oxycodone. These opioids may be in a combination pill that includes acetaminophen or ibuprofen. If a pill includes an opioid and acetaminophen or ibuprofen, do not give your child acetaminophen or ibuprofen in addition to that pill.

Doctors may also prescribe the opioids morphine and dilaudid. Giving codeine to your child routinely is generally not recommended because it does not always improve pain.

Fentanyl is an opioid that works rapidly. The pain relief doesn't last long though. It is commonly given through the nose to the patient while the emergency room is being set up for pain management. This is used to get initial pain control while the medical providers are working on placing an IV for IV pain medicine.

Ketamine is not an opioid, but it is another important medication for sickle cell disease pain. It is not available at all hospitals. Some ERs and hospitals are now able to give ketamine to people with sickle cell disease through an IV. Ketamine can really help with sickle cell disease pain. This is because it treats pain and also makes opioid medications work better.

There are other medications that can be placed directly on the skin as a patch (lidocaine patch) or a cream or ointment (lidocaine or diclofenac). These can be very helpful to place directly on the arms, legs, back, or other areas of pain.

There are also other medicines that can be taken by mouth that were previously used for other conditions, such as muscle spasms or depression. These can work very well to lessen sickle cell disease pain.

It's hard to keep track of all the medications. The table below is a general guide. Medications and doses should be individualized and discussed with family during a scheduled visit. Every child with sickle cell disease should have an individualized pain plan in their medical chart that outlines how their sickle cell disease pain should be treated in the ER and hospital. These individualized pain plans lead to faster and better care.

Table: Pain medicines

Medication	Type	How is it given?	Can this be given at home?	How frequently is it usually given?	What side effects?
Acetaminophen (Tylenol)	**Non-steroidal anti-inflammatory drug (NSAID)**	By mouth or by IV	Yes	Every 4 to 6 hours	No common side effects but overdosing can cause liver damage
Ibuprofen (Advil, Motrin)	**NSAID**	By mouth	Yes	Every 6 to 8 hours	Can cause stomach irritation, and long-term use may affect kidney function
Ketorolac (Toradol)	**NSAID**	By mouth or by IV	Yes	The medical provider should discuss this	
Oxycodone (Percocet)	**Opioid**	By mouth	Yes	Every 4 to 6 hours	All opioids can cause constipation, nausea, itching, sleepiness. People may experience withdrawal symptoms after 7 days, which is corrected by slowly decreasing doses. Uncommon side effects related to overdosing can result in trouble breathing and over-sedation
Hydrocodone (Norco, Vicodin)	**Opioid**	By mouth	Yes	Every 4 to 6 hours	
Morphine	**Opioid**	By mouth or by IV	Yes	Every 3 to 4 hours	
Hydromorphone (Dilaudid)	**Opioid**	By mouth or by IV	Yes	Every 3 to 4 hours	
Fentanyl	**Opioid**	Through the nose or by IV	No	Given only in the ER for 1 to 2 doses when trying to get an IV placed for IV pain medications	
Ketamine	**Anesthetic**	By IV	No	The medical provider should discuss this	Can cause disorientation, confusion, or altered mental status

Getting the right dose

In order to get control of severe pain it is important to get the right dose of pain medication. If your child's pain is not better, they may not be getting enough pain medication. On the other hand, too large of a dose can cause problems, like sleepiness or pneumonia. If you are concerned about how much pain medicine your child is getting, talk to the doctor.

Side effects

Pain medications can have some side effects. Often, opioids cause constipation. Constipation is having difficulty "pooping." If this happens, give your child something to help soften their poops, such as prune juice. Feed them a diet high in fiber, with lots of fruit, whole grains, and beans. Give them a lot of fluids. If the constipation continues, contact your medical provider for advice.

Other side effects of opioids include itching and mood changes. Itching can be treated with diphenhydramine (Benadryl). There are some serious problems that can happen with pain medications but these are very rare.

Short-term pain medication use does not cause addiction

Short-term use of pain medications for sickle cell disease pain is not addicting. Many parents worry their children will get addicted. However failing to treat your child's pain can lead to serious complications.

Addiction stems from other problems. Children with sickle cell disease are not more likely to abuse drugs than other children.

Most cases of addiction occur in teenagers and young adults who have other problems besides sickle cell disease pain. A teen or adult who is having problems with home, school, or friends may abuse pain medication. When this happens, it is likely that other drugs would be used if the pain medication wasn't around.

Addiction prevention begins at home. Speak to your social worker or nurse if you are concerned about drug abuse. Take action early if you think your child is having problems at home or in school. Get involved with drug education programs at school and in your community.

Pain medication dependency and withdrawal

Long-term use of some medications can cause the body to become used to it. If the body has become used to the drug, suddenly stopping the medication will create discomfort. This is called withdrawal. Withdrawal signs include fatigue, upset stomach, anxiety, and trouble sleeping.

When someone has taken pain medication for a long time and the pain has stopped, the doctor

will gradually decrease the dose of medication to prevent a problem. Withdrawal does not happen in people using pain medications for a short period of time. Dependency is not the same as addiction.

Pain medication tolerance

Some medications become less effective if they have to be used for a long time. When that happens, higher doses are given to decrease the pain.

Chronic pain syndrome

The strong pain medications described above are only for sudden, short-term pain. They are not meant to be used for long-term (or chronic) pain. People with chronic pain need to learn other ways to control their pain besides taking very strong pain pills. Some special pain problems require different medications that your medical provider will talk with you about.

Fewer than 1 in 20 people with sickle cell disease have chronic pain that may result in the frequent use of pain medication. If your child is one of these, you and your child should see a team of pain experts for help. This team may include a psychologist, social worker, physical therapist, neurologist, and pain medicine doctor. The team will draw up a treatment plan for home and the hospital. It is important to follow all parts of the plan.

Transcutaneous Electrical Nerve Stimulation (TENS) unit

A Transcutaneous Electrical Nerve Stimulation (TENS) unit may help block the pain. It is a small device prescribed by the physical therapist. Your child can use it at home when they start to feel sickle cell disease pain.

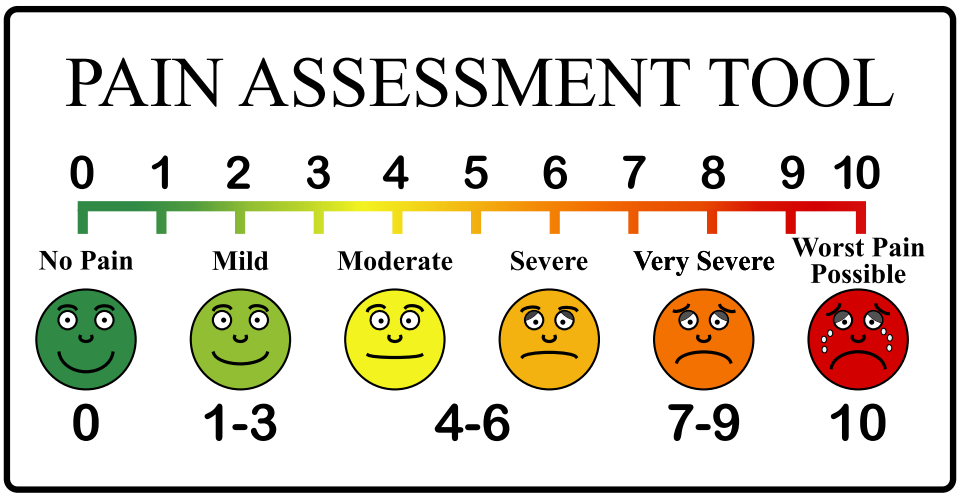
Describing the pain

You and your child must know how to describe the pain to get the right treatment. There are many ways to describe pain. Stick to the one that works best for your child. Make sure their doctor uses the same one too.

How severe is the pain?

First, your child needs to be able to say how severe the pain feels. They can give the pain a number from 1 to 10, with 1 as the mildest pain and 10 as the worst pain. They can also pick a face, from a big smile for no pain to a big frown for the worst pain. The pain rating scale below shows these different ways to rate pain.

Figure: Pain Scale



Where is the pain?

The doctor will also want to know where your child feels the pain. Sometimes it is easiest to describe where it is. Other times, it is helpful to point or mark the places on an outline of the body.

How does the pain feel?

Pain may feel sharp or dull, hot, or throbbing. If your child can't tell you how it feels, sometimes they can show you with color. Ask them to color how the pain feels and you can show it to the doctor.

Living with pain

Understanding and sympathy are important for helping your child when they are in pain. Pain itself is not a cause for panic. While sickle cell disease pain hurts, it rarely causes serious problems. Most of the time, it can be managed at home. This pain is usually not a sign of something worse. It is part of the disease and needs treatment. Also, even in a child with sickle cell disease, not all pain is caused by the sickle cell disease. If you have questions about the cause of your child's pain, ask their medical provider.

Get help with pain problems

If your child has many problems with pain, counseling may be helpful. Counseling can help your child learn to manage the pain. A counselor can also help other family members cope. One person in pain can put a strain on the whole family.

Don't let pain take over your child's life. Help your child find ways to ease their pain and to live with it and learn to not think about it. Also, make sure your child is taking the medication hydroxyurea. It is well known that hydroxyurea decreases pain episodes.

Pain in teens and young adults

Some teens with sickle cell disease have more pain as they get older. The pain may feel worse or just happen more often. The treatment for pain is the same for teens as it is for younger children. By the teenage years, if your teen has had a problem with pain, they have most likely tried lots of ways to ease it. It makes sense for your teen to use the ways to manage pain that work best for them.

Pain triggers

Dehydration (lack of fluid in the body) is a common trigger for pain. Teens often get involved in sports and don't take the time to stop and drink or rest. Teens need to take responsibility and find ways to take care of themselves since you can't always be there to remind them.

Although it's rare, some people get severe menstrual pain that brings on sickle cell disease pain. The doctor may be able to prescribe a medication to prevent this pain.

As your child gets older and becomes a teenager, they will learn more about what triggers their pain. As they do new things, suggest that they watch to see what happens with the pain. If they find that something often leads to pain, they can stay away from it or take special precautions.

Individualized pain plans

Your child's medical provider should have a pain plan created by your child's sickle cell disease or pain doctor with your child at a clinic visit when they were not in pain. The individualized pain plan can prepare your child to manage the pain when it starts.

Every medical provider might provide different looking pain plans. All pain plans should contain at least three detailed sections on how pain will be treated 1) at home, 2) in the emergency room (ER), and 3) the hospital. All older children and teens should have a copy of their individualized pain plan.

You and your child should have their individualized pain plan with them at home and whenever they seek care for pain at the clinic, ER, or hospital. An individualized pain plan will show how your child has received treatment in the past. The information can be either on paper or in an electronic form. You and your child can bring a photo of the individualized pain plan.

See the online Appendix for pain plans that can be individualized for your child in discussions with their medical provider.

Chapter 7:
Common Medical Problems (not Including Pain)

Introduction

This chapter describes the most common problems that your child may have because of sickle cell disease. Pain is not included here because it is written about in the previous chapters.

Signs of problems, ways to prevent problems, and treatment approaches are included to help you understand these medical issues.

This chapter discusses the most common medical problems in younger children:

- General infections (including an increased risk of life-threatening blood infections and meningitis)
- Pneumonia or acute chest syndrome
- Problems with the spleen or splenic sequestration
- Anemia (low red blood cell count)
- Hand-foot syndrome (also called dactylitis)
- Gallstones
- Problems with the kidneys and urine

This chapter also covers problems that increase with age:

- Eye problems (retinopathy)
- Leg ulcers
- Avascular necrosis (hip and shoulder pain)
- Delayed growth and puberty

This chapter additionally covers uncommon but serious medical problems. If your child has one of these problems, you can learn more about it. Be aware that the treatments discussed in this book could be different from the treatment that is given at your child's medical center. Many problems have multiple possible treatment approaches. Follow your medical provider's advice.

General infection

Infection has always been the leading cause of death in young children with sickle cell disease. However, with early treatment and preventive measures, death is uncommon.

Infections can be caused by bacteria or viruses. Bacteria are tiny organisms that can live inside or outside the body. Viruses are organisms that thrive by entering and multiplying inside the body. Many types of bacteria and viruses can cause infection.

The most common cause of serious illness or death in sickle cell disease is infection with a bacteria called “pneumococcus.” Many of the methods used to prevent infection are specifically focused on preventing pneumococcal infections.

Serious infections can occur in many parts of the body, including these parts:

- Blood
- Lungs (pneumonia, acute chest syndrome)
- Spinal fluid (meningitis)
- Kidneys or bladder (urinary tract infections, kidney infections)

Prevention

A study of young children with sickle cell disease from all over the country showed that taking penicillin twice a day prevents pneumococcal infections. The study showed penicillin does not cause harmful side effects. Now, all children 5 years old and younger with sickle cell disease are given penicillin twice a day to protect them from bacterial infections.

Children with sickle cell disease also receive pneumococcal and meningococcal vaccines (shots). These shots greatly reduce the chance of infection by pneumococcus and a related bacterium called meningococcus. This meningococcus bacteria causes meningitis.

Other routine vaccinations are also extremely important for your child. Your medical provider will give your child these other vaccines to prevent many other serious infections.

Signs

Fevers are one of the most important signs of infection. If your child has a fever of over 101°F, contact your medical provider and take your child to the clinic or emergency room (ER) right away.

Contacting your medical provider first, or while on the way to the medical facility, allows your medical provider to call the clinic or ER before you arrive. Your medical provider will tell them

that your child should be seen quickly. The call will make sure that your child gets the tests and treatments needed for a person with sickle cell disease. If your medical provider calls first, it will help your child get the best care possible.

Treatment

When you get to the clinic or ER, the medical provider will examine your child. They will take samples of blood and possibly urine and do a chest X-ray. The vital signs are important indicators of illness. They include the heart rate, breathing rate, and temperature.

It is also important that they check the oxygen level in the blood. This is checked by a pulse oximeter. A pulse oximeter is a machine that measures how much oxygen is in the blood.

If your child's fever is very high, if your child is very young, or if your child seems to be very sick, they may need a spinal tap to check for an infection in the spinal fluid. A spinal tap takes fluid using a needle from the spine in the lower back to check it.

The samples of blood, urine and spinal fluid will be sent to the laboratory (lab). The lab will look for what is causing the infection so that your child can get the proper treatment.

While waiting for the lab results, your medical provider may decide to admit your child to the hospital for antibiotics through an IV. If this is the case, your child will have to stay in the hospital on the antibiotics until there are results from the lab tests. This usually takes 1 to 2 days. If the tests don't show that there is a bacterial infection, it might mean the fever was caused by a virus. If your child's fever improves and your child seems well, your child will be able to go home.

If the tests do show that bacteria are causing the infection, your child will have to stay in the hospital on the IV medicine for several more days. Sometimes the medical provider will also tell you to give your child antibiotics at home for another week or so. It is important for you to give your child all the medicine the doctor ordered, even if your child seems well. This will help prevent the infection from coming back.

Meningitis

Meningitis is a severe infection of the membrane around the brain and spinal cord. It can be caused by a bacteria or virus. It is an uncommon but serious infection in people with sickle cell disease. Children with sickle cell disease are at greater risk for developing meningitis than other children. The signs of meningitis are the following:

- Sudden high fever
- Stiff neck
- Severe headache
- Nausea or vomiting

- Confusion or trouble concentrating
- Seizures
- Unexpected sleepiness or trouble waking
- Sensitivity to light

If you are concerned that your child has meningitis, contact your medical provider and take your child to the clinic or ER right away. If a medical provider is concerned about meningitis during an ER examination, they will do a spinal tap.

Sometimes, after children get over this infection, they may have some hearing problems. Hearing tests should be done to check for these problems.

Pneumonia or acute chest syndrome

Acute chest syndrome is the term used to describe pneumonia in people with sickle cell disease. Children with sickle cell disease are about 300 times more likely to get sick with acute chest syndrome than other children.

Acute chest syndrome can have many causes, including infection or sickle cells getting stuck in the lungs. Often pneumonia and sickling in the lungs happen together. They frequently develop during pain episodes. Low oxygen in the blood can happen at the same time.

Acute chest syndrome can also develop from an asthma attack. People with sickle cell disease appear to have a higher rate of asthma than other children. Treating asthma will minimize the risk of acute chest syndrome. Early treatment will keep the acute chest syndrome from getting worse. Warning signs that your child might be developing acute chest syndrome are the following:

- Fever
- Severe coughing
- Rapid breathing
- Shortness of breath
- Difficulty breathing or “grunting”
- Chest pain
- Extreme tiredness

If you see any of these signs, contact your medical provider and describe your child’s symptoms.

Prepare to go to the clinic or ER right away. Treatment for acute chest syndrome is given in the hospital.

Your child will have laboratory (lab) tests done and will be given antibiotics. They may need oxygen or other breathing treatments (like an asthma inhaler). Your child may also need a blood transfusion. Most of the time, these treatments make people feel better.

Your child may have multiple chest X-rays while in the hospital. They may also need to come back to the hospital to get a repeat chest X-ray within a few weeks after leaving the hospital. The medical provider may also repeat lung tests and an echocardiogram (an ultrasound of the heart) in a month or two to see how well the lungs have healed and to check if the heart was strained. Children who have had one episode of acute chest syndrome or asthma or both have an increased chance of having another episode in the future.

See Chapter 2, Routine Medical Care, for information on common medical tests.


Incentive spirometer

Acute chest syndrome can develop when a child does not take deep enough breaths. This condition can be triggered by a lot of things, including when children are given too much medication for pain or surgery.

To prevent acute chest syndrome, when your child is hospitalized for any reason (pain, surgery, dental work), your child should expand their lungs. They do this by blowing bubbles, blowing into a balloon, or using a device called an “incentive spirometer.” The incentive spirometer is a little plastic tool with a mouthpiece that will teach your child to take deep breaths, even when lying in bed.

A small number of children get acute chest syndrome often. They may have one infection after another. These children often need to get blood transfusions in clinic every month.

Sleep apnea in children with sickle cell disease can lead to acute chest syndrome and other sickle cell disease complications. Sleep apnea is difficulty breathing while sleeping. If you notice your child snoring or having trouble breathing, it might mean they are having lung problems. Ask your child’s medical provider for testing.

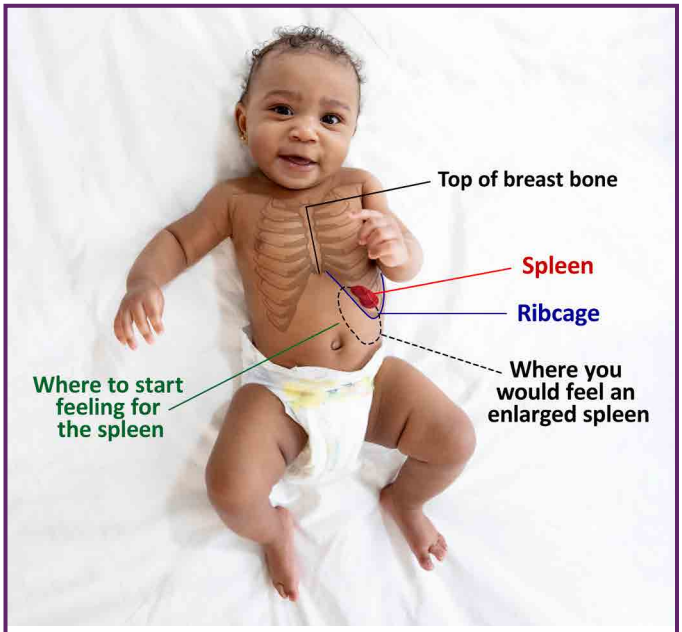
Problems with the spleen

The spleen is in the left upper corner of the abdomen, just under the edge of the rib cage. The spleen's job is to filter out damaged red blood cells from the blood and to help fight infections.

Enlargement and scarring

Most young children with sickle cell disease get enlarged spleens sometime in their first two years of life. One reason it is important for your child to see a medical provider often is to have the size of the spleen checked when they are feeling healthy. Then, if your child becomes ill, the medical provider will know whether the spleen is larger than usual.

Parents can learn to check their child's spleen's size following the instructions below. If you have not been instructed, you should ask your provider to teach you. Your child's medical team can show you how to do this at each visit. If your child's spleen seems to be getting larger or firmer, call your medical provider so they can check your child.



The spleen is a small organ (see red oval) that can get bigger and firmer if there is a problem (see dotted lines)

Different types of sickle cell disease affect the spleen differently. In most children with sickle cell anemia (SS disease) and sickle beta zero thalassemia (SB^0) disease, the spleen stays enlarged for several years. By 6 years of age, their spleen usually becomes small again because of scarring from sickling. However, the scarring can be delayed in children who are taking hydroxyurea. Also, children with sickle C disease (SC disease) and sickle beta plus thalassemia (SB^+) disease often have enlarged spleens for a longer time, sometimes for their whole lives.

A mildly enlarged spleen doesn't usually cause any problems. However, scarring keeps the spleen from working well. This is one of the reasons that children with sickle cell disease can get serious infections.

See Chapter 1, Basic Questions, for more information on the spleen.

Steps for feeling for your baby's spleen:

1. Have your baby lie comfortably on their back with something entertaining. Practice feeling the spleen when the child is not ill.
2. Look at your baby's chest and belly area. Locate their nipples, belly button, and the bottom of their neck where the breastbone starts. The breastbone meets in the middle. You can feel the start of the breastbone below the neck.
3. Run your finger down in a straight line from where the breastbone starts to the end of their belly. Stop an inch and a half above the end of the belly button and feel the area where the spleen is located. It is on the left side of your baby's belly, below and under the ribcage. For most spleens, if you move too far across it will no longer feel soft and you will be touching the bone of the ribcage. The spleen is very close to the surface of the belly (right under the skin), and you do not have to push hard to feel the spleen. The baby might be ticklish so don't rush.
4. Press lightly on the skin (act like you are just playing with your baby so you don't give them any discomfort or pain) and keep pushing your fingers down softly to the baby's left side (from your view, moving to the right) until you reach your baby's rib bone. Most likely when you do this, the belly feels the same. The belly should feel soft like a water-filled balloon. It should feel this way until you move across the belly to the baby's left side.
5. If the spleen is enlarged, it will feel different from the soft belly (not feel like a water-filled balloon), and it will be more firm and dull. It will feel more like muscle or tissue. A good way to see this is when your baby breathes in. Take a few minutes to watch your baby breathe. Sometimes you may be able to see a little bit of an outline in the area below the left side of your baby's rib cage.
6. Repeat these steps multiple times and mark any areas (with a child safe marker) that feel different from the soft belly with a dot. You can take a picture of it to know where you found it and repeat the exam later.
7. You may or may not be able to feel the spleen. What is important is to know how the spleen area usually feels. That way you will notice any change from the usual.
8. If you think you feel an enlarged spleen or anything that doesn't feel normal, mark the area, take a picture, and contact your doctor right away.
9. Always remember to be gentle. You can avoid causing discomfort or pain to your baby during the exam.

Splenic sequestration

Normally, blood is pumped into the spleen to be filtered and rapidly returns to the bloodstream. In some children, the spleen becomes larger very quickly. This is because the spleen swells with blood trapped inside it. This can cause a sudden drop in the red blood cell count causing severe anemia. This can lead to heart failure and death if not treated promptly with a blood transfusion. Rapid spleen enlargement with dropping red blood cell count is known as a "splenic sequestration."

Signs

Splenic sequestration can happen when your child has a fever or a cold. Signs to watch for are abdominal pain or swelling, pale color, and tiredness.

Treatment

If your child has one episode of splenic sequestration, they are more likely to have another. If your child has splenic sequestration and is at least 2 years old, it is very likely their spleen will be removed with surgery so that this won't happen again.

Your child does not need their spleen to live and be healthy. However, if your child has their spleen taken out, they should keep taking penicillin twice a day to help prevent infections. Some doctors recommend continuing penicillin for life. Other doctors stop daily penicillin after about 3 years past the spleen surgery. For children without a spleen who stop taking penicillin, they must have a small supply of penicillin at home. They take this any time that they get a fever.

Anemia (low red blood cell count)

People who have sickle cell disease have fewer red blood cells than usual. The degree of anemia or decreased blood cells varies between the types of sickle cell disease. Sickle cell anemia (SS disease) is the most common type and often has more severe anemia. Sickle C disease (SC disease) usually has mild or no anemia.

Because red blood cells carry oxygen to the muscles, severe anemia may cause an increased need to rest during exercise or strenuous activity. However, even in people with severe anemia, the body learns to adjust to the low red blood cell count. This means that children with sickle cell disease can participate in most activities that children without sickle cell disease do, including exercise and playing sports.

There are times when your child's red blood cell count may fall much lower than usual. This can happen with a fever or an infection. Either the body stops making new cells or the cells are destroyed more quickly than usual. When this happens, the destroyed red blood cells in the body fluids can make the eyes look more yellow and the urine look darker than usual. Yellowing of the skin and eyes is called jaundice.

Signs

Bring your child into the doctor to have a red blood cell count checked if you notice any of these signs of an extra low red blood cell count:

- More tiredness than usual
- Pale color
- Loss of appetite
- Yellow eyes or skin (jaundice)
- Dark urine

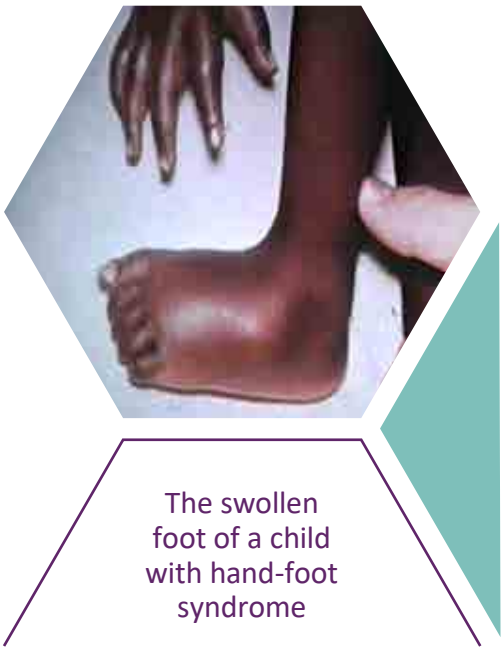
Treatment

If your child's red blood cell count falls very low, they may need a blood transfusion. An extremely low red blood cell count can result in heart failure and death if not treated in time. Since the blood count often falls at the time of other medical problems, your medical provider will usually check it frequently when your child is in the hospital.

Hand-foot syndrome (dactylitis)

Signs

About a third of children with sickle cell disease younger than 3 years old may get painful swelling of their hands and feet. This is known as the hand-foot syndrome or dactylitis. It is caused by the sickled red blood cells blocking the tiny blood vessels in the bones of their hands and feet.



The swollen foot of a child with hand-foot syndrome

Treatment

In most children, the pain is mild enough to be treated at home. Acetaminophen and extra fluids help with the pain. The swelling goes down within a day or two.

Your doctor may also prescribe stronger medicine like oxycodone or another opioid. Follow your doctor's instructions in giving this medicine. It is safe when taken as directed and will decrease the pain. Many children have constipation (hard poops) while taking opioids. If this happens, give your child extra fruits and other foods high in fiber.

If your child has a fever or if the pain is very severe, contact your medical provider. Your child might need to go into the hospital for a few days to receive fluids through an IV and stronger pain medicine. Almost always, dactylitis will go away without any lasting effects.

Gallstones

About a third of children with sickle cell disease have gallstones by 7 years of age. Gallstones are formed from the waste products of broken-down red blood cells. These waste products collect in a small organ called the gallbladder and form thick sludge or stones.

Gallstones are not harmful unless they cause a clogging of the gallbladder. But if they get stuck in or near the gallbladder, they can cause blockage, liver swelling (the liver is right next to the gallbladder), or a serious infection. Most short-term episodes of gallbladder pain can be treated right away. Planned surgery can happen later. Occasionally the blockage could be severe enough to require urgent surgery or other procedures to remove the gallstone.

Signs

Jaundice (yellow skin and eyes) can be a sign that gallstones are stuck. Often, there is a warning before the stones get stuck. The warning is pain on the right side of the belly that happens when the stones pass on their way from the gallbladder. The pain can often be triggered by fatty foods.

Treatment

If your child has an episode of gallstones that causes pain and jaundice, taking out the gallbladder is often the treatment to avoid future problems. People do not usually have any medical problems because they are missing a gallbladder. Your child's diet does not have to change if the gallbladder is removed.

Yellow eyes or jaundice

People with sickle cell disease can have yellow eyes or jaundice from time to time even without gallstones. This is caused by a yellow-colored substance called "bilirubin" that comes from broken down red blood cells. In some people, the yellow color lasts for a long time. It may always be there.

It is not a medical problem unless the eyes are much more yellow than you've seen before. In that case, it might be a sign that red blood cells are breaking open more than usual. It might be a sign that your child has a problem with the gallbladder. Both of these problems can be dangerous. If the color is much stronger, contact your medical provider to see if your child needs to be checked.

Problems with kidneys and urine

The kidneys help the body keep and get rid of fluid. In sickle cell disease, the sickle cells damage the kidneys so that even young children urinate (pee) more frequently than other children.

Dehydration

Children with sickle cell disease drink more fluids and urinate more often than other children. When a child becomes sick and drinks less than usual or loses fluid by vomiting, diarrhea, or fever, they can get dehydrated. Strenuous exercise can also cause dehydration. This can be prevented by making sure your child has easy access to fluids before and during exercise.

Signs of dehydration:

- Tiredness
- Dry, sticky mouth and lips
- Sunken eyes or sunken soft spot (on a baby's head)
- Urinating much less (fewer wet diapers for babies)

Urinating less may not be a reliable sign, though, because children with sickle cell disease may continue to urinate the same amount despite dehydration. If you notice any of these signs, give your child extra fluids to drink and contact your child's medical provider.

Bedwetting

Frequent urination can cause bedwetting, particularly at night. This is a common problem in all young children without sickle cell disease. However, older children with sickle cell disease may continue to wet the bed. Bedwetting is not your child's fault. In many children with sickle cell disease, nighttime bedwetting happens because there has been early sickle cell disease damage to part of the kidney. The general function of the kidney may be normal when your child wets the bed, but they might be unable to control the bedwetting. Most children stop bedwetting when they are older and can get up by themselves to use the bathroom.

Providing positive support can help your child stop wetting the bed. Do not use punishment or make your child feel guilty. It may help to wake your child or set an alarm clock to go off during the night so your child can go to the bathroom. But children also need their sleep. Your medical provider may have other helpful ideas. As a last resort, you can discuss the use of the medicine desmopressin with them. Desmopressin is a pill or a liquid taken before bed.

Kidney and bladder infections

Blood is filtered by the kidney. After it is filtered, the urine is put into a tube called the ureter. The ureter empties into the bladder. Its purpose is to store the urine and remove it through

a tube in the penis or near the vagina. Urinary tract and bladder infections are common in all people. They are more dangerous in children with sickle cell disease, however, because there is a greater chance the infection will spread to the kidney. This is serious and can cause kidney damage.

Contact your doctor or nurse if you notice any of these signs:

- An increase in the number of times your child goes to the bathroom
- Bedwetting after your child has stopped wetting the bed
- Losing control of urine or urine accidents during the day
- Foul smelling or cloudy urine
- Fever
- Burning and pain when urinating
- Abdominal or back pain

Your child will have urine tests every so often to look for infection of or damage to the kidneys. The infection can show up in the test as protein in the urine. If your child has a bladder infection, they may need to have a urine test more often to make sure that the infection has not returned.

If your child gets repeated bladder infections, your child will need to have testing to make sure the bladder and kidneys are normal. Your child may have to take an antibiotic other than penicillin every day to prevent repeated infections.

Blood in the urine

Another problem caused by sickle cell disease is blood in the urine because of bleeding from the kidney. When this happens, the urine usually looks bright red or brownish. In more severe cases, there may be back pain and small pinkish specks or lumps in the urine (these are blood clots). Usually, the blood disappears from the urine within hours. In some cases, the bleeding can go on for days and become a serious problem.

Always contact your medical provider right away if you notice blood or blood clots in your child's urine or diaper. Tests should be done to find out what is causing the bleeding. In most cases it is due to sickle cell disease damage to a small blood vessel in the kidney. This usually does not cause permanent kidney problems.

However, there are uncommon causes of kidney bleeding that should be ruled out. It is very important to get plenty of fluids, sometimes through an IV in the hospital, and to rest in bed during the bleeding. Many people may need a blood test and an ultrasound of their kidneys. If the problem continues, there are medications to stop the bleeding.

Problems that increase with age

Teens have many of the same problems as younger children with sickle cell disease. Some of the problems that we have already described are more common in older children and teens than younger children. These include:

- Pneumonia or acute chest syndrome
- Gallstones
- Pain (aside from dactylitis, which occurs in infants)

In addition to the problems listed above, there are other common health problems that usually only affect teens and adults with sickle cell disease. These are:

- Eye problems (retinopathy)
- Leg ulcers
- Avascular necrosis (hip and shoulder pain)
- Delayed growth and puberty

Eye problems (retinopathy)

Sickle cell disease can cause eye damage and, rarely, blindness. This is why your teen needs to be checked by a special eye doctor once a year. They are called an ophthalmologist. Early detection and monitoring can prevent the development of poor vision and eye complications.

The back of the eyes contain tiny blood vessels that can become clogged by sickle cells. This can cause bleeding or scarring at the back of the eyeball. When the damage starts, you can't see it. Only an eye doctor using special techniques can see it. If the doctor finds damage, it can be treated. Without treatment, these early changes can lead to loss of vision.

Signs

In the early stages of retinopathy, there may be no symptoms of eye problems but there can still be damage. In later stages, your child may complain of the following problems:

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

Treatment

Early treatment by an eye doctor may include laser treatment.

Hyphema (blood in the white part and front of the eye)

Hyphema is also related to the eye. It is not uncommon for children to get eye injuries that result in bleeding in the front or white part of the eye. This could be serious but usually goes away completely without any treatment.

In children with sickle cell disease, once the bleeding happens, sickle cells in the blood can block the drainage of the eye. This can cause the pressure in the eye to rapidly increase (called "glaucoma"). Glaucoma can damage the eye. The child should see an ophthalmologist as soon as there is bleeding. If the pressure in the eye is too high, a simple procedure will fix it.

Leg ulcers

Leg ulcers may be a problem for teens with sickle cell disease. They usually start as a small sore on the ankle. They can grow large and get infected. Some leg ulcers heal quickly, but most take a long time to heal.

Sickling happens more in the lower legs and ankles because of the pressure of standing. With sickling, small blood vessels get blocked. Blood can't get through to all of the cells. A leg ulcer forms when skin cells in the ankle die because of lack of blood flow.

Men are more likely to get leg ulcers than women. People with sickle cell anemia (SS disease) are also more likely to get leg ulcers than those with other kinds of sickle cell disease.

It is much better to prevent and treat leg ulcers when they are small than when they are larger. Leg ulcers, once they happen, frequently happen again. Before the leg ulcer is formed, some children show swelling and eczema in the area. Once the leg ulcer forms, it can hurt.

Prevention

Leg ulcers develop in a warm environment. They are sometimes aggravated by badly-fitting shoes or socks.

- Make sure socks and shoes are comfortable and well-fitting
- If the area shows any changes, have your medical provider examine it
- It may be helpful to treat the skin over the ankles when there is any cut or rash

Any child who has had one leg ulcer has a higher chance of getting another one, so they need to be careful.

Signs

Take your child to see the doctor if you see any of these signs of leg ulcers:

- A cut or wound that doesn't heal
- A patch of dry, itchy skin
- A small dark spot that is surrounded by painful swelling

Treatment

These are the basics for good treatment for leg ulcers:

- Keep the area very clean
- Stay off the feet as much as possible
- Elevate the feet as much as possible
- Put on a fresh bandage twice a day or as often as advised
- Wear clean white cotton socks and flat protective shoes until the leg ulcer is healed
- Use lotion or ointment to keep the skin moist
- If the leg ulcer looks infected, see your doctor for antibiotics

If the leg ulcer is large or has not started to heal in a few weeks, your child may need to go into the hospital. They will be on strict bed rest and get special wound care there. Medical providers may use transfusions to try to bring more oxygen to the tissues.

If the leg ulcer doesn't heal, surgery may be needed. Surgery can move a piece of skin from the thigh to cover the leg ulcer. This is called a skin graft. It requires a longer stay in the hospital for strict bed rest. If it doesn't work, another skin graft may be needed.

Avascular necrosis (bone damage)

Avascular necrosis of the hip is not a common problem in the general population. It occurs in the top part of the thigh bone (femur) called "the head." The head attaches to the pelvis. In a person without sickle cell disease, avascular necrosis usually occurs in much later years. However, with sickle cell disease, avascular necrosis can happen in children as young as 5 years of age and in young adults.

The symptoms are usually in the hips, but the shoulder joints can be affected and cause pain. Many people who have one hip affected will eventually have the other hip affected.

When the blood flow to the hip joint is slowed by sickle cells, the bone in the joint becomes flat and crooked. Then the hip can't move freely. Walking puts pressure on the joint, and more damage is done.

Signs

Your teen will know if they have this problem. Their hip will hurt when they walk or run. They may also have lower back or thigh pain and develop a limp. Many people have this problem and do not complain of severe pain. Sometimes the groin or buttocks hurt. In most cases, weight bearing or a lot of motion on the hip will cause pain.

Sometimes hip replacement surgery is needed and can leave a scar

We often see young people who justifiably want to engage in dance or play vigorous sports. These activities aggravate early avascular necrosis and cause more hip pain. Bed rest following the hip injury may eliminate the pain temporarily. When adults return to dancing or other activities with ongoing repetitive movements, the pain often recurs. These young people often have to be readmitted to the hospital shortly after being released, with pain in the same place.

Sometimes severe pain can be stopped by seeing a physiatrist, rehabilitation doctor, or physical therapist. A physiatrist is a doctor who specializes in physical medicine and rehabilitation. These medical providers can fit the child with a special lift in the shoe. At your sickle cell disease clinic visit, you can have your child's medical provider examine the hip range of motion. The provider can do this even if your child has no symptoms. They do this to make sure range of motion is not limited. Limited range of motion can be an early sign of injury to the joint.

Report any severe pain in the hip joint, legs, or shoulders to the medical provider as soon as possible. Many young people who have avascular necrosis of the hip may develop some injury to the shoulders. This is usually pain free because the shoulders do not carry a lot of weight. If your child has this problem, make sure they do not hang a purse or backpack on their shoulder.

Testing

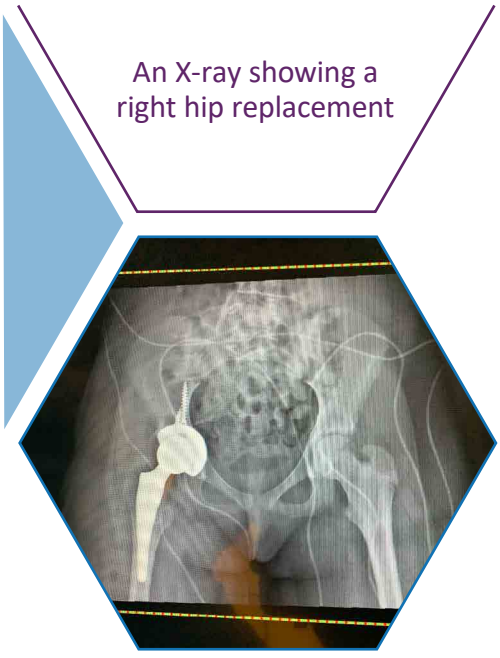
Regular examination of the hips, as well as magnetic resonance imaging (MRIs) and X-rays, are commonly used to test for avascular necrosis. MRI of the hips is the best test to find the disorder. MRI can pick up early damage that is not seen on an X-ray.

Moving joints in the full range of motion regularly is the best prevention care. If your child cannot move their hip completely, physical therapy may help. Physical therapy teaches the child how to walk and strengthen the muscles around the hip to avoid injuring the hip.

Treatment

Early treatment helps. The treatment depends on the amount of change on the imaging tests and the amount of pain the person is having. Sometimes small changes are found that may progress over time. Repeat imaging can show if the condition is worsening and if your child should change treatment.

Sometimes a person needs to use crutches for a few months to take the weight off of the joint. Other times, physical therapy or surgery might stop the hip from changing shape. A detailed physical therapy plan is recommended with multiple weekly physical therapy sessions. Then your child can do the exercises at home themself without ongoing physical therapy visits. Keeping a healthy level of vitamin D is important for the body to repair the bone.



An X-ray showing a right hip replacement

Surgery

There are two main types of surgery: hip core decompression and hip replacement. Hip core decompression is a simple operation that sometimes may not require a hospital stay.

Hip core decompression provides pain relief. It may also slow the progression of necrosis. The operation requires an orthopedic surgeon to take out a core (or several cores) of damaged bone. Coring stops pain by decreasing the pressure in the joint.

Hip replacement is a major operation only considered in the later stages of avascular necrosis for pain relief. When the head of the thigh bone is severely damaged, it may need to be replaced. This usually occurs in older people. Hip replacements are not done in children whose bones are still growing. Other therapies are offered.

If your teen can't walk without severe pain, the hip may need to be replaced. This can only be done when the bones have stopped growing. If your teen needs this treatment, they will need to wait until they have grown to their full size.

Delayed growth and puberty

Most children with sickle cell disease grow in the usual way when they are young babies. After their first birthday, they may start to grow more slowly. Throughout the rest of childhood, some children with sickle cell disease are shorter and thinner than other children their age or than their siblings were at the same age. Puberty can also be delayed due to this slow growth.

In almost all cases, this difference in size is only temporary. Children with sickle cell disease keep growing after their friends have stopped. After a while, they will reach the height expected based on the size of their parents. If your child is concerned about being small, let your child know that they will grow bigger like their friends and other children, it will just take a little longer.

If your child is very thin and much smaller than normal for their age, eating extra food between meals or before bed at night may help with weight gain and better growth. Your medical provider may also want to give your child extra vitamins or minerals. Most children don't need any special diet or vitamins and won't grow any faster if these are given.

As with all children, they should eat three meals and several snacks each day. Limit the amount of candy, sugar-sweetened beverages (like sports drinks, energy drinks, fruit drinks and sodas), and other "junk foods." These are not good for children in general. While being thin has historically been an issue, obesity is increasingly becoming an issue in people with sickle cell disease. Junk foods are especially unhelpful for children who have sickle cell disease because they can lead to additional health problems.

Late puberty

Children with sickle cell disease may begin puberty later than other children. This may result in emotional issues. With puberty, there are many physical and emotional changes. These changes are the same for teens with and without sickle cell disease. There is a difference in the age when the changes happen for biological males and females. Females usually go through puberty at a younger age than males. For females, puberty is when their breasts grow and their periods start. In males, their face hair grows, their muscles get bigger, and their voices deepen. Once a person has gone through puberty, that person can get pregnant or can make someone pregnant.

Some teens with sickle cell disease reach puberty about two years later than others their age. Late puberty is not a problem in most cases. Sometimes a referral to a medical specialist called an endocrinologist is helpful. You should discuss this with your medical provider. While most growth problems will resolve on their own, they may result in your teen feeling bad about themself. Talk to your doctor or a counselor if you or your teen feel concerned about this issue.

Sickle cell disease and appearance

Some children and teens with sickle cell disease can be concerned about their looks. They may

be jaundiced, smaller than their peers, delayed in puberty, or have another reason. If your child feels this way, reassure them. You and other family members can support them in feeling better about their appearance and who they are. It may also be helpful for them to talk with a counselor or other teens with sickle cell disease.

See the online Appendix for a link to the California Clinical Network and Community Based Organizations web page.

Uncommon but serious medical problems

Less common, but very serious problems for people with sickle cell disease of all ages are:

- Stroke
- Priapism (prolonged, painful erection)

Strokes

Strokes are caused by sickle cells blocking blood vessels in the brain. This can cause the brain to not work properly. A full stroke is usually paired with weakness, or inability to speak, and other symptoms.

Strokes that cause sudden symptoms are a very serious but fairly rare problem caused by sickle cell disease. They are called “overt,” or “clinical,” strokes.

There are multiple risk factors for stroke. First is very severe anemia. Second is a history of symptoms of stroke that last hours and then disappear (this is called a “transient ischemic attack,” or a “TIA”). Third is a history of silent strokes on radiographic studies. Fourth and extremely important is an abnormal transcranial doppler during screening. This is further explained below.

Three things that will decrease your child’s chances of having a stroke are:

- Taking hydroxyurea regularly
- Getting regular stroke screens (also called transcranial doppler, explained below)
- Monitoring blood pressure

Signs

Strokes happen to fewer than one in 20 children who have sickle cell anemia (SS disease) and even fewer children who have sickle C disease (SC disease) and sickle beta plus thalassemia ($S\beta^+$) disease.

The following can be signs of a stroke:

- Sudden weakness of an arm or leg or the whole body
- A difference in the way one side of the face or one eye moves compared to the other side
- Sudden strong headache
- Seizure (shaking that can't be stopped)
- Difficulty speaking
- Fainting

If you see any of these signs, contact your medical provider and bring your child to the hospital right away. The sooner the child is seen at the hospital, the better. Overt strokes are diagnosed by their signs and symptoms and an abnormal brain scan. A common type of brain scan used to look for a stroke is a computed tomography or CT scan (also sometimes called CAT scan).

Treatment

If diagnosed, an overt stroke requires your child to be admitted to an intensive care unit (ICU) and to receive an immediate red blood cell transfusion. The team taking care of your child will generally include an intensive care doctor and medical specialists. Specialists include a brain doctor (neurologist), a rehabilitation doctor (physiatrist), and a physical therapist, in addition to a hematologist (blood doctor).

In addition to a CAT scan, another kind of brain scan is called an MRI. Your child will need an MRI to get a more detailed picture of what happened in the brain. Most children treated promptly with a transfusion do very well. Occasionally surgery or other therapy is needed to remove a blood clot in the brain.

After a child with a stroke is well enough to go home, they will require a blood transfusion in the clinic every month for about 5 years to minimize the chance of another stroke. Eventually, these children will also need to take a medication called a chelator to remove the excess iron from all the transfusions.

To help your child recover from a stroke, take your child to a center that has all of these services:

- Rehabilitation program, including physical, occupational, and speech therapy
- Neuro-psychological testing and care
- Experience with iron chelation

Small, silent strokes

Overt strokes are rare. By contrast, silent strokes are common and can have no symptoms. Silent strokes may increase in frequency and size as a child gets older.

The chances for a silent stroke are one reason it is so important for your child to see their medical providers regularly.

These small, silent strokes are because of sickling in small blood vessels. Though they don't cause sudden, severe symptoms, silent strokes can cause learning, memory, and behavior difficulties. Difficulties are caused by small injuries of certain learning areas. Silent strokes increase the chances of future strokes and brain problems.

MRIs give a detailed picture of the brain. MRIs are used in sickle cell disease to detect both overt and silent strokes.

Screening for strokes

Transcranial Doppler (TCD). Transcranial Doppler or TCD is a mini ultrasound. It is similar to the ultrasounds done during pregnancy. The TCD is placed on the side of the head. The TCD does not detect strokes. It is a screening test done once a year in children 2 to 16 years old to find children with a higher chance of strokes in the future.

The testing is most helpful in sickle cell anemia (SS disease) and in sickle beta zero thalassemia (SB^0) disease. In sickle C disease (SC disease) and other sickle types that are usually milder, the benefit of TCD screening has not been proven.

A TCD can detect narrowing of the major blood vessels of the brain. Blood vessels can be narrowed because of sickle cell disease damage. When this happens, the blood travels more quickly through the narrow area. It is similar to holding your finger over a hose forcing the water to rapidly flow through the smaller open area.

The TCD may find that there is faster flow in one of your child's vessels. That means that the blood vessel may be narrower. It also means there is a greater possibility for having a stroke in the future.

The results of the TCD are reported as either "normal," "conditional," or "elevated." Finding an elevated TCD early is important to prevent a full stroke. People with an elevated TCD are at about 40% increased risk for developing a full stroke. A child with an elevated TCD result requires more frequent TCDs. They often require monthly red blood cell transfusions, and then may be transitioned to hydroxyurea.

Neurocognitive and academic screening

Anemia and sickling can result in learning challenges. This is true in all children with sickle cell disease. It is especially true in children who have had strokes. While children with sickle cell disease have the same intelligence as children without the disease overall, they may have specific areas of learning difficulty. These include memory and the speed at which they process questions. These learning differences could make learning and studying harder than it is for other children.

All children with sickle cell disease should be screened for learning differences through their medical provider's office, through school, or both. Finding these learning differences through screening tests means that teachers and school staff can provide support for your child. Support will improve your child's success in school. Just as strokes can cause learning difficulties, sometimes, learning difficulties may be an early sign of sickling in the brain that require more frequent monitoring with MRI, TCD, or both.

Priapism

Priapism is a painful unwanted swelling of the penis that can occur in male children with sickle cell disease. When this happens, the penis becomes erect, hard, and painful.

Priapism is a serious problem in boys with sickle cell disease. It often goes undetected until it's an emergency. Priapism can happen if sickle cells block the blood vessels in the penis.

This is different from a normal erection. It can happen at any age. It is important to educate the child of this potential complication and the importance of telling the family and medical provider when it happens. Repeated untreated episodes can result in permanent damage. If children have had priapism, they may need to take medications.

Stuttering Priapism

Stuttering priapism is the most common type of priapism. In general, it refers to short episodes of priapism that last for a few minutes to an hour. A history of stuttering priapism is associated with an increased chance for longer episodes of priapism in the future. Most often, these short episodes occur in boys or young men in the middle of the night or the morning and the penis becomes soft without any treatment. If your child has a case of stuttering priapism, it is important to discuss it with your medical provider. There are medications that can be helpful.

At home, the initial management of episodes for priapism include drinking more water, pain medication and warm or cold compresses. If it does not improve after 2 hours, bring your child to the ER and speak to a urologist. A urologist is a specialist for the genital area. Your child may require IV hydration, pain medications, oxygen, or transfusions. They may need to undergo a minor procedure to stop the priapism.

Chapter 8:
The Emergency Room and the Hospital

Introduction

Emergency room (ER) visits and hospital stays can be difficult times for your child and your family. Your child's medical team will do whatever they can to keep you and your child as comfortable and informed as possible. They want to answer your questions. They want to help you in every way they can while taking the best care of your child's medical problems.

Many ER visits and hospital admissions for children with sickle cell disease are unplanned and happen because the child has become suddenly ill. This adds to the stress of being in the ER or hospital. It may make parents feel they have no control over what is happening. This is normal, and you should let a social worker or your child's medical provider know what you are feeling. They can help you.

This chapter will describe what you can expect and things that can help with each part of an ER visit and hospital stay. We have also included information about some of the procedures that may be done to your child in the hospital. The following sections will be included in this chapter:

- The hospital emergency room (ER)
- Getting admitted (checked in)
- Getting ready for the hospital
- In the hospital
- Medicines and IVs
- Surgery
- Going home

Different centers and medical providers may use other approaches to treating these problems. Follow your doctor's advice.

The emergency room

There are times when you may need to take your child to the ER. If your child is having any of the signs listed on the last page of Chapter 1, your medical provider may tell you to go to the ER.

See Chapter 1, Basic Questions, Danger signs: When to contact a medical provider.

Always try to contact your medical provider before going to the ER. If you can't reach your medical provider, then contact the ER so that they can get ready to see your child. Make sure they know that your child has sickle cell disease and that you have a true emergency.

Your medical provider may give you or your child an individualized pain plan or treatment information that describes your child's type of sickle cell disease and how it is treated. If you don't have one, ask about it. Don't forget to bring this information with you and show it to the ER staff.

It is also wise to keep trying to contact your medical provider. Your medical team will help advocate for your child. They will try to make sure your child gets the right care in the ER. You should ask the ER staff to contact your medical provider as well.

Starting with the ER

Several people will help you with the process of being checked into the ER. First, you will usually meet someone who asks questions about you and your child. This is the admission clerk or triage nurse. It is very important that you let them know that your child has sickle cell disease. You should describe any medications your child is taking.

Do not assume the ER staff know what sickle cell disease is. They will not know why you have brought your child to the hospital. Tell the admission clerk the name of your child's sickle cell disease provider. Let the clerk know that your provider has told you that your child should be seen right away.

Next, your child's name will be called. An ER Nurse will take you and your child to a treatment room. They will examine your child and pass this information onto the ER doctor. The doctor will also examine your child and ask you questions. The doctor might consult with additional doctors.

Your main doctor in the ER will decide whether to admit your child to the hospital or to treat your child in the ER. Check that the ER staff have spoken with your child's primary sickle cell disease or medical provider so that the ER has information about your child's sickle cell disease. For example, the ER doctors should find out your child's usual hemoglobin level.

Planning ahead for emergencies

To plan how you would manage getting your child to the hospital in an emergency, think about the answers to the following questions:

- ☐ Where is the nearest ER?
- ☐ Who will take your child to the ER?
- ☐ Who will watch your other children if you need to take your child to the ER?

The hospital

Getting admitted to the hospital

If your child is admitted (checked in) to the hospital through the ER, several people will help you with the admission process. A team of hospital support staff will help you and your child to physically move from the ER to the hospital floor. It often takes a while (up to several hours), for your child to be set up in the hospital room.

This can sometimes cause a break in giving medication like pain medication. It is very important that your child's pain is as well controlled as possible before the actual move to the hospital room. If you are concerned about a break in your child's medication, ask for your child to get a dose of medication just before leaving the ER for the hospital floor.

If your child is admitted directly to the hospital on orders from their doctor's office or clinic (meaning you do not first go through the ER), the doctor will tell the hospital why your child is being admitted. Then once you arrive, an admissions clerk will register your child.

Whether your child is admitted from the ER or directly to the hospital, once you reach your child's room you will meet a nurse who will examine your child. They will help both of you prepare for the hospital stay. You will then meet the admitting doctors. These doctors will ask a lot of questions, examine your child, and decide what orders they need to write for your child. The orders could include medications, breathing treatments, and consultation with doctors from other teams.

While all this is going on, you and your child may have some worries or concerns about what may happen. Many people feel fearful about coming into the hospital. It can be helpful to talk about your concerns with the hospital staff. A social worker can help get your questions answered and provide support. You can also talk to other parents or the chaplain at the hospital.

In most hospitals, children share a room with other children. Because of this, each floor or unit has rules of conduct they expect all children to follow. To help the hospital stay go well, it is important that you and your child learn these simple rules. If you have any questions, ask the nurse or social worker about them.

Getting ready for the hospital

Thinking about your child's hospital stay can be overwhelming. Who will take care of your child? Who will visit your child and how often? Who will watch your other children when you are at the hospital? How will you deal with hospital costs? Talk to the social worker at the hospital if you have these concerns and need help to make a plan.

The best way to prepare for future hospital stays is to learn about what will happen at the hospital. At your routine medical visits, ask about what to expect at your child's first hospital stay.

Your hospital may also be able to give you tips on how to talk to your child about going to the hospital. You may want to visit the children's floor of your hospital to see it and meet the nurses. If you have other children, include them on the hospital tours.

Often, your hospital will have information that can help you and your child. The hospital may have information that explains the resources available to help children and their families. Some hospitals have child life specialists who use play to help children manage their feelings about being in the hospital. On or before arrival, get the hospital's brochure that explains the details about services and operating procedures. The brochure can also provide information about services such as financial support, food, overnight housing for parents, playrooms, social services, visiting hours, and approval for family members to stay overnight.

There are often many medical providers who will care for your child in the hospital, including the following:

- Supervising pediatrician, who is called "the attending doctor"
- Pediatrician who is training to be a supervising doctor in hematology (a blood specialist), called a "fellow"
- Pediatrician who is still in the final stages of being trained to be able to work independently as a pediatrician, called a "resident or intern"
- Primary nurse who is at the bedside and will provide the hour-to-hour care and support for you and your child

These providers usually work in shifts and rotate in and out of the hospital. Find out the names of these providers at the beginning of the hospital stay. Try to stay updated when these providers change. They change often, which can be hard.

It may be helpful to know the charge nurse. This nurse manages and organizes all the nurses on the hospital floor. This nurse can give you guidance on issues. These issues may include justified reasons to move to a different room. If there are problems with your room, please make a list and tell the charge nurse about them and ask them to correct it. In general, the hospital maintenance staff will fix issues such as burned-out light bulbs and the television not working properly.

You can use play to help your young child understand what may happen to them in the hospital. Get a toy doctor's kit and let your child "doctor" stuffed animals, dolls, and even you. Watch your child's play. Correct any wrong ideas they might have about what will be happening in the hospital.

There are some things that you should plan for before your child needs to go into the hospital. Figure out who will take your child to the hospital. You should also make plans for who will stay with your child during the day and overnight if they go into the hospital.

For children who have to stay in the hospital overnight, it is best to have a parent spend the night. At night, your voice and physical presence can comfort your child. During the day, if a parent can't be there, the hospital may have volunteers who can sit with your child for short visits. They can keep them company and help them be entertained and comfortable. Often some families plan in advance with relatives who know the child. Grandparents and other significant people can provide relief and backup for the parents.

You may have other children or other things you need to take care of. If you don't think that you will be able to spend time at the hospital, talk this over with the hospital staff. They may know of resources to help you and your child.

Keep in close contact with your child's nurses. If you can't stay at the hospital, you might ask to set up a schedule for video or phone calls and visits so that the nurse knows when to expect you. Your child's nurse can then give you an update on your child's progress. They can make sure that your child is available when you come. In most hospitals, parents can visit their child any time they want and stay overnight, so you can get reports from each shift nurse.

Your child may also see other specialists, like pain specialists, lung specialists, or kidney specialists. Who they see will depend on the reasons they are in the hospital. These people may visit your child throughout the day or may need to see your child only once. It is important that you talk with these specialists too.

It is important that you understand what the doctors have to say about your child's illness. It is important that you understand what medications and treatments are being given. Sometimes providers share a lot of complicated information at one time. If you do not understand, ask them

for more of an explanation until it is clear. You should feel empowered to ask questions about your child's medical care.

Hospital activities for your child

There are often special activities in the hospital that can help make your child's stay more pleasant. For young children, these include games and other preschool activities like playing with toys, doing crafts, and reading stories. The hospital may also sponsor special events such as magic shows, visits from the animal shelter, visits from athletes, and birthday parties. For older children, there are sometimes "teen rooms." They may have activities more appropriate for their age level.

If the hospital has a playroom or a teen room, you won't need to bring too many things for your child to have from home. This way your child won't lose a favorite toy. It might help your child to pick out a special "hospital toy" that is just to take to any hospital visits.

Dealing with behavior changes

In the hospital, you may notice some changes in the way your child behaves. They may not seem happy to see you when you visit or may cry a lot when you leave. You may notice them eating things in the hospital that they don't eat at home. Or, for younger children, they may need diapers in the hospital even though they are toilet-trained at home. It can be confusing for your child if you expect different behavior at the hospital than what you expect at home. You can be a little more patient with your child when they aren't feeling well, though.

Talk with the staff about any concerns you have about your child's behavior. It can be helpful for the staff to know how, within the hospital routine, they can be as consistent as possible with your child's home routines. Consistency between home and hospital can help a child return to their "usual self" much more quickly when they leave the hospital.

Talking about needs or concerns

Your child may feel fear or anxiety while in the hospital. If they are old enough to make requests or share their feelings, let them know that they can ask the staff for help or support. If your child is not able to talk to the staff themself, you should step in and let the staff know about these needs or fears. The medical team will do what they can to help your child.

You know your child better than anyone. By telling the staff about your child's needs and concerns, you will help your child get the best care.

Teens and self-advocacy

Pain is a common reason for teens to be admitted to the hospital. Your teen should have an individualized pain plan that is tailored for them specifically. Some teens who require frequent hospitalizations may have a plan that uses high doses of medication. There may be multiple reasons for this, including the development of their drug tolerance.

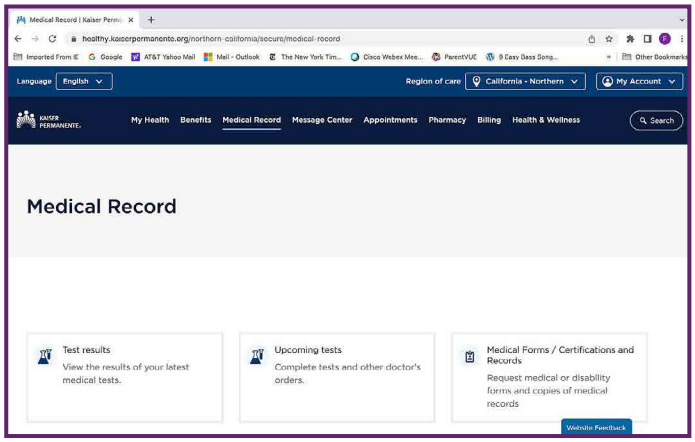
Your teen should know their own pain plan and have a copy of it, either printed or on their phone. While individualized pain plans improve patient treatment and satisfaction, problems may still happen. As teens age, parents should educate them to advocate for themselves using concrete actions that improve their care.

Unfortunately, teens and people of all ages with sickle cell disease are sometimes questioned about their reports of pain.

Children, teens, and adults with sickle cell disease pain can be mistakenly thought of as drug seekers by medical providers who are not very experienced with sickle cell disease. As a result, providers may not want to give the appropriate doses of pain medications needed.

Teens can be prepared for this situation. In addition to having access to their established individualized pain plan, your teen can ask the hospital to look up their pain plan from earlier visits to the hospital or ER. They should point out that their treatment information is in their electronic health record.

Teens should have the information needed to contact their primary medical providers. They should inform them about any problems and ask for their help. The main message is to be prepared by knowing the pain plan and having a copy of it. It may be helpful to always keep a secure copy at home to bring to the hospital if you are in a different hospital than your typical hospital. You or your child can show this information to medical providers to show the care your child received in the past.



An example of an online patient portal that can be used to contact a medical provider

Your teen's primary medical provider should also help and make sure the pain plan is followed. Whenever you or your teen feels not heard in the ER or in the hospital, you should reach out to their regular sickle cell disease provider so that person can call and help your teen get the appropriate care.

Adolescent and transition sickle cell disease programs should include education around these issues. They should help your teen develop the skills needed to communicate and access their support team and treatment plan. Using these methods will help your teen successfully advocate for their own care.

See Chapter 11, Bias, Racism, and Discrimination in Sickle Cell Treatment and Care.

Early aggressive treatment of severe pain

When your teen arrives in the ER or hospital in severe pain, they need immediate treatment.

You and your teen should know that there are no laboratory tests that can show the extent of your teen's sickle cell disease pain. Only your teen can say how much pain they are in.

Pain treatment should be based on the teen's report instead of how much they are moving around or their laboratory report. The reaction of pain varies among children. Some appear visibly hurt and others may be quiet or withdrawn. Pain medication should not be withheld before laboratory tests are done.

You can tell the providers that early and frequent treatment of pain not only improves the quality of life for your teen, but it also results in fewer hospital admissions in the ER and shorter hospital stays.

You may want to take a photo of the below section of this handbook and take it to the ER to help you advocate for your child's needs. Online version of this handbook (go.cpdh.ca.gov/SCParents):

- There are guidelines from the American Pain Society, the National Institutes of Health, and the American College of Emergency Physicians that say that for severe pain the wait time should be under 15 minutes. Some type of pain medicine should be given within 30 minutes.
- Opioid medications through an IV are first line medications to treat severe painful episodes in sickle cell disease. However, since there are often delays in getting an IV placed in the ER, many centers will give pain medications through the nose while waiting to get an IV placed. Pain medications that can be given through the nose include fentanyl and ketamine. Fentanyl is an opioid that works rapidly but its effects do not last long. It is very useful for initial pain management before there is an IV for IV opioids.

- Revisiting pain relief should be done about every 20 minutes. The next dose of medicine should come within 30 minutes if your teen remains in severe pain.
- Similarly, facilities should give antibiotics to a feverish teen with sickle cell disease within 30 minutes.
- About 30 minutes after treatment, the medical staff should reassess your child for fever, pain, or other symptoms. If there are major delays in your child's treatment for severe pain or fever, it is critical that you speak up. If your child does not receive care in a timely way, ask to speak to a supervisor or ER staff whose job it is to make sure patients get the care they need.

Medicines and IVs

There are common treatments that children of all ages with sickle cell disease require during hospital stays. These treatments are generally discussed below.

Medicine types

The most common types of medicines your child will receive in the hospital are:

- Antibiotics to fight infections
- Pain medications
- Medications to reduce fevers

Common Antibiotics

- Ceftriaxone
- Cefuroxime
- Penicillin
- Ampicillin

Pain Medications

- Morphine
- Dilaudid
- Codeine
- Hydrocodone
- Ketorolac
- Ketamine
- Ibuprofen (Advil or Motrin)
- Acetaminophen (Tylenol)

See Chapter 6, Pain and Pain Medications, for more information on these medications.

Medications for side effects of pain medications

- Stool softener – Colace, MiraLAX
- Diphenhydramine (Benadryl)

Blood products

- Red blood cell transfusions

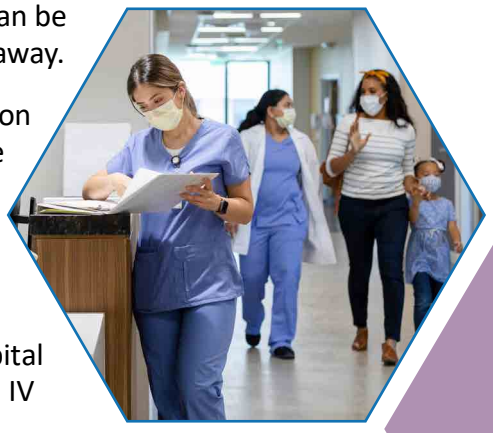
See Chapter 9, Blood Transfusions.

IVs

When your child is first admitted (checked in) to the hospital, they may be given fluids, medicine, or transfusions by an IV. "IV" stands for "intravenous," which means "into the vein." The IV will be left in the vein so that your child can get the fluids and medicine needed. It may take a few tries to get the IV in the right place. If your child is in the hospital for surgery, they will need an IV before, during, and after surgery to give them fluids and medicine.

The IV "pump" is on a stand next to the hospital bed. This little machine makes sure that the fluid and medicine go into the vein at the right speed. Occasionally, children who require frequent transfusions and hospitalizations may have a semi-permanent IV placed called a central IV line. This avoids many needle sticks, but it does have some risks you should review with your health team.

Surgery



Surgery can be elective or an emergency. Elective surgery means that it can be planned ahead. Emergency surgery means that it needs to be done right away.

If your child needs surgery, they will often receive a red blood cell transfusion to quickly reduce the amount of sickle hemoglobin in their body before the operation. This is true whether it is elective or an emergency surgery.

For elective surgery, your child will usually be admitted to the hospital the day before. This is so that lab tests and other preparations can be done. Prior to surgery, children must not eat or drink. This is called "nothing by mouth" or "NPO." One reason they are admitted to the hospital the night before surgery is so that they can receive hydration through an IV while they are NPO so that they do not get dehydrated.

Before surgery, it is important to make sure that asthma and other lung conditions are checked and are under control. The surgeon will talk with you about the surgery and possible problems that could happen. Other doctors or providers will explain anesthesia. This is when your child is put to sleep for the surgery. They will explain recovery time and any special care your child

will need after the operation. If you still have have questions about the surgery and anesthesia, make sure you speak with them before the operation.

People with sickle cell disease have a higher rate of pneumonia after surgery and should be alert, breathing normally, and able to walk before leaving the hospital. As soon as your child wakes up from surgery, it is important to work with them to take deep breaths frequently by blowing bubbles or using a device called an "incentive spirometer."

All surgery requires your consent if your child is under 18 years old. You must sign an "informed consent form" before any procedure can be done unless your child is 18 or older. In this case, your child will have to sign the consent.

Going home

When your child is ready to go home, a member of the medical team should talk with you about how your child is doing. They will share important issues to look out for at home. Try to allow time to speak to them when your child is getting ready to go home. This is also a good time to discuss any questions or concerns you have about caring for your child at home.

You may be given medicines for your child to take at home. Make sure that you understand how much and how often the medication should be taken. If you have questions when you are at home, contact the number on your follow up instructions. It is important that your child get the medicine as often and as long as the medical provider says it is needed.

Sometimes children don't return to their old patterns when they come home. They may have some of these problems:

- Trouble sleeping at night
- Wanting more attention
- Acting more unruly than before
- Wetting the bed or themselves when they had already stopped doing that at home
- Asking for a bottle when they didn't use one at home

Keep in mind that these problems can be managed. If the problems don't last long or are not too big, consistency in your approach before and after the hospital stay will help. Your child needs to have the same expectations as any child the same age who doesn't have sickle cell disease. To help return to normal, your child can be helped by a routine bedtime pattern, such as a bath, pajamas, and story at the same time each night.

If any of these problems last beyond a few weeks or are disrupting your household, talk about them. Contact your child's regular medical provider, social worker, or a member of the sickle cell disease team.

Finally, after a hospital or ER visit, if there is anything that you or your child want to share about your child's care, speak with the hospital's staff member responsible for improving medical care for patients. This person may be called an ombudsman. This person's job is to hear about what experience you and your child had in the hospital, what went right or wrong, and how it should be improved.

Chapter 9:
Blood Transfusions

Introduction

Children with sickle cell disease sometimes need blood transfusions. Transfusions mean blood from one person (called a donor) is put into another person. There are two major reasons why transfusions are given. The first is to decrease the number of sickle cells in the blood in order to prevent sickle cell disease complications. The second is to correct anemia and deliver more oxygen to the body.

There are many reasons why your child may need a transfusion. Your child might need a transfusion for a complication or for prevention of a complication. Transfusions will not help with all sickle cell disease complications. Transfusions are not generally done for the treatment of ongoing pain. Some of the reasons to give a transfusion include:

- Severe anemia. This is when red blood cell counts drop to a very low level. This anemia can be caused by:
  - Splenic sequestration – when the spleen enlarges and traps a lot of blood cells
  - Aplastic episode – when the body stops making new blood cells. Usually this is caused by a viral infection.
- Life-threatening problems, like a stroke or acute chest syndrome or severe pneumonia
- Prolonged, painful erection of the penis (priapism)
- Preparation for surgery
- An abnormal TCD

Until your child is 18 years old, the medical team will always try to tell you and get your consent before giving your child blood. Once your child turns 18 years old, the medical team will get consent directly from your child.

Types of transfusions

There are two types of transfusions your child may get. They are “simple” and “exchange” transfusions. Transfusions can be performed in the clinic or in the hospital, depending on the resources available.

Simple transfusions are the most common. Your child will get a set amount of donor blood through an IV.

An exchange transfusion is more complicated. It involves taking out your child's sickle red blood cells and replacing them with donor blood. The result is that the normal red blood cells replace sickled red blood cells.

An exchange transfusion can be the best and fastest way to increase the amount of blood flowing in your child's body and decrease the number of sickle cells. Your child may be put in the intensive care unit to be closely watched. This procedure can be done by nurses with syringes. It can also be done with a machine. When the blood exchange is done by a machine it is called "apheresis." Sometimes your child will need a new IV line or a special, big IV line that needs to be placed by surgery in order to have apheresis.

Each time your child is transfused, they will need blood tests ahead of time to match the blood that they are getting very closely with their own blood. Even though it requires an extra visit to a medical facility to get these tests before a transfusion, it is critically important to giving your child a safe transfusion and avoiding dangerous reactions.

Transfusion complications

Every effort will be made to give your child blood only when it is necessary because of possible complications. These complications are rare, but they can be serious. Possible complications include allergic reactions, allo-immunization, and infection.

Allergic reactions. Rashes, hives, and itching are common but not serious problems that can be treated. Rarely a person can develop fever or breathing problems.

Allo-immunization. Your child may develop antibodies that destroy the donor blood. Most people with sickle cell disease receive detailed matching of the donor blood with their blood to reduce the chance that this will happen.

Infection. All blood products are thoroughly screened for human immunodeficiency virus (HIV), hepatitis B virus, hepatitis C virus, human T-Lymphotrophic virus, West Nile virus, and syphilis. Blood from first-time blood donors is screened for even more infections.

It is very rare that infections are transmitted by transfusions.

Infection by the viruses that are screened for have less than a 1 in 2 to 3 million chance of happening. Infection by bacteria that may be in the blood is also rare but is more common than viral infections (less than 1 in 30,000 or 40,000).

See the Glossary for "Cross-matching Blood" and "Red Blood Cell Phenotyping" definitions.

Chronic transfusion

Some children need to have transfusions on a regular basis to stay well. If your child needs transfusions once a month, they are said to be on chronic or long-term transfusions. Your medical provider will not start such a program without discussing it with you first. You should understand why this is necessary. You will be asked to agree that this is a reasonable therapy before your child starts this treatment.

Inside red blood cells are deposits of iron. If a child is on chronic transfusions, they will get too much iron in their body. This is because the iron in the red blood cells they receive during the transfusions gets deposited in their body. This condition is called “iron overload.” It can lead to iron poisoning. It happens because the body does not have a way to get rid of the excess iron that it gets from red blood cell transfusions.

This extra iron may build up in their heart, liver, or kidneys and damage these organs. If your child is on chronic transfusions, your child will need to take a special medicine every day to get rid of the extra iron, called a “chelator.” This medication attaches to the extra iron in the body and removes it in the urine and stool.

Iron chelators

If your child is on chronic transfusions, they must take one or a combination of these chelator medicines. Jadenu, Exjade, and Ferriprox are all pills that need to be taken by mouth either once or twice a day. Jadenu also comes in “sprinkle” form, which is like little flakes that can be mixed with food.

An additional iron chelator called Desferal (deferoxamine) is not commonly used though it is an excellent and safe iron chelator. It is an infusion that is given through a needle placed just under the skin over an 8- to 10-hour period each day by an infusion pump. This pump slowly lets out the medicine through a needle under the skin. Most children take it at night when they are sleeping. Older children and adults may find it easier to take it during the day at school or work. You can give your child a choice about the timing.

As long as your child gets transfusions, they must take iron chelation. Even after the transfusions stop, it can take a while to get rid of all the iron. Your medical team will tell you when your child can stop.

Measurement of iron level

A test of the blood is used to measure iron in the body. It is called a ferritin test. The amount of iron in the liver can be measured by MRI of the liver. The test of ferritin concentration and the liver MRI are used to monitor iron levels. Medical providers use them to decide if iron chelation should be started, increased, or decreased. The liver iron MRI is done every 1 to 2 years.

Designated donors

Friends and families often want to donate blood. They can be donors if they don't have sickle cell trait or sickle cell disease. This is called being a "designated donor." Getting this blood donation is not covered by insurance and must be paid for by the family. They are not safer than volunteer blood donors at your blood bank. Generally, designated donors are not recommended. Having a designated donor may affect the success of a bone marrow transplant in the future.

Religious considerations

Some children and families with sickle cell disease are Jehovah's Witnesses. These families deserve respect and care from medical providers. The teachings of the religion talk about blood transfusions. We recommend talking to religious elders about this issue. Many hospitals have ways to address this issue in a manner that respects the religion. Even an informed and supportive medical provider may get a judge's order to allow a child to get a blood transfusion against the parents' advice. A medical provider would only do this when they fear the child's life is in danger.

Chapter 10:
Research and Treatment

Introduction

Over the last several decades, there has been an explosion of new treatments and new ideas for treatment of sickle cell disease. This chapter summarizes the drugs and treatments that are available. It also discusses some of the ongoing research to cure sickle cell disease. This information is current at the time of this revision in 2024. However, advancements are rapid, so check in with your sickle cell disease medical provider about updates and the status of therapies during your regular visits.

Fetal hemoglobin

The hemoglobin that all of us are born with does not sickle. It is called fetal hemoglobin. Fetal hemoglobin does not sickle even in babies with sickle cell disease. Many of the modern treatments for sickle cell disease aim to increase the amount of fetal hemoglobin in the body.

Hydroxyurea, discussed in more detail below, is the most common and the first Food and Drug Administration (FDA)-approved drug to prevent sickle cell disease complications. Its main action is to increase fetal hemoglobin.

Hydroxyurea has been used in sickle cell disease since 1995. It has been demonstrated to prevent or minimize sickle cell disease complications. Standard recommendations are to start it in all babies with sickle cell anemia (SS disease) and S beta zero thalassemia (SB^0) disease by 9 months old. Hydroxyurea's job is to prevent sickle cell disease complications from developing.

Overall, the use of hydroxyurea has been associated with cutting the risk of hospitalizations, pain, and other complications in half. These complications include the need for blood transfusions. Research on hydroxyurea seems to show that people who take it for a long time have improved brain functioning.

All people, including those with sickle cell disease, still have the ability to make fetal hemoglobin. Bodies naturally turn off this function after birth, though. For decades, researchers have tried to understand how we can get children and adults who usually don't make fetal hemoglobin to start making fetal hemoglobin again.

Researchers have learned more about how the body turns off the ability to make fetal hemoglobin.

There are genes (or instructions) in the body that block fetal hemoglobin production. These genes are called “modifier genes.” The most important “modifier” of fetal hemoglobin production is a protein called “BCL11A.” This protein is what shuts off fetal hemoglobin production once someone is born.

Exciting new sickle cell disease research shows that new treatments can block “BCL11A” so that the body can produce more fetal hemoglobin. These new treatments are called “gene therapy.” Research studies called clinical trials are also testing medications (aside from hydroxyurea) that increase fetal hemoglobin.

Hydroxyurea, and three more medicines that help people with sickle cell disease without increasing fetal hemoglobin, are discussed next in more detail.

Available drugs

In addition to pencillin, there are four prescription drugs available to help patients with sickle cell disease, as of 2024. More drugs are on the way in clinical trials. The available drugs in the United States are hydroxyurea, L-glutamine, voxelotor, and crizanlizumab. Below is a summary of the drugs. Parents of children with sickle cell disease should talk to their child’s medical provider about the best drug or drugs for their child.

1. Hydroxyurea (HU) – liquid or pill, taken once a day

How does it help people with sickle cell disease?

People with sickle cell disease who take hydroxyurea live longer lives and have half as many hospitalizations, pain episodes, and lung problems as those who don’t take hydroxyurea. In addition to reducing sickle cell disease complications, hydroxyurea also improves anemia. In other words, it makes hemoglobin levels higher. It does this by increasing the amount of fetal hemoglobin.

How long has hydroxyurea been around?

Hydroxyurea has been used to treat sickle cell disease since 1995.

Should my child be on hydroxyurea? What age should my child start?

Hydroxyurea is so effective and safe that it is recommended for all children with sickle cell anemia (SS disease) and S beta zero thalassemia (SB^0) disease. Hydroxyurea is recommended for many other people with sickle cell disease if they have severe symptoms of the disease, such as frequent pain. They include people with SC disease and other types.

Once children reach 9 months old, parents and providers should discuss starting hydroxyurea. The provider should explain the required regular lab tests and describe the drug’s potential side effects.

Sometimes families with children who have sickle cell disease are scared to use hydroxyurea because they have heard that it has been used for chemotherapy in cancer. It is one of many medications that are used for both cancer and other diseases. The purpose of hydroxyurea in sickle cell disease is different from the purpose in cancer treatment. Hydroxyurea is very safe and very effective for sickle cell disease.

What are the side effects and monitoring guidelines?

The most common side effect of hydroxyurea is a drop in the number of white blood cells. You need white blood cells to fight infections. Infections include colds, pneumonia, and skin infections. However, people with sickle cell disease have too many white cells. This is one of the factors that may worsen sickle cell disease symptoms. Lowering the white blood cell count to a safe level is helpful. However, sometimes the white cell count may drop too low. This is completely reversable by adjusting the hydroxyurea dose. In order to stay in a safe range, regularly scheduled blood tests are needed.

At the start, the lab monitoring can be as frequent as every few weeks. Once a child has been on the medicine for several months, it can usually be spaced to every 3 months or so. The actual frequency of lab tests will be decided by your doctor. There is no set rule.

One other possible side effect of hydroxyurea is stomach upset, but this is generally mild. There are other minor side effects that do not happen in most people. Hair thinning and nail changes can happen, though. Usually if the side effects from hydroxyurea are causing your child problems, your doctor will lower the dose. For people whose symptoms are not well controlled with hydroxyurea, one of the other three approved medications can be safely combined with hydroxyurea.

2. L-Glutamine or Endari – powder that is mixed with a drink or soft food, taken twice a day

How does L-Glutamine help people with sickle cell disease?

L-Glutamine is another treatment option. The brand name of the prescription drug is Endari. Endari reduces the number of annual pain episodes from 3 to 2. It reduces hospitalizations and may reduce the number of acute chest syndrome problems in people with sickle cell disease.

Endari works by acting as a natural antioxidant in red blood cells. An antioxidant can sop up chemicals that damage red blood cells. The benefits of Endari are more significant in people also taking hydroxyurea.

How long has L-Glutamine been around?

As a medicine for people with sickle cell disease, L-Glutamine or Endari has only been available since 2017. However, glutamine is a natural amino acid that is commonly found in food. An amino acid is a component of protein. It is available without a prescription as a health supplement, but there is variability in the quality and purity of the over-the-counter products.

As a medicine for people with sickle cell disease, it is relatively new, so there are no data on long-term benefits. Glutamine has a history of being safely used for other medical conditions, though.

Should my child be on L-Glutamine? What age should my child start?

If your child is taking hydroxyurea and still has pain that is not well controlled, you may want to consider adding Endari. It is approved for children of 5 years and older.

What are the side effects and monitoring?

Constipation (hard poops) and stomach upset are the most common side effects. There is no lab monitoring required.

3. Voxelotor (Oxbryta) – tablet that can be swallowed or crushed and is taken once daily

How does Voxelotor help people with sickle cell disease?

Voxelotor protects red blood cell health. It raises hemoglobin levels by decreasing red blood cell destruction and preventing jaundice. Long-standing anemia is associated with many long-term problems. Taking voxelotor to prevent anemia may protect the brain, kidneys, and other parts of the body from harm. It can also slightly cut down on pain episodes.

How long has Voxelotor been around?

Voxelotor has been available for people with sickle cell disease since 2019.

Should my child be on Voxelotor? What age should my child start?

Voxelotor is approved for children who are at least 4 years old. Your doctor may recommend the medication to raise your child's hemoglobin or to decrease red blood cell destruction. It may have a beneficial effect when given with hydroxyurea.

What are the side effects and monitoring guidelines?

The most common side effects are upset stomach and diarrhea. Standard monitoring of the hemoglobin is needed.

4. Crizanlizumab (Adakveo) – 30-minute IV infusion given initially 2 weeks apart, then monthly

How does Crizanlizumab help people with sickle cell disease?

Sickle cells are sticky. Sickle cells, white blood cells, and platelets often attach to the inside of blood vessels leading to decreased blood flow and blockage of blood flow.

Blockage and reduced flow cause pain and injury. Drugs that reduce the stickiness of cells are helpful in the treatment of sickle cell disease. Crizanlizumab works by blocking sickle cells from sticking in blood vessels.

How long has Crizanlizumab been around?

Crizanlizumab has been available for people with sickle cell disease since 2019.

Should my child be on Crizanlizumab? What age should my child start?

Crizanlizumab is available for people who are at least 16 years old. It is a particularly good option for people who have trouble taking medicine at home since it is a medicine given by IV in the clinic every month.

What are the side effects and monitoring guidelines?

Muscle aches are the most common side effect of this medication. Most people who experience these aches only do so with the first few infusions.

Table: Drug treatments

Table includes approved ages, benefits, side effects, how drugs are administered and who they could benefit.

Drug	Approved for ages	Main benefit	Major side effects	Administration	Good for?
Hydroxyurea (HU)	9 months or older	Less anemia, less pain, less hospitalizations, less acute chest syndrome	Low white blood cell numbers (mild and rare)	Oral liquid or capsule daily	Everyone
L-Glutamine (Endari)	5 years or older	Less pain	Nausea, abdominal discomfort (mild)	Oral powder twice daily	People who have either persistent pain on HU or would like complimentary or natural therapy
Voxelotor (Oxbryta)	4 years or older	Healthier red blood cells and less anemia	Headache, abdominal discomfort (mild)	Oral tablet daily	Very anemic people or those who can't tolerate HU
Crizanlizumab (Adakveo)	16 years or older	Less pain	Back pain, muscle aches, nausea (mild), infusion site reaction	IV infusion. The first two doses are 2 weeks apart, then it is given every 4 weeks	Those who can't tolerate HU or it does not work for them. Those who have trouble taking medicine at home

Clinical trials

There are more drugs being tested for sickle cell disease in clinical trials. Clinical trials are research studies involving people. They are the primary way that researchers find out if a new drug, diet, medical device, or other new treatment is safe and effective. These trials are required to protect the rights, interests, and safety of the people who participate in them.

Participating in clinical trials is a way to receive the newest treatment. It is a way to have additional care and attention from clinical trial staff. It could also help researchers find better treatments for sickle cell disease. National Library of Medicine (https://clinicaltrials.gov) is one place to find out about clinical trials to join.

See the online Appendix for more links to information about clinical trials.

Therapies to cure sickle cell disease

There are currently two ways to cure sickle cell disease. First, there is stem cell transplantation. This is also called bone marrow transplantation. Stem cell transplantation has been successfully used to cure sickle cell disease since the 1990s.

Gene therapy is the second cure. The FDA approved some gene therapy treatments in 2023.

Transplanting stem cells

Stem cell transplantation (which is also called bone marrow transplantation) is sometimes recommended for people with sickle cell disease. It is especially recommended for those who have frequent pain, acute chest syndrome, stroke, and other bad complications.

In this type of transplant, the blood stem cells of a person with sickle cell disease are replaced by the blood stem cells of another person, called a donor. This works the best when the donor is a sibling with very similar genes. Most children survive and are cured permanently. Overall about 10% die or have the sickle cells come back. There are multiple factors that affect the success of each transplant.

If a person with sickle cell disease does not have a sibling who is a genetic match, it is still possible for them to receive a transplant from an unrelated donor who is a genetic match. At the time of publishing this book in 2024, transplants with donors who are not matched siblings are usually done through clinical trials.

If a family is interested in stem cell transplantation, they should request to meet with a stem cell transplant specialist who has experience in sickle cell disease and access to clinical trials. At the time of the first meeting with the transplant team, it is good to get what is called human leukocyte antigen or "HLA" matching. HLA matching finds out if there is a genetic match between the person with sickle cell disease and their sibling. This testing also will provide the sample to look for a genetic match that is not related to the person. Information on genetic matching with non-family members exists in a worldwide database.

During this meeting, the family will also learn the risks of the stem cell transplantation. The risks can include long hospitalization, serious infection, and graft versus host disease (GVHD). GVHD is a complication in which the donor cells start to attack parts of the body in the person with sickle cell disease. The closer the genetic match, the less chance there is of GVHD.

A successful bone marrow transplant requires strong chemotherapy to eliminate sickle blood stem cells. The chemotherapy clears out the sickle cells and makes space for new healthy stem cells to grow. Additional "immune" therapy prevents the body from rejecting the new stem cells. It can take over a month before the healthy stem cells grow enough to produce normal cells.

Transplantation takes several months but, if successful, the person with sickle cell disease will be cured of sickle cell disease for their whole life. In addition to 3 to 4 weeks in the hospital for the transplant, there is a lot of preparation and follow-up time.

The overall success rate of stem cell transplantation in sickle cell disease is always improving. The success rate and complications are based on the person's risk factors, such as age and previous medical complications.

In a review of over 1,000 transplant cases from multiple sites, 9 out of 10 people survived and did not have sickle cell disease. Although the death rate from transplant varies, families should consider that five out of 100 people could die from the transplantation. There could also be other problems aside from death.

People with sickle cell disease who get a transplant could have a dramatic improvement in pain, lung disease, and stroke. However, the majority of people lose their ability to have children. Because of this, people will be given an option to save and store sperm or eggs before transplantation. In addition, people may develop hormone problems after the transplantation. As a result, they may need to take hormones to keep them healthy and growing.

Alternative options for families without a sibling donor match

Several options are available if there is not a sibling who is a good genetic match. These options are best explored as part of an ongoing clinical trial by an experienced sickle cell disease team.

The most common alternative transplant involves a fully genetically matched unrelated donor. Other options include a stored umbilical cord sample or a half-matched relative (also called "haplocompatible donor").

Gene therapy

Gene therapy is very similar to a stem cell transplant. It is a stem cell transplant in which the person's own stem cells are removed from the body. Stem cells are removed by a large surgically placed intravenous line. The cells are then modified in a lab to produce non-sickle blood stem cells. They are then returned to the person who has sickle cell disease. Since the transplantation uses the person's own stem cells, people who get gene therapy are not at risk for graft versus host disease (GVHD) because GVHD is only associated with standard transplants.

Gene therapy is an exciting treatment. Until 2023, only people in clinical trials could try gene therapy. That year, the FDA approved gene therapies called Casgevy and Lyfgenia. The clinical trials done on these gene therapies demonstrated that gene therapy is generally safe and effective. However, the treatment is so new that all the long-term effects are not known yet.

The gene therapy clinical trials have already had two people with sickle cell disease who received gene therapy and developed blood cancers. The cancer was not considered related to the gene therapy, but nobody knows for sure whether it was related. As of 2024, this issue continues to be intensely investigated.

Types of gene therapy

As of 2024, there are two types of gene therapy being tested in clinical trials or available for use:

1. Gene addition therapy. For this type, a healthy anti-sickling gene is added to the stem cells of a person with sickle cell disease to block sickling.
2. Gene editing therapy. For this type, a person's genes are modified to produce more healthy hemoglobin.

The FDA has approved Casgevy, a type of gene editing therapy that uses CRISPR technology. CRISPR changes a piece of DNA to cure sickle cell disease.

Using CRISPR is expensive. There are no long-term results to show patient safety over the long-term, yet. CRISPR has cured a growing number of people of sickle cell disease, however. Talk to your child's medical provider to find out if this treatment is an option for your child if you are interested.

Chapter 11:
Bias, Racism, and Discrimination in Sickle Cell Treatment and Care

Introduction

People of all races and backgrounds can be born with sickle cell disease. In California, sickle cell disease is most commonly found in Black and Latino people. As with any health issue affecting communities of color, people with sickle cell disease may experience bias, racism, and discrimination in medical care.

There can also be a lack of awareness and education about sickle cell disease among medical providers. For example, some medical providers ask, “How long have you had sickle cell disease?” without knowing that the disease is a life-long condition passed down to children from their parents. You may find that you may have to repeat information on your child’s condition and be more forthcoming with your expectations for care.

How can bias affect the care your child receives?

Most medical providers want to deliver the best care for your child. But everyone has some unconscious bias, even at work. This means an attitude they might not even know they have that affects the way they think about and treat others around them.

This bias can present as racism and discrimination. Bias and lack of awareness in medical systems can affect whether your child can access medical care and services. It can influence the quality of care received. It can affect your child’s treatment and health. It can also have an effect on the mental health of you and your child.

You or your child may not feel heard while seeking medical care or pain medication. This is not how your medical care experience should feel. When people with sickle cell disease are discriminated against by their medical providers or system, it may increase their stress and cause them to avoid seeking care or help when they need it. All of this can cause people living with sickle cell disease, including your child, to have poorer health.

More people and organizations are realizing that this needs to change. The former head of the U.S. Centers for Disease Control (the CDC), for example, asked both medical providers and health

systems to make changes. Changes include using non-harming language and adding more anti-bias training. These and other changes are designed to reduce health inequalities and address racism when caring for patients living with sickle cell disease.

The California Department of Public Health (CDPH) similarly champions anti-racism changes in medical care. CDPH helps ensure hospitals know they are required to have implicit bias training. CDPH helps ensure hospitals have resources and tools for medical staff to deliver fair, safe, and high-quality care to California's diverse communities.

Since some birthing parents and their infants can face unique challenges, CDPH offers families supportive programs such as the Black Infant Health; California Home Visiting; Women, Infants, & Children (WIC); and Perinatal Equity Initiative programs. These programs are part of WIC or the CDPH Maternal, Child, and Adolescent Health (MCAH) Division. You can do an online search for "CDPH" and "Center for Family Health" to find these programs.

If you or your family believe you're not getting the care your child needs and deserves, it is your right to ask providers for better care for your child.

Even though it shouldn't be this way, your family may need to advocate strongly for better medical care in the clinic, hospital, or emergency room to make sure your family gets the care they deserve.

If you feel that you did not receive the care that your child needed after a hospital or emergency room visit, discussing the experience you had with administrators at the hospital or emergency room gives you a voice in the care of your child. It may prevent the same experience for your child and other children with sickle cell disease in the future.

What can you do?

Here are a few things that may help you and your child work with the medical system:

- Stay in touch with your child's usual sickle cell disease doctor if you can. It is your right to bring up issues of discrimination and racism in your child's medical care. You can also bring it up with other members of your child's comprehensive care team.
- During your regular outpatient visits, make it a priority to talk to your providers about your family's experience with your child not receiving the care they deserve. Ask your doctor to directly address the person or medical facility where they faced discrimination. Your doctor backing you up and helping with these issues will likely help with both your child's treatment and your family's experience in the future.
- It is important to work with your sickle cell disease doctor to create an individualized pain plan during your regularly scheduled visit. This plan should provide the guidelines

for your child's hospital and emergency room visits, no matter where they get care. Taking a photo of it and having it in your phone or your child's phone may be helpful. Many patients may need treatment at facilities they don't regularly use. Having this information available for these medical providers should help inform them of the care you expect and deserve.

- If providers do not follow your child's individualized pain plan, make sure the medical staff makes a note in the medical record when pain medication or treatment is refused by the provider. That way there is a record you can go back to if you decide to report what happened.
- It is your right to report when medical care has not been high-quality or takes too long. Many hospitals, offices, and clinics have a staff member responsible for improving medical care for patients. This person may be called an ombudsman. This person's job is to hear about the experience you and your child had with medical care, what went right or wrong, and how it could be improved in the future. In addition to filing a complaint with your medical provider, you can also file a complaint with your health plan or insurance provider. Ask your ombudsman or your health plan member services representative how to file a complaint.
- You can also file a complaint with the state if your rights were denied. To file a complaint with CDPH, please do an online search for "CDPH" and "Cal Health Find Database" (https://www.cdph.ca.gov/Programs/CHCQ/LCP/CalHealthFind/pages/home.aspx).
- You can also contact your local CDPH district office. A list of those offices can be found in an online search for "CDPH" and "CHCQ Field Operations District Offices" (https://www.cdph.ca.gov/Programs/CHCQ/LCP/Pages/DistrictOffices.aspx).
- Once CDPH receives your complaint, the licensing teams conduct a full investigation and ensure all applicable laws were properly followed. If they weren't, the hospital could face citations or heavier penalties.
- You can also file a complaint with the federal government if your rights were denied. To find the Complaint Portal Assistant, do an online search for "US" "HHS" and "Complaint Portal Assistant" (https://ocrportal.hhs.gov/ocr/smartscreen/main.jsf).
- If you need help with your health insurance plan, have concerns, or want to file a complaint about your health insurance plan, you can submit an online complaint with the California Department of Managed Health Care (https://www.dmhc.ca.gov/). Do an online search for this agency name for more information.

See the online Appendix for additional resources on racism and sickle cell disease. It includes links to community groups that support families and websites with more information on standing up for your rights.

Chapter 12:
Sex and Reproduction

Introduction

In the teenage years and through adulthood, people face choices about sex and having children. It is not always easy to know what is right to do. This is true for both children and their parents.

People with sickle cell disease face the same challenges as other people. Teenagers and young adults with sickle cell disease need to decide if and when they want to have sex and whether they need birth control. Sometimes young people with sickle cell disease have concerns about whether they can have children, if their disease will affect their dating relationships, and what kind of birth control is right for them.

Young adults with sickle cell disease may not feel comfortable discussing these issues with their family or medical provider. Creating an environment that encourages communication around these issues is important. It will enable the young adult to make informed choices. It will help them address their fears and concerns.

Young people can decide if they want to have children. It is very important that they understand the chances of their baby having sickle cell disease. The chances of any child having sickle cell disease is dependent on the genes passed down from both the mother and father. (Genes are in our cells. They are the blueprint for our bodies.)

There are reproductive options that a person with sickle cell disease and their partner should know about. Learning more may improve the health of the pregnant person and their pregnancy. People with sickle cell disease may want to speak with a genetic counselor, get hemoglobin testing, and make sure they have access to comprehensive care before having children.

The choices a young adult with sickle cell disease needs to think about are discussed in this chapter. Learning about these choices will enable young adults to make the best decisions for their future.

Parents can help their children face important questions. You can talk to your children so that they know the facts. You can also share your feelings and values to help guide their choices. This chapter will help you talk to your child with sickle cell disease about these issues. It will also inform you about the impact of the disease on these areas of your child's life.

This chapter will cover:

- Sex and teens
- Birth control basics
- Sexually transmitted infections (STIs)
- Pregnancy

Sex and teens

Like all teens, teens with sickle cell disease face challenges with sex. As they reach puberty, their bodies go through many changes. They begin to have strong sexual feelings. Some are able to get pregnant. Others are able to make someone pregnant. All teens have choices to make that will have a major impact on their lives.

Questions for teens to answer

Here are some of the questions that teens need to answer for themselves:

- How will they respond to their sexual feelings?
- When will they have sex? With whom? Will they wait to have sex until they are married?
- Do they want to have children?
- If they have sex but don't want children now, what kind of birth control will they use?
- How will they protect themselves from the HIV and other STIs (sexually transmitted infections) like chlamydia or genital warts?

These are not easy questions. Teens need to give these questions a lot of thought. They need to make good choices for themselves and their future families.

Concerns with sickle cell disease

Teens with sickle cell disease may have concerns about maturing late. Biological males may wonder whether they can have sex if they have not reached puberty or if they have a problem with priapism. Some teens may worry about whether they will be able to have children.

These concerns can add to usual fears about dating. Teens with sickle cell disease often worry about whether they will find a romantic partner. Having friendships and a good time with other teens of both sexes that they are not dating may ease some of these worries.

Teens should know that there is a high likelihood that they can have sex and get pregnant like other teens without sickle cell disease. Some people with sickle cell disease may have complications during pregnancy. Early monitoring by gynecologist or obstetrician sickle cell disease specialists will minimize or prevent these problem.

Talk to your teen about sex

You don't have complete control over what your teen does about sex, but you can talk to them. You can give them the facts and help them make sense of all that they have heard. You can share your feelings and values so that they know where you stand. You can listen to their feelings and thoughts and help them decide what is best.

Don't wait until your child becomes a teen to start talking about sex. Your child may ask you questions about these issues. Answer what they ask. You don't have to explain everything at once.

If your child doesn't ask you about sex, bring up the subject. Look for times to ask about their thoughts or feelings and to share yours. It could be while watching TV or when you hear or read something about sex or love.

Don't assume your teen knows it all even if they act like they do. Research shows that teens who know the least about sex are more likely to have sex sooner. More often than not, it is what teens don't know about sex that can hurt them.

Birth control basics

Teens who have reached puberty can get pregnant or can get someone pregnant. If they choose to have sex and don't want children, they need to use birth control. Birth control is also called "contraception." There are many kinds of birth control available.

Condoms and foam

One of the best forms of birth control for people with sickle cell disease is the condom. Condoms are good because they also help protect a person from STIs, including HIV and syphilis.

Condoms don't always keep someone from getting pregnant. It is important to use them according to instructions. For more protection, many people also use a special foam with a condom. This foam is put in the vagina before sex. It kills sperm that the condom doesn't stop.

Birth control pills

Birth control pills can be used by some people with sickle cell disease. People with sickle cell disease are at increased risk for clots and stroke. High-estrogen-containing birth control pills

increase the risk of clots and stroke. People with sickle cell disease who have a history of clots, stroke, severe migraines, high blood pressure, or kidney failure should not take estrogen-containing birth control pills.

Some providers believe estrogen-containing birth control pills are not safe for people with sickle cell disease. However, the World Health Organization (WHO) and the CDC do not agree. They say that low-dose, estrogen-containing or progestin-only birth control pills are an acceptable option for people with sickle cell disease. They say the benefits outweigh the risks. Your teen should discuss these risks with their medical provider.

Birth control pills are very effective in preventing pregnancy if taken every day. Remember that pills don't give protection for HIV and STIs.

Depo-Provera (DMPA)

Depo-Provera is a shot given every 12 weeks. The hormone in it keeps women from getting pregnant. It does not prevent STIs. Biological females should ask their doctor about whether it would be a good method for them. People with sickle cell disease using Depo-Provera should have bone density studies and may need calcium supplements. It can also cause weight gain with long-term use. This can lead some people to switch to a different method of contraception.

Intra-uterine devices (IUDs)

Intra-uterine devices or IUDs are small devices inserted into the uterus. They are an acceptable method for people with sickle cell disease. These devices are safe to leave in for years and are effective forms of birth control. They do not protect against STIs. For people with sickle cell disease, the progestin-containing IUD, which can stay in for 5 years, is preferred over the copper IUD, which can stay in for 10 years.

Choosing a birth control method

In addition to the other methods discussed above, people can also use a diaphragm, cervical cap, vaginal ring, implant, or other newer methods as they become available. Your teen should talk with their partner and their medical team about what method of birth control to use. Then they can choose what will work best.

Sexually transmitted infections (STIs)

Sexually transmitted infections (STIs) are a group of diseases that are spread by having sex. They include diseases like genital warts, chlamydia, gonorrhea, HIV, and syphilis.

Signs of STIs

Most STIs can be cured if they are treated early. If your teen is having sex, they should watch for these signs. If they have any of these symptoms, they should contact the doctor right away.

In biological males:

- Drip from the penis
- Pain or burning when passing urine (peeing)
- Sores, rashes, or growths on or near the genitals

In biological females:

- Strange discharge from the vagina
- Pain in the lower abdomen and fever
- Sores, rashes, or growths on or near the genitals

Some people never develop symptoms even though they may have an STI. Because of this, it is very important that sexually active biological females visit a gynecologist every year. A gynecologist is a reproductive health doctor. Early treatment can prevent some of the more harmful effects of these diseases.

Your teen should be checked by the doctor right away if they are worried about any of these signs. STIs can make them very sick or unable to have children if they are not treated.

Talk to your teen about STIs. Explain what they are and how they are spread. If they don't have sex, they won't get an STI. If they choose to have sex, condoms will help protect them and their partner.

HIV

Like other STIs, HIV can also be spread through sex. Once you have the HIV virus, you can't get rid of it. Over time, HIV can cause people to get very sick. Some people get sick quickly. Other people stay well for years.

Most people who are diagnosed early and take medicines for HIV can live long, healthy lives. But there is no cure for HIV infection. The best way to deal with HIV is to protect yourself from infection. The main way that people get infected with HIV is by having sex with someone who has it. Some people also get HIV by sharing needles through IV drug use.

Talk to your teen about how they can protect themself from this very serious disease. If they have sex or use IV drugs, they are at risk of getting infected. Go over these guidelines with them:

- The only sure way to protect yourself from the virus is not to have sex or use IV drugs.
- If you have sex, condoms will help protect you from the virus. Also use foam or cream with Nonoxynol-9 to help protect you from the virus.

Pregnancy

Some people with sickle cell disease choose not to have children. Others choose to have children or adopt. The choice is up to each person.

Planning a pregnancy

Planned pregnancies are very important with sickle cell disease. Without planning and special medical care, pregnancy can be dangerous for the pregnant person and the baby. If your teen is going to have sex and they aren't ready for children, they need to use birth control.

If your teen wants to have a baby, it is important that they understand the baby's risk of having sickle cell disease and how that might affect their decision.

Genetic counseling and hemoglobin testing for your teen and their partner before pregnancy can show the couple's chances of having a baby with sickle cell disease.

Even though someone has sickle cell disease, it does not automatically mean that their baby will have sickle cell disease. Pre-pregnancy counseling and testing will tell them the chances of this happening. The information in this section also applies to you (the parent) if you want to have more children.

If a couple has an increased chance of having a baby with sickle cell disease, there are ways to make sure the baby is born without sickle cell disease. The couple can choose a biological way to get pregnant called "in vitro fertilization" or "IVF." IVF involves retrieving eggs from the female and fertilizing them with sperm in a laboratory. The eggs are tested and then can be implanted in the female's body. There may be other options that a genetic counselor can discuss as well.

Genetic counseling and hemoglobin testing

Whether you (the parent) or your teen are planning a pregnancy, we recommend finding out all you can about it.

You and your partner are able to get genetic counseling if having more children is in your future. If your teen wants to have children, or is pregnant or has fathered a child, your teen should also get genetic counseling.

Many people do not know it is their right to talk to a genetic counselor or medical provider when planning a pregnancy. If it is not offered, you can ask for it. A genetic counselor is a person who

is trained to help you, your teen, or other members of your family understand your chances of passing sickle cell disease or sickle cell trait to a baby.

In addition to getting genetic counseling, a couple wanting to become parents should have hemoglobin testing before a pregnancy. This lets a couple know before they are pregnant if they might have a baby with sickle cell disease or a different hemoglobin disease. It is important for both people to get tested. Hemoglobin testing can also be done as soon as a pregnancy begins. Talk to a genetic counselor and your medical provider to discuss your particular situation.

See Chapter 13, How Sickle Cell Disease Is Inherited, for information on hemoglobin traits and disease.

Ending unwanted pregnancies

When someone gets pregnant, it is their choice whether to have the baby or not. Some people may want to end a pregnancy for a variety of reasons. In 2024, in California (and about half of the other states), people can choose to end an unwanted pregnancy by getting an abortion. Do an online search for "California Surgeon General" and "Let's Talk Reproductive Health" for more information (https://osg.ca.gov/reprohealth/).

Pregnancy and health

People with sickle cell disease may wonder if they can get pregnant and have healthy children. There is a high likelihood they can. But if they choose to have the baby, they need early care to prevent or lessen problems.

While a person with sickle cell disease can birth a healthy baby, there are risks. Both they and the baby need to be watched closely. It takes planning and work to have the best chance for a healthy pregnancy and baby.

For people who decide to continue a pregnancy, early prenatal care helps the baby stay healthy. Early care can lower the risk of miscarriage. It can also decrease the risk of having a baby who is too small. Early prenatal care also helps the pregnant person. They need to be monitored so that any problems can be found and treated early.

It is best to start prenatal care before getting pregnant or right after. The obstetrician and sickle cell disease medical provider can work together to help keep the parent and their baby healthy. For all people with sickle cell disease, prenatal care should be done by an obstetrician who is an expert in high-risk pregnancy, especially pregnancies complicated by sickle cell disease.

A person with sickle cell disease needs to plan ahead when pregnant. From the start of the pregnancy, they have to be careful about what they do.

They may have to live with more pain while they are pregnant or go without treatment for certain problems.

Alcohol, certain medicines, and other drugs can all harm their baby. This can be hard for people who rely on certain drugs to help them manage their disease.

A pregnant person needs to check with the medical provider before taking any medicine. Their provider will tell them what medicines might cause problems.

Some pregnant people may need to receive blood transfusions near the last 3 months of pregnancy. This decision is best made with both the obstetrician and the sickle disease cell doctor.

Testing the fetus for sickle cell disease

Many couples want to find out before birth if their fetus has sickle cell disease. The unborn baby can be tested for sickle cell disease in the early months of pregnancy. These tests include amniocentesis or chorionic villus sampling.

If tests show that the baby has sickle cell disease, the couple can make choices. Some people use the time to prepare for their baby's special needs. Others choose not to continue the pregnancy. It is up to the couple to decide what is right for them. The medical staff will support their decision.

Cord blood banking

Parents of a child with sickle cell disease who plan to have more children may want to find out about the option to save the new baby's cord blood. This is blood in the placenta. If the new baby does not have sickle cell disease, their cord blood might be usable to cure the child with sickle cell disease.

The process of curing sickle cell disease from cord blood requires special tests and a stem cell transplant. A number of both public and private cord blood banks offer free storage to families with medical needs. Talk to your medical provider about how to find out more about cord blood banking.

See Chapter 10, Research and Treatments, for more information on curing sickle cell disease.

Teen pregnancy

Teens who get pregnant have more problems than older people who get pregnant. Pregnancy is stressful. Since teens are still growing themselves, pregnancy places an added strain on their bodies. Teens with sickle cell disease have increased chances for even more pregnancy problems.

Some teens with sickle cell disease try to get pregnant to prove that they can. Talk to your teen about the risks before they do something that may not be right for them. Reassure them there is a high likelihood that they will be able to get pregnant when they plan it.

Make sure your teenager knows that they are fertile. Tell them that they do not have to prove that they are fertile now. If they have sex and don't want a pregnancy, they need to use birth control.

If your teen gets pregnant or gets someone else pregnant, they will need your support. If your child doesn't want the baby or didn't plan to have children, your teen may be upset or scared. Talk to your child. Listen to your child's feelings and let them know that you will help. Share your feelings in a way that doesn't turn your child away. It won't help if you stop talking to each other.

Pregnant teens have some hard choices to make. Some teens choose not to continue the pregnancy. Others choose to have their babies. They may raise them or give them up for adoption. No matter what their choice is, teens with sickle cell disease need to see both an obstetrician and their sickle cell disease medical provider if they get pregnant. An obstetrician is a doctor who delivers babies.

Where to get help for pregnant teens

- **Obstetrician.** It is helpful to find one who is an expert in high-risk pregnancies.
- **Sickle cell disease genetic counselor and social worker.** They will provide you with supportive guidance throughout the process.
- **Local Health Department.** They may have a public health nurse or social worker who helps teens get the care they need. They may also have a prenatal clinic or classes for teens.
- **Family Planning Clinics.** These clinics offer low-cost pregnancy tests and family planning services. Some of them also provide prenatal care.
- **Community Health Centers.** These clinics often offer low-cost pregnancy tests and prenatal care.
- **Medicaid (Medi-Cal).** This government program pays most or all prenatal care costs for low-income women.
- **Women, Infant and Children (WIC).** This program provides free food and formula to low-income people who are pregnant or parents of young children.

Chapter 13:
How Sickle Cell Disease Is Inherited

Introduction

Sickle cell disease is a genetic condition that is present at birth. The term “genetic” is related to how genes and traits are passed down from parents to children. People can be born with a genetic condition because of a change in a gene. A gene is a written instruction or a blueprint that the body uses to make proteins. A body needs proteins to develop and grow.

A genetic condition happens when there is a change in these instructions. For example, a change in the gene for hemoglobin causes sickle cell disease. Hemoglobin is a protein that carries oxygen in red blood cells. This chapter answers common questions about sickle cell disease genetics and history.

What causes sickle cell disease?

Sickle cell disease is an inherited disease, passed from parents to their children through their genes.

Every person has two copies of a gene for hemoglobin, one from each parent.

Each parent has two copies of a gene for hemoglobin. Each parent passes only one of these copies on to a child, so that child ends up with one copy of a hemoglobin gene from each parent.

How is the disease passed from parents to children?

To inherit sickle cell disease, a child must get the sickle cell gene from one parent and a gene that is not for normal hemoglobin A, from the other parent. If a baby inherits a normal hemoglobin A gene from either parent, the baby will not have sickle cell disease.

Random chance decides which copy of the hemoglobin gene from each parent is passed to each child. It is like tossing a coin and getting heads or tails. The parent can’t control which copy of the hemoglobin gene is passed on to their child. However, they could find out which copy of the gene was given by specific testing during pregnancy or after birth.

Hemoglobin types are sometimes named according to the two types of genes they contain. The most common hemoglobin type is AA, meaning the person has two genes to make hemoglobin A, the usual type. As noted earlier, this is not the same as the A blood type.

What is sickle cell trait (or another hemoglobin trait)?

Often a person may not know they have a chance of having a child with sickle cell disease because both they and their partner are healthy. Having a child with sickle cell disease can happen because the parents both have sickle cell trait or another hemoglobin trait.

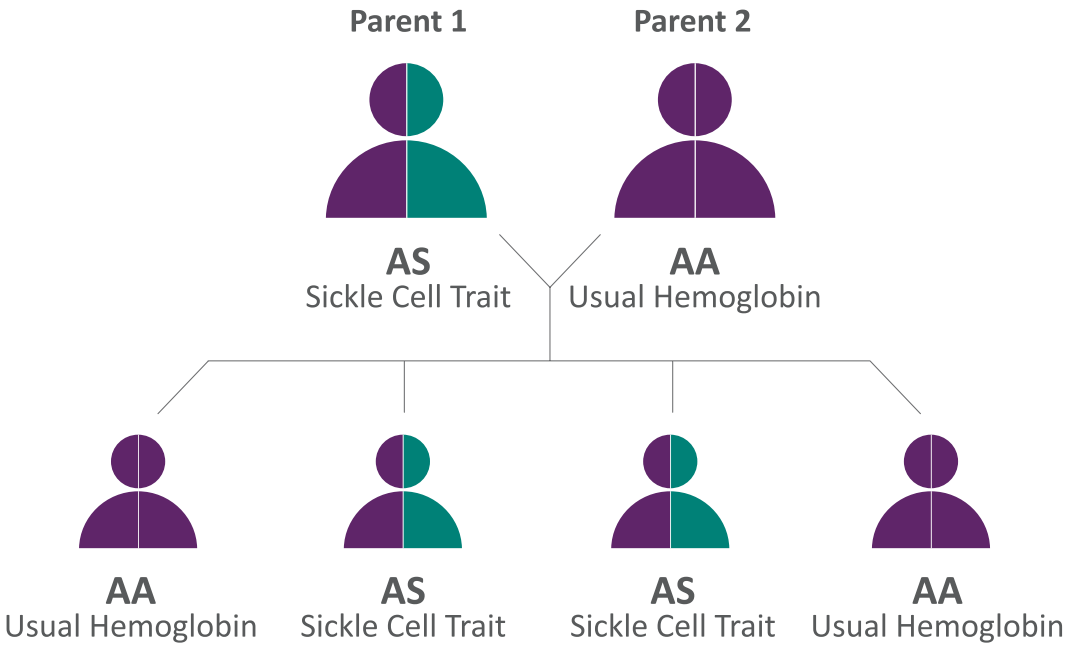
Someone with a hemoglobin trait has one gene for hemoglobin A and one gene for a different type of hemoglobin. People who have one gene for Hemoglobin S have sickle cell trait. Types of traits can be sickle cell trait (AS), hemoglobin C trait (AC), or beta thalassemia trait (AB^0 or AB^+). In addition, there are other less common traits.

Having a hemoglobin trait is not a disease. Most babies with a hemoglobin trait do not have any health problems caused by the trait. They do not have a mild case of the disease. They do not have a "trace" of the disease. Sickle cell trait does not cause health problems for most people. It never changes into sickle cell disease. It will not go away, either.

If both parents have a hemoglobin trait, some of their children may be born with sickle cell disease.

What are the chances of having a baby with sickle cell disease?

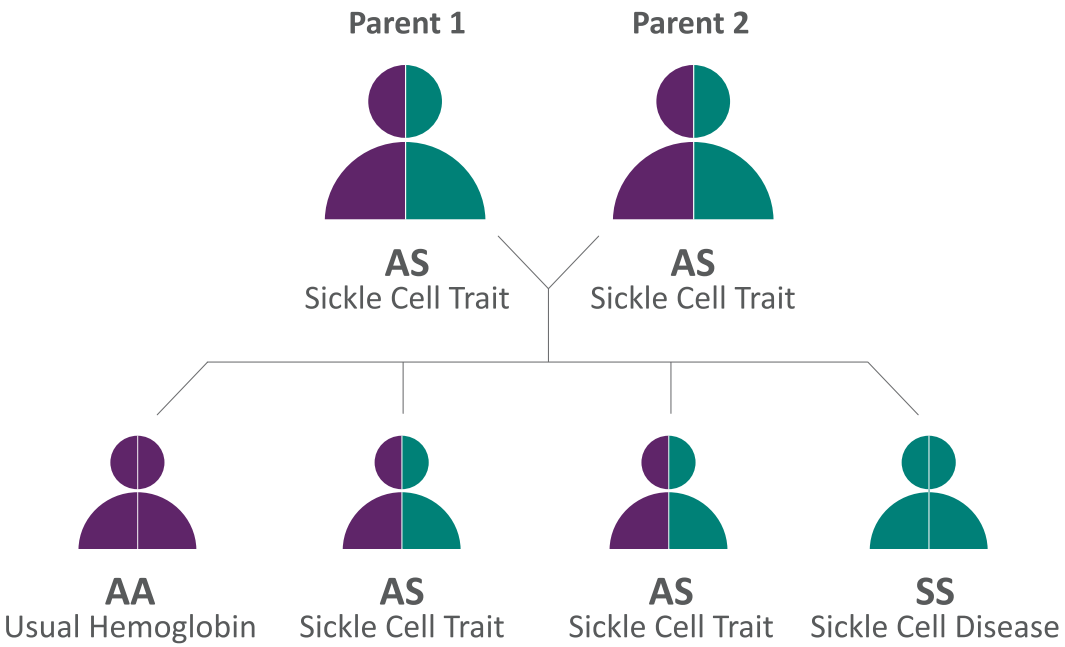
Example 1:
When one parent has sickle cell trait



A = usual hemoglobin A
S = sickle cell

When one parent has sickle cell trait and the other parent does not have either sickle cell trait or sickle cell disease, there is no chance that the parents will have a child with sickle cell disease. However, there is a 1 in 2 chance (50%) that the parents will have a child with sickle cell trait in every pregnancy.

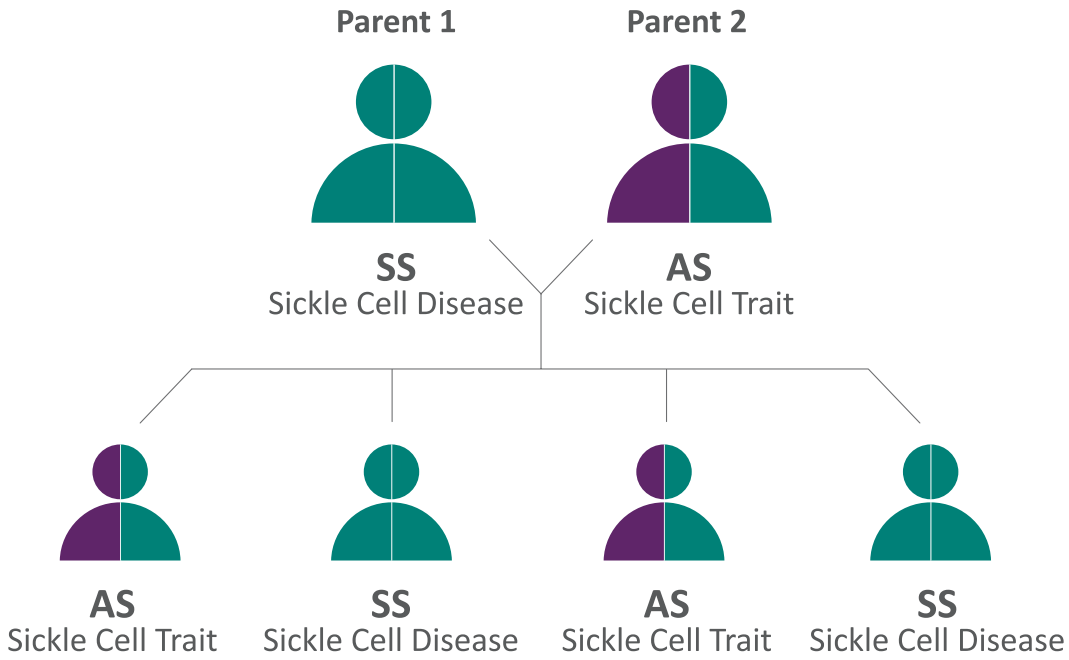
Example 2:
When both parents have sickle cell trait



A = usual hemoglobin A
S = sickle cell

When both parents have sickle cell trait, there is a 1 in 4 (25%) chance that the parents will have a baby with sickle cell disease. Every time this couple gets pregnant, they have the same 1 in 4 (25%) chance.

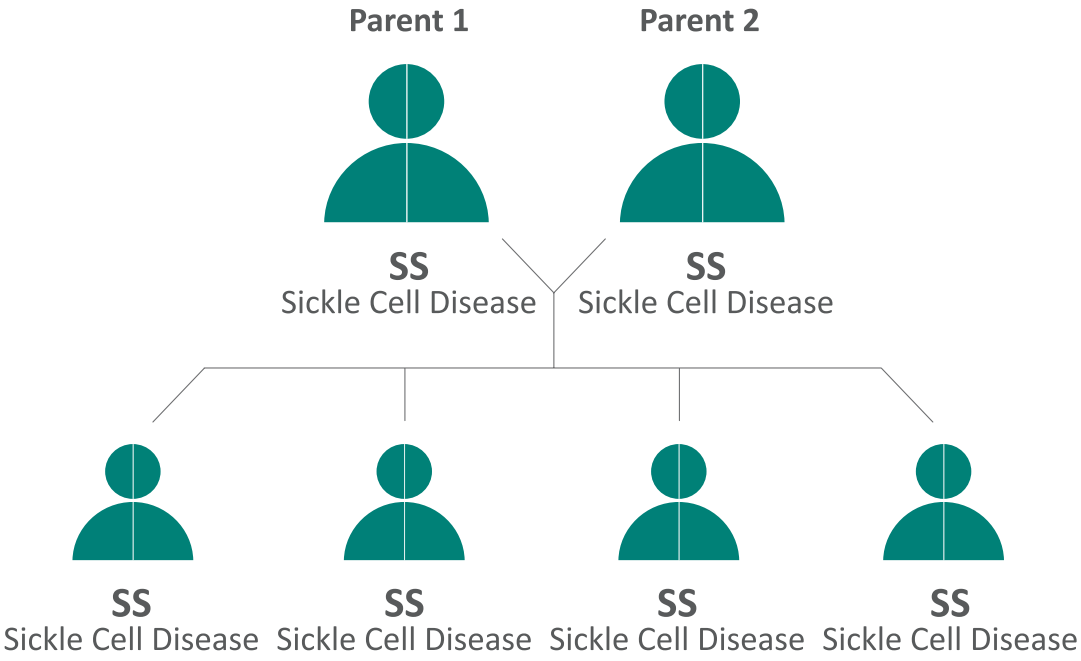
Example 3:
When one parent has sickle cell disease and the other has sickle cell trait



A = usual hemoglobin A
S = sickle cell

When one parent has sickle cell disease and the other has sickle cell trait, the parents will have a 1 in 2 (50%) chance of having a baby with sickle cell disease. Each pregnancy has the same 1 in 2 (50%) chance. If their child does not have sickle cell disease, the child will have sickle cell trait.

Example 4:
When both parents have sickle cell disease



A = usual hemoglobin A
S = sickle cell

If both parents have sickle cell disease, all of their children will have sickle cell disease.

See the online Appendix for charts to fill in hemoglobin types for you and your partner. Your genetic counselor or medical provider can help you understand what they mean for your family.

How can you find out what hemoglobin genes you have?

To know what type of hemoglobin you (the parent) or any member of your family has, you can ask your medical provider for a blood test. It is called hemoglobin electrophoresis with a complete blood count (CBC). This is the same test that was done to diagnose your child with sickle cell disease.

Sometimes a gene test is also done to detect the exact trait you might have. Many families carry genes for hemoglobin traits without knowing it. Your doctor or sickle cell disease care center can order this test for you. It's a good idea for all family members to know their own test results.

In California, screening for sickle cell disease in newborns started in February 1990. The California newborn screening method also began identifying babies with a hemoglobin trait, including sickle cell trait, at that time.

When a newborn is found to have a hemoglobin trait, the state sends their family a notification through the mail with a phone number to call for more information. A baby with a hemoglobin trait does not require special medical care. But the family will usually want to know this information and share it with their medical provider. They will want to keep this information in mind when the person with this trait plans a pregnancy.

People can contact the California Newborn Screening Program (www.cdph.ca.gov/NBS) for their trait result if they were born after February 1990. People with a hemoglobin trait can contact the Newborn Screening Hemoglobin Trait Follow-up Program for information, counseling, and free, voluntary, and confidential testing at 1 (866) 954-2229.

See Chapter 12, Sex and Reproduction, for more information about what to do before planning a pregnancy.

How many people have sickle cell disease?

Babies with sickle cell disease are born in every country and in every part of the world. It is most common in sub-Saharan Africa and South Asia.

Sickle cell disease affects approximately 100,000 Americans. In the United States, sickle cell disease affects:

- 1 in every 365 births in people of African descent
- 1 in every 16,300 Latino or Hispanic births

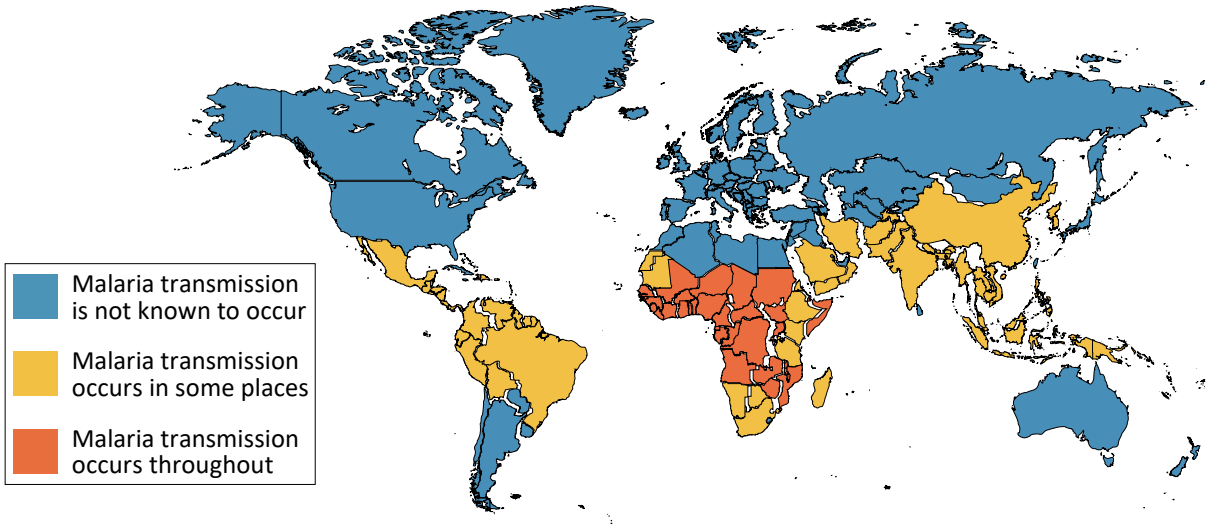
In California, an average of 84 babies were born with sickle cell disease each year from 2014 to 2018.

How many people have sickle cell trait?

In the United States, sickle cell trait happens in:

- 1 in every 14 people of African descent
- 1 in every 145 persons who are Latino or Hispanic

Sickle cell trait is also found in other ethnic groups such as Greeks, Yugoslavians, Western Asians, Turks, Southern Iranians, Mexicans, Puerto Ricans, Cubans, Spaniards, Native Americans, and South Asians, including people from India. Although sickle cell trait is more common in some groups of people, anyone can be a carrier of sickle cell trait.



This map shows areas where malaria happens. These areas are where sickle cell trait developed in people thousands of years ago. Source: CDC

Why is sickle cell trait so common?

Sickle cell trait developed in people thousands of years ago in areas where malaria was a major cause of death. Malaria is a disease caused by a parasite that lives inside red blood cells. A parasite is a tiny bug. Hemoglobin C trait, beta thalassemia, and other hemoglobin traits all developed at the same time as sickle cell trait.

The geographical regions with malaria include Africa, Central and South America, and various other regions. The reason that these traits developed is that they prevented severe malaria. They also helped people who caught malaria survive better compared to those without the trait. This survival advantage then allowed them to pass sickle cell trait to their children. Their children, with the trait, were healthier and more likely to grow up and pass the trait on to their children.

Because of this advantage, the genetic change or mutation that caused sickle cell trait (or C trait or beta thalassemia) increased over generations in the areas of the world where malaria exists. Sickle cell trait does not cause health problems for most people who carry it. However, if two copies of sickle cell trait are passed down from parents to their child, the child will develop sickle cell disease with all of its complications.

See the online Appendix for a history of sickle cell disease and a list of sickle cell “greats” - people with notable accomplishments.

Glossary of Sickle Cell Terms

Acute Chest Syndrome
Pneumonia caused by infection and sickle cell disease damage or just sickle cell disease damage in the lungs. Signs of acute chest syndrome may include fever, chest pain, coughing, shortness of breath, or difficulty breathing. This is a medical emergency.

Anemia (low red blood cell count)
When there is less hemoglobin in the blood than usual so that the blood can't carry as much oxygen.

Aplastic Episode
When the bone marrow stops making red blood cells. The blood count may fall much lower than usual. If it happens, it is usually with a fever or infection.

Avascular Necrosis
When sickle cell disease damages joints.

Carrier
A person who has one gene for Hemoglobin A and one gene for another type of hemoglobin. This person is also referred to as having a hemoglobin trait. A carrier doesn't have the disease, but two carriers can have a baby with sickle cell disease.

Chromosome
Structures containing the genes in the body. Most people have 46 chromosomes. Prenatal testing can be done to study the chromosomes of a fetus (developing baby) to determine their hemoglobin type.

Complete Blood Count (CBC)
A blood test that measures the size of the red blood cells and the amount of hemoglobin. It tells the number of red blood cells, white blood cells, and platelets.

Cross-Matching Blood
Before a blood transfusion, the blood bank mixes a sample of the donor blood with the blood of the person receiving a transfusion to check if the donor blood is a good match with the blood of the person who will receive it. A cross match is done with each transfusion.

Dehydration
A condition caused by not having enough water in the body. Dehydration can happen with diarrhea, fever, or exercise. It may cause a sickling episode in someone with sickle cell disease.

Electrophoresis
One of the best blood tests to find out a person's hemoglobin type. It shows most hemoglobin traits and can identify different types of sickle cell disease.

Exchange Transfusion
Blood transfusions can be given through an intravenous (IV) line to increase the hemoglobin to a normal level. They can also be done with a machine, which removes the sickle cell blood and replaces it with blood from donors. An exchange transfusion requires two IV lines: one to take the sickle blood out and one to transfuse the donor blood in.

Ferritin
Ferritin is the protein that carries iron in the body. Ferritin testing is used to show if the amount of iron in the blood is too high.

Gene
Genes are in our cells. They are the blueprint for our bodies. Genes are passed on by a mother in the egg and by a father in the sperm. People have about 35,000 genes that define many characteristics, including hemoglobin type.

Genetic
Related to genes and how physical traits and characteristics pass from one generation to the next. Sickle cell disease is a genetic condition.

Hemoglobin
The substance that carries oxygen in red blood cells. People with sickle cell disease often have lower hemoglobin levels.

Infarct
A blockage of blood flow that causes tissue to die because it doesn't have enough oxygen.

Inherited
A characteristic passed on from parents to their children. Sickle cell disease is an inherited disease.

Jaundice
Yellowish color of the skin or eyes. It is caused by chemicals from red blood cell breakdown.

Leg Ulcer
A breakage in the skin that begins as a small sore on the lower leg (above, over, or around the ankle). It can be caused by injury and decreased blood flow.

Neuropsychological Testing
This is testing done in young children or adolescents to find out how to help them if they are having a hard time in school. A specially trained psychologist will ask the child questions. There may be some questions for the child to read and then write answers. Sometimes testing can take as long as eight hours and has to be done in two sessions.

Pain Medication Dependency
Dependency is when the body has become so used to a dose of medication that when it is stopped, the person can get sick or feel bad. The doctor will gradually decrease the dose of medication to prevent a problem. Dependency is not the same as addiction.

Pain Medication Tolerance
When medications become less effective because they have been used for a long time, the body has developed pain medication tolerance. When that happens, higher doses are given to decrease the pain.

Pain Medication Withdrawal
Long-term use of some medications can cause the body to become used to it. Stopping the medication suddenly can create discomfort. Withdrawal discomfort includes fatigue, upset stomach, anxiety, and trouble sleeping.

Priapism
A persistent, painful, unwanted erection of the penis caused by sickling.

Prophylactic Penicillin
Penicillin that is given to reduce the number and severity of infections in children with sickle cell disease.

Pulmonary Hypertension
In sickle cell disease, sickling causes damage to the lungs. If pulmonary hypertension results, the heart has to pump harder than usual to get blood to the lungs. An echocardiogram will help find out if pulmonary hypertension is happening.

Red Blood Cell Phenotyping
Before a blood transfusion, the blood bank matches the proteins in blood between a blood donor and the person getting a blood transfusion. They do this because people who lack certain proteins may have a transfusion reaction to that protein if they are given blood with that protein from a donor.

Retinopathy
When the back of the eye (retina) is damaged by blockage in the small blood vessels and scarring in that area. This can lead to poor vision and even blindness. After ten years of age, every person with sickle cell disease should see an eye doctor once a year.

Sickle Cell Anemia
Another name for SS disease, the most common type of sickle cell disease.

Sickle Cell Disease
A term for all types of sickle hemoglobin disorders, such as sickle cell anemia (SS disease), SC disease and S beta thalassemia disease.

Spleen
An organ on the left side of the body that may be felt below the rib cage. It is a filter to remove bacteria and damaged red blood cells from the blood. This organ does not work well in people who have sickle cell disease. It can trap blood and become enlarged.

Splenectomy
Surgery to remove the spleen. This is done to cure the serious anemia that happens when blood is trapped in the spleen (splenic sequestration, see below).

Splenic Sequestration
A medical emergency in people with sickle cell disease that is caused by blood being trapped in the spleen very rapidly, depriving the rest of the body of blood. It can be life threatening.

Stroke
A loss of blood flow to part of the brain, which damages brain tissue. Sickle cell disease increases the chances of a stroke in children. Seeing medical providers regularly can help decrease the chances of a stroke in children with sickle cell disease.

Trait
A person who has one gene for Hemoglobin A and one gene for another type of hemoglobin has a hemoglobin trait. This person is referred to as a carrier (see above).

Transfusion
Blood from one person given to another person. People with sickle cell disease receive transfusions for many reasons, including a very low red blood cell count, to prepare for surgery or to treat certain complications of sickle cell disease.

Vaso-Occlusive Episode
Also called a pain episode. This can happen when sickle cells block the flow of blood. It causes pain and, if severe, tissue damage.

Appendix

The Appendix and PDF versions of this handbook are available online (go.cdph.ca.gov/SCParents). Appendix includes:

Scan this QR code with a mobile phone camera to go to the Handbook and Appendix web page

1. Information or Links:

- Glossary of Sickle Cell Terms
- History of Sickle Cell Disease
- How to Feel for Your Child's Spleen
- Sickle Cell Disease and Nutrition
- Sickle Cell Disease Greats
- California Children Services (CCS)-approved Sickle Cell Disease Centers
- California Clinical Network and Community Based Organizations
- Child Development Assessment: Milestones and Denver Developmental Screening Test
- Danger Signs: When to Contact a Medical Provider
- Diagram of Sickle Cell Disease Inheritance Tool (Punnett Squares)
- Fahrenheit to Celsius and Celsius to Fahrenheit Temperature Conversion Charts
- Resources for Sickle Cell Disease, Including on Clinical Trials, Transition, and Filing Complaints
- Sickle Cell Disease and Adulthood: What Medical Issues to Expect
- Sickle Cell Disease and an "IEP" and a "504-Plan" in Public Education
- Suggested Ibuprofen and Acetaminophen Dose Charts
- Type of Medical Providers List
- When to Get Shots (Vaccines or Immunizations) for Your Baby, Preteen, or Adolescent

2. Sample Forms, Letters, or Plans

- Authorization for Medical Treatment Form
- Authorization for Release of information Form
- Comprehensive Sickle Cell Disease Care Plans: Birth to 6 Years and 6 Years to Adult
- Doctor Letter to Schools about Physical Education
- Doctor Travel Letter
- Pain Plans
- School Plans: "IEP" and "504"

3. **Handbook Acknowledgements**

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California Department of Public Health

California Newborn Screening Program
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Parent and Caregiver Handbook for Sickle Cell and Online Appendix
(go.cdph.ca.gov/SCParents)

Scan this QR code to go to the Parent and Caregiver Handbook for Sickle Cell and Online Appendix web page