A PARENTS' HANDBOOK FOR SICKLE CELL DISEASE

PART I

Birth to Six Years of Age

Modified: 1/6/06

Referral Components For Pediatric Comprehensive Sickle Cell Services In Virginia

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A PARENTS' HANDBOOK FOR SICKLE CELL DISEASE

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PART I

Birth to Six Years of Age

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Note to Health Care Providers

This handbook was developed in response to requests from parents of children with sickle disease for more detailed information that they could refer back to as needed. The parents' role in early identification of infection and other problems is difficult yet so critical to the health of their child particularly during the first few years of life. It was hoped that the document would assist in empowering parents' to assume this role as partners in the delivery of health care to their child. It was also hoped that the handbook would be useful for parents to share with their primary care provider and emergency room staff unfamiliar with the treatment of sickle cell disease.

The role that parents are being asked to play in the treatment of sickle cell disease has become very complex, particularly with statewide newborn screening and prophylactic penicillin treatment. This handbook is one method that can be very useful to providers in teaching this important information to parents. The intent was not for parents to sit down at one session and read the document, rather for health care providers to utilize it as a teaching tool at visits. Parents should be encouraged to keep the booklet and write down their notes and questions in it. The various sections could be reviewed as appropriate at different visits.

Also, the parents could use the guide to look up terms and explanations given at clinic visits that they either did not understand, or needed to review.

Parents of children with sickle cell disease were involved in the development of this booklet from the very early stages. The formal field test with consumers consisted of asking for feedback on the readers understanding of the material, usefulness, quantity of information, usability as a reference tool, completeness, messages of graphics, cultural sensitivity and overall appeal of handbook. Field testing was done both with individuals and focus groups. Approximately thirty families throughout California participated in the formal field testing process.

Reading level was an important issue to us from the onset, and testing was done throughout the process of developing this booklet. We never set out to develop an easy-to-read pamphlet; however we did want the handbook to be at a reading level suitable to as many adult readers as possible without compromising the original purpose. The final version of the handbook is at a ninth grade reading level using the Fry test. It varies from section to section but most of the document ranges from eighth to tenth grade.

We realize that this reading level is high. However, when the document is used as intended in conjunction with clinic visits, with specific information being highlighted by the staff, the document has been useful even to people with low reading skills. Furthermore, the format of the booklet such as size of margins and use of graphics to reinforce messages, as well as the amount of empty space on each page all contributed to making the document overall more readable and appealing. The feedback from parents has been very positive.

Another consideration in use of the booklet is time of its introduction. We do not recommend giving it out to all parents of infants with sickle cell disease at their first visit. Only when parents seem ready for more information should they be given the handbook.

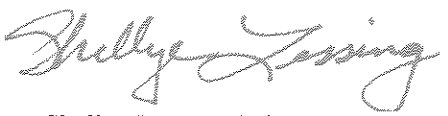
Preface

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 health care professionals about your child's future. It may seem like there is so much to learn, and 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 for you to use.

Use this handbook to help you get involved with your child's care! Learn about sickle cell disease and how to deal with some of the most common symptoms so that you can help your child lead a full life. By working together with your child's doctor and other health care providers, you can make sure that your child gets the best care.

We hope that you will share this handbook with others that are close to you so that they can learn about sickle cell disease, too. Other family members, friends, child care providers, and your family doctor may all have questions that this book can answer. The more they know, the more they can help you and your child. You don't have to manage it all yourself.

The families and staff of the Children's Hospital Sickle Cell Center in Oakland, California who have worked to put this book together hope that this book will help your child live a happy and healthy life.



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Editors

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Sickle Cell Center

NOTE: In half of the chapters of this book, your child will be referred to as male. In the other half, your child will be referred to as female. **All of the information applies to both girls and boys.**

Acknowledgements

We would like to thank **Ghaliyah Roberts** for preparation of the manuscript; **Caroline Hastings, MD,** for review of the manuscript and helpful additions; **Barbara Gaffield, RD,** for her comments on nutrition; **Melinda Lassman, MA, MS,** Regional Network Coordinator, **Linda Lustig, MS,** Chief, Prenatal Genetic Services Section, **M. Eileen McElroy, RNC, MSN,** Nurse Consultant, **Virginia Mordaunt**, Chief, Newborn Screening Section, **Rhonda Schonberg, MS**, Genetic Disease Program Specialist and **Karen Whitney, MS**, Genetic Disease Program Specialist of the California Department of Health Services GeneticDisease Branch for their review; and the Department of Health Services Genetic Disease Branch, **George C. Cunningham, MD, MPH, Chief,** for funding the development and distribution of this book.

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Special thanks to the families of children with sickle cell disease who made suggestions during production of the book, modeled for the illustrations, and who were the inspiration for the project.

For more information, contact your local Sickle Cell Program office listed below:

Table of Contents

Basic Questions

This section provides answers to many of the first questions asked by parents of children with sickle cell disease. These questions include:

- **What is sickle cell disease?**
- **What problems are caused by sickle cell disease?**
- **How serious is sickle cell disease?**
- **What causes sickle cell disease?**
- **How can I help my child with sickle cell disease?**

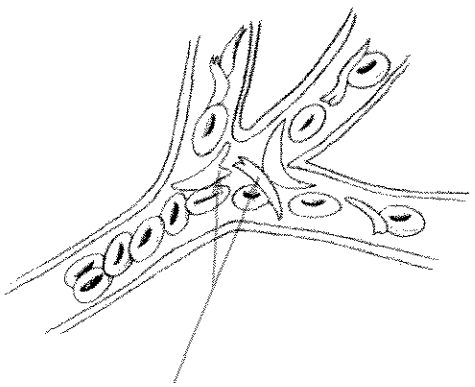
The answers to these questions and the suggestions in this handbook will help you give your child the best start. With good medical care and home care, most children with sickle cell disease can grow up to lead full and productive lives.

Q What is Sickle Cell Disease?

Sickle cell disease is a disease that affects a special protein inside our red blood cells called hemoglobin. Red blood cells have an important job. They pick up oxygen from the lungs and take it to every part of the body. It is the hemoglobin in these cells that carries the oxygen to different parts of the body.

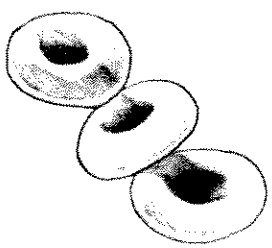
A person with sickle cell disease makes a different kind of hemoglobin. This causes the red blood cells to change their shape. Instead of being smooth and round, the cells become hard and sticky. Their shape looks like a banana or like a sickle, a hand tool used to cut wheat or tall grass. It is this sickle shape of the red blood cells that gives "sickle cell" disease its name.

The hard, sticky sickle red blood cells have trouble moving through small blood vessels. Sometimes they clog up these blood vessels so that blood can't bring oxygen to the tissues. This can cause pain or damage to these areas.

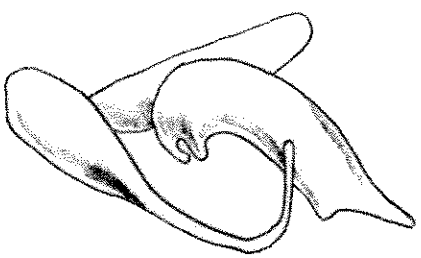


Sickle cells

Hard, sticky sickle cells clogging up a small blood vessel.



Normal red blood cells.



Sickle red blood cells.

Q What Problems are Caused by Sickle Cell Disease?

Playing like other children.

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

- ❑ Infections
- ❑ Pain
- ❑ Anemia (low blood)
- ❑ Damage to the body organs

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

Infections

One of the most serious problems that people with sickle disease have is infections. Infections, like pneumonia, pose a special problem for infants and small children who can get very sick or even die if they don't get prompt treatment.These infections are caused by problems with the spleen, the biggest lymph node in the body. Lymph nodes, like the spleen, help the body kill germs. The sticky sickle cells will clog the spleen so it can't do its job. This leaves the body open to infections.

Thankfully, we can prevent many of the sickle cell infections by giving young children penicillin every day until they are at least five years old, or as recommended by the National Institute of Health (NIH) Consensus Development Conference. There are also many ways to treat infections, especially if they are found quickly. Later sections of this book describe ways to help prevent and treat these deadly infections.

Pain

The hard, sticky sickle red blood cells caused by sickle cell disease can sometimes cause pain. The shape of these cells makes it hard for them to get through tiny blood vessels. When they get stuck, these cells pile up and block the blood vessel. This cuts off the blood supply to nearby tissues so that no cells can get through to bring oxygen. Without oxygen, the area starts to hurt. This is the source of the pain that comes from sickle cell disease.

Some sickle cell pain can be very strong and needs to be treated in the hospital. Most pain is milder and can be handled at home. There are many ways to treat the pain to make your child feel better. This pain is sometimes called a "crisis." In fact, most sickle cell pain is not a crisis. Later sections of this book will describe ways to help you deal with this pain.

Leading full, productive lives.

Anemia (low blood)

Besides causing pain, something else happens because of the shape of the sickle cells. These cells are pulled out of the blood and broken down faster than regular red blood cells. The body can't make enough new red blood cells to replace the old ones. This decreases the number of red blood cells and the amount of hemoglobin in the body. This "low blood count" is called anemia. If the anemia becomes severe, your child may need to be given blood to prevent heart failure and other problems.

Damage to the body organs

Over many years, lack of oxygen due to clogged blood vessels can lead to tissue damage. This damage can happen to any organ.

While not all tissue damage can be prevented, some of it can. With early treatment and good self care, people with sickle cell disease can lessen the damage to their bodies.

Most children and adults with sickle cell disease can lead full lives

Not everyone with sickle cell disease will have all of these problems. Many have very few problems and may go years without pain or hospital care. Only a few 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 lives like other people. While they may miss school or work or go to the doctor or hospital more often than others, their lives can still be full and happy.

Note: Sickle cell disease affects the body, not the mind. Sickle cell disease does not affect how smart a person is.

Q How Serious is Sickle Cell Disease?

Sickle cell disease is a chronic disease. It can't be cured, but it can be treated. People with sickle cell disease can live well into middle and late adulthood. With penicillin to help stop infections, today's children with sickle cell disease are living longer than ever before.

Sickle cell disease affects different people in different ways. No one can know how serious the disease will be for your child. But we do know that three things can make a difference:

a. The type of sickle cell disease.
b. The kind of care a person gets.
c. How the person and the people around him deal with the disease.

The type of sickle cell disease

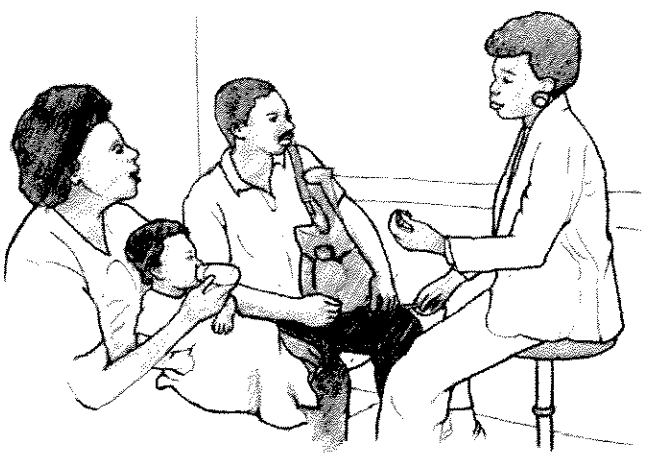
There are many different types of sickle cell disease. The two most common types are sickle cell anemia (SS disease) and sickle "C" disease (SC disease). Sickle beta thalassemia disease (S beta thal disease) is another type, but it is less common.

Some types of sickle cell disease cause fewer problems than others. For example, SC disease is often less serious than SS disease.

The kind of care a person gets

Poor medical care and home care can make a chronic disease like sickle cell much more serious. For example, if a fever is not treated early, a child can become very sick. On the other hand, getting the best medical and home care can help a person live longer and better. Good medical care includes frequent visits to a doctor who has experience with sickle cell disease. It can also mean getting help from other health care providers, like social workers, counselors and physical therapists.

Good home care includes many things, from giving young children penicillin twice a day to having them drink lots of fluids. Since we spend much of our time at home this care can often have a big impact.

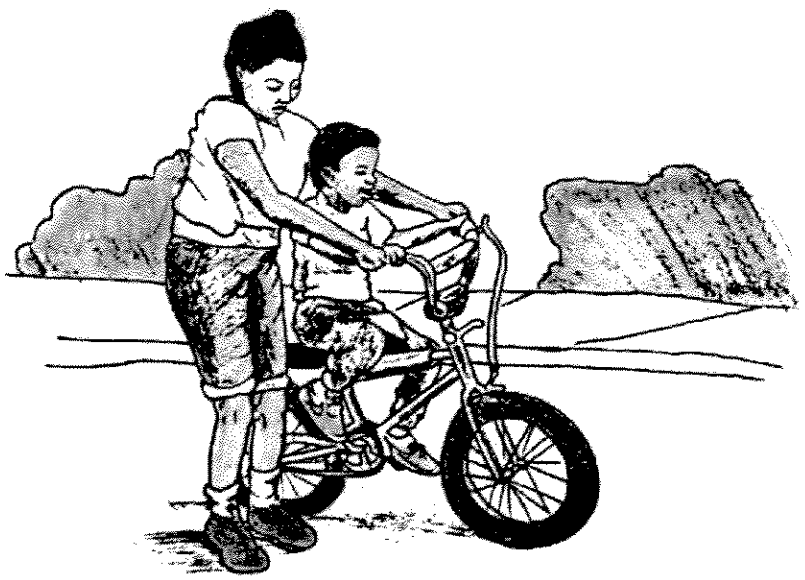


Getting good medical care.

How the person and the people around him deal with the disease

While people with sickle cell disease share common experiences, the way they deal with them can be very different. For example, when it comes to pain, some children are able to deal with it as if it is a part of their everyday life. Other children may have the same amount of pain, but have a tougher time handling it.

People can learn to handle these experiences better, and families can react in ways which help their child. If a person with sickle cell disease learns positive ways of dealing with his problems, the disease will often feel less serious.



Living with sickle cell disease.

Q What Causes Sickle Cell Disease?

Hemoglobin type and blood type are not the same. Everyone has both a hemoglobin type and a blood type.

Sickle cell disease is inherited

Sickle cell disease is an inherited disease. An inherited disease is one which is passed from parents to their children through their genes.

Genes are our body's map for development. We have pairs of genes for the color of our eyes, for our height, for our blood type and for each of our other features, including our hemoglobin type.

A baby gets one hemoglobin gene from each parent

To make up our pair of hemoglobin genes, we get one gene from our father and one gene from our mother. Each of our parents has two genes for hemoglobin, but they pass only one of these genes on to each child. Which gene is passed on is a matter of chance, like having a boy or a girl or like tossing a coin and getting heads or tails.

To inherit sickle cell disease, a child must get the sickle (S) gene from one parent and a sickle (S), C, or B (beta thal) gene from the other parent. If a baby inherits at least one hemoglobin A (usual adult hemoglobin) gene, he won't get sickle cell disease.

Both parents have a hemoglobin trait

Someone who has one gene for hemoglobin A and one gene for a different type of hemoglobin has a hemoglobin **trait**. This trait could be AS (sickle cell trait), AC (C trait) or AB (Beta thal trait). In addition, there are other less common traits.

A trait is not a disease

People with a hemoglobin trait are healthy. They do not have a mild case of the disease. They do not have a "trace" of the disease. A trait does not cause health problems. It never changes into sickle cell disease.

If a man and woman both have a hemoglobin trait, some of their children may be born with sickle cell disease.

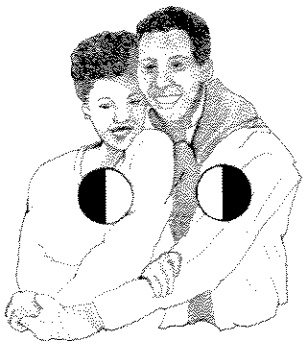
How to find out about your hemoglobin genes

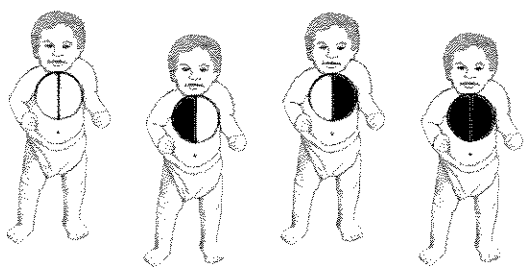
The only way to know for certain what type of hemoglobin you have is to have a special blood test called hemoglobin electrophoresis with a complete blood count (CBC). Many families carry genes to make other types of hemoglobin besides hemoglobin A without knowing it. Your doctor or sickle cell center can order this test for you.

Chances of Having a Baby With Sickle Cell Disease

Both parents have sickle cell trait

Sickle Cell Trait (AS) **Sickle Cell Trait** (AS)



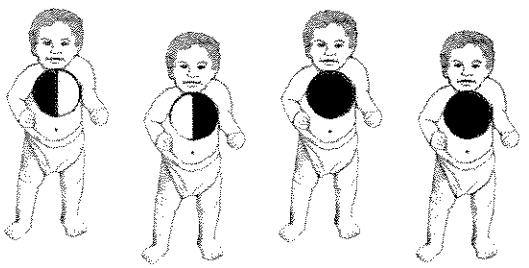


No Trait (AA) | Sickle Cell Trait (AS) | Sickle Cell Trait (AS) | Sickle Cell Disease (SS)

When both parents have sickle cell trait, they have a 25% chance (1 out of 4) of having a baby with sickle cell disease. Each time they get pregnant, they have the same chance.

One parent has sickle cell disease and one parent has sickle cell trait

Sickle Cell Disease (SS) **Sickle Cell Trait** (AS)



Sickle Cell Trait (AS) | Sickle Cell Trait (AS) | Sickle Cell Disease (SS) | Sickle Cell Disease (SS)

When one parent has sickle cell disease and the other has a trait, they have a 50% chance (or 1 out of 2) of having a baby with sickle cell disease. Each time they get pregnant, they have the same chance.

A 25% or 1 out of 4 chance does not mean that if you have 4 children, 1 will have the disease. You have the same 25% chance with each baby.

How many people have sickle cell trait?

In the United States, one out of every 10 to 12 people of African descent has sickle cell trait (not the disease). About 1 out of 35 to 50 African-Americans have C trait.

Sickle cell trait is also found in other ethnic groups, such as Greeks, Yugoslavians, Western Asians, Turks, Southern Iranians, Asiatic Indians, Mexicans, Puerto Ricans, Cubans, Spaniards, and American Indians. As more babies are screened for sickle cell disease, we will learn more about these groups.

How many people have sickle cell disease?

Sickle cell disease affects about one out of every 400 African-American babies born in the United States. Information about the disease in other ethnic groups is not well known.

Q How Can I Help My Child With Sickle Cell Disease?

There are many things you can do to help your child manage his disease. While you can't get rid of the disease, you can help your child get sick less often and feel better. You can also help your child learn how to live with the disease so that he can make the most of his life.

Here are some of the important things you can do.

Get the best medical care

❑ Find a doctor for your child who is experienced in sickle cell disease. Take your child for well child exams as often as the doctor orders.

❑ Make sure your child gets all his baby shots on time.

Getting the best medical care.

❑ Help the doctor give your child the best care. Answer his or her questions fully and ask any questions you have about what to do for your child.

❑ Learn as much as you can about the disease. The more you know, the better care you will be able to give your child.

Take good care of your child at home

❑ Give your child penicillin twice a day, until at least age five. Don't stop until your doctor tells you to stop.

❑ Give your child lots of fluids to drink when he is sick, in pain, very active or taking a trip and when it's hot outside. The rest of the time, just make sure your child gets fluids to drink whenever he is thirsty.

❑ Call the doctor right away if your child has a fever of 101°F or higher. Check your child's temperature when you think he might be sick.

❑ Help your child handle any pain he has from the disease. Try more fluids, quiet play, warm baths, heating pads or warm towels, massage or Tylenol. If these home remedies don't ease the pain, then call the doctor.

Get help for your child when it's needed

Read the list of danger signs on page 33. There is a copy of these signs in Appendix A for you to hang on your wall or refrigerator. Put it up in a place you can find when something is wrong with your child. Check the list to see what you should do. If your child should be seen right away, call your doctor or nurse first so they can tell you where to take him.

You know your child better than anyone else. If you think something might be wrong, call your doctor or nurse. They will help you decide what to do.

Take care of yourself and your family.

Having a child with sickle cell disease is not easy. The disease affects the whole family. Help those who are close to your child learn about the disease. Let them read this book or talk to them about what you've learned. When they know more, they can help both you and your child.

You can also get support from family and friends. Let them know how they can help you. You might need someone to listen to you or someone to watch your other children. Think about what you need and ask for it.

Other parents who have children with sickle cell disease can give you a special kind of support. Find out if there is a parent support group near you and join it.

If things get to be too much for you, get help. Talk to your doctor, clergy, or a social worker. You are not alone. Make sure that you get what you need to give your child the best care.

Read this handbook

The rest of this book will describe these things in much more detail. We hope that you will use this book to learn how to best help your child live with the sickle cell disease. Together with your child's health care team, you can help your child live the fullest life.

Routine Medical Care For Your Child

2

Regular check-ups are a must for all children with sickle cell disease. At these visits, the doctor will check your child's growth and development. Your doctor will also check to see if your child is having any health problems.

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

- **Where to go for care**
- **Well child exams**
- **Baby shots**
- **Common lab tests**
- **Questions to answer**
- **Information to share**
- **Learning from your medical team**

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

Where To Go For Care

Your child can get basic medical care from a pediatrician or family practice doctor. Depending on where you live, a combination of these may work out best.

Pediatrician or family practice doctor

Your family doctor or pediatrician can provide most of the care your child will need. This includes well-baby care, routine shots and treatment for some illnesses. Your family doctor will refer you to a sickle cell center or a hematologist for special care. These specialists will work together with your family doctor to make sure your child gets the care she needs.

Sickle cell center

In some places, there are special centers which have a team of experts trained in sickle cell disease. These centers do most of the research on sickle cell disease. They also train other doctors and health care professionals.

The health care team at sickle cell centers includes a pediatrician and other doctors who can provide both routine medical care and special care related to sickle cell disease. Besides doctors, this team may also include nurse practitioners, nurses, genetic counselors, social workers, psychologists and others who can help you and your child.

See Appendix B to help you find the nearest sickle cell center.

Getting a complete evaluation

To help your family doctor give your child the best care, we suggest that your child start with a complete evaluation at the nearest sickle cell center. The medical team at the sickle cell center will share the results of this evaluation with you and your family doctor. The sickle cell center will also let you know how often they think your child should come back to the center to keep track of how she is doing.

Taking your child to a sickle cell center for a complete evaluation.

Talking about your child's case at a health care team meeting.

Well Child Exams

During the first five years of life, your child will have many check-ups. From birth to six months old, your child should be checked once a month. Between six months of age and one year, your baby should go to the doctor every two months. After one year, visits should be set up for every three to four months until your child is five. If your child is sick, she will need to be seen more often.

How often your child should see a doctor

Age	How often
Birth - 6 months	every month
6 months to 1 year	every 2 months
1 year to 5 years	every 3-4 months

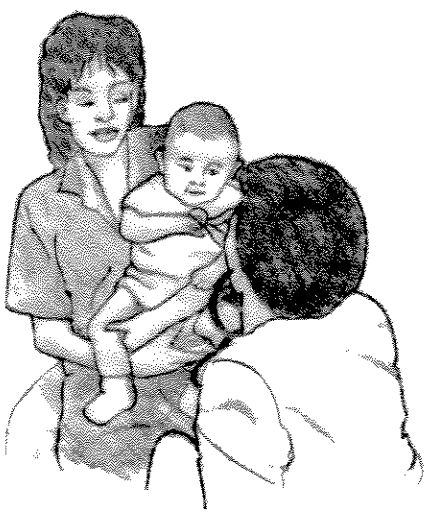
At these doctor visits, the medical staff will check your child's:

1. Temperature
2. Heart rate and breathing rate
3. Height and weight
4. Blood pressure (when she is old enough)
5. Head size

Next, the doctor or nurse practitioner will check your child's:

1. Eyes, ears, mouth and throat
2. Soft spot on the head (until one year of age)
3. Neck
4. Heart and lungs
5. Abdomen
6. Spleen size
7. Skin
8. Penis and scrotum or vaginal area
9. Joints
10. Back

See Appendix C for a sample Comprehensive Sickle Cell Disease Care Plan. It describes what should be checked at different ages.



Getting a well child exam.

Baby Shots

(Vaccinations)

"Baby shots" are a very important way to protect your child's health. Because children with sickle cell disease get more infections than other children, they need these shots even more.

Your child will get the same baby shots that other children get. Plus, she will get some other shots to help her fight infections.

Giving the doctor a record of your child's shots.

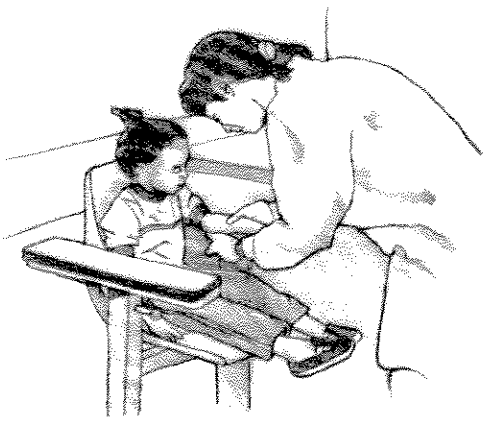
Here is a list of the shots your baby will get and why they are given.

Regular Baby Shots*		
Shots	**Protects Against**	**Age Given**
DTP	Diphtheria Tetanus (lockjaw) Pertussis (whooping cough)	2 months, 4 months, 6 months, 15-18 months, before entry to kindergarten
OPV	Polio	2 months, 4 months 15-18 months, before entry to kindergarten
MMR	Measles Mumps Rubella (German measles)	15 months**, before entry to kindergarten
(HIB)	Meningitis	2 months, 4 months, 6 months, 15 months***
Extra shots for children with sickle cell disease		
Pneumococcal Vaccine	Some pneumonias and blood infections	9 months optional 24 months, 5 years
Flu shot	Influenza (flu)	varies
Hepatitis B Vaccine	Hepatitis (liver infection)	varies

* Source: California Department of Health Services, Immunization Unit
** In some areas the original dose is given at 12 months.
*** HIB Schedule depends upon brand of vaccine and age started.
In addition to shots, children with sickle cell disease need to have a TB (tuberculin) skin test done yearly, starting at 12 months old.

See Appendix C-1 for a chart of when these shots and tests are done .

Common Lab Tests



Getting your child's blood drawn.

When your child goes to the doctor, she may be sent to the lab for blood or urine tests. For a blood test, they will take some blood from her finger or arm with a tiny needle. This may hurt like a little pinch. It is important to tell your child that taking blood is not the same as a shot, and it won't hurt afterwards. If a urine sample is needed, the nurse will explain how to get it. These are some of the most common tests:

Hemoglobin electrophoresis

This is the test which is used to find out a person's hemoglobin type. It is this test which 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 is in your child's blood. Before a blood transfusion, doctors use the test to help decide how much 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 complications from sickling.

Complete blood count (CBC)

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

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

Reticulocyte (Retic) count

Reticulocytes are young red blood cells. The number of these cells shows whether the bone marrow is doing its job well, making and releasing young cells into the blood.

Kidney and liver function tests

These tests show if these organs have been damaged by sickle cell disease. In young children, damage is rare. However, over time, the sickle cells can plug up the small blood vessels of these organs so that they can become damaged.

Urine test (urinalysis)

In sickle cell disease, children can get kidney or urinary tract infections. Urine is checked under a microscope for signs of infection. With a bacterial infection, urine is cloudy, smells bad and tiny swimming bacteria and white blood cells can be seen. Red blood cells in the urine can be a sign of slight bleeding from the kidney. A lot of protein in the urine can mean kidney damage, but a small amount doesn't usually matter.

Blood Chemistry Tests

These tests measure substances in the blood which are important for health and growth, such as iron, glucose (sugar), and minerals.

X-Rays

X-Rays are used to see if there is an infection in the lungs and to look at bones which may be damaged by sickle cell disease.

Learning From Your Medical Team

An important part of every visit is to get answers to questions that you have. Be sure to ask your doctor or nurse about anything that you don't understand or that concerns you. No questions are silly or stupid. All of your questions are important, and the staff will be happy to answer them.

Sometimes it helps to write down your questions or concerns before you take your child in. Then you can check your notes to make sure that you remembered everything.

Your child's visits are a time for you to learn more about sickle cell disease and how to help your child. Get to know the people who are caring for your child and learn from them at each visit. The more you learn about the disease, the better care you will be able to give to your child.

Getting answers to the questions you wrote down.

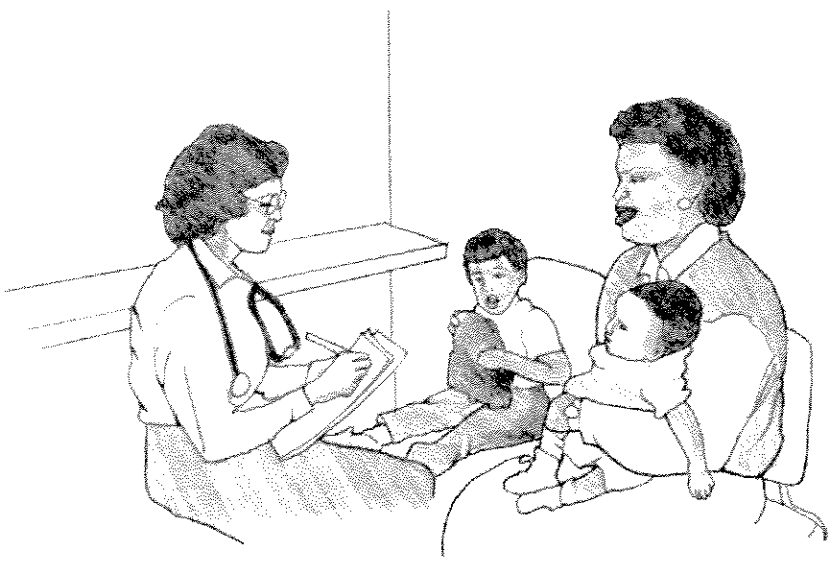
Questions To Answer

During the exams the doctor or nurse 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 these questions before you see your doctor.

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

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

Your doctor needs to know about any illnesses you may have treated at home or had treated elsewhere. It is very important to always call or see your doctor right away if your child has a fever, jaundice or seems sick.



Answering the doctor's questions.

2. What is your child eating?

This helps the medical staff to 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.

3. What medicines are you giving at home and how much?

Your doctor also 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. Bring all your child's medicines to each visit to make sure you give the doctor the right information.

4. Is your child having any problems taking prescribed medicine, like penicillin, at home?

Your doctor needs to know if your child is having any problems taking medicine. For example, you may have trouble getting the prescription filled or getting your 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 she gets the medicine she needs.

5. What does your baby do? Smile? Roll over? Talk?

It is important to know if your baby is developing normally.

Information to Share

Many things can affect your child's health. The more the doctor or nurse knows about your child's life, the better care they can give. Be sure to tell your doctor or nurse at the sickle cell center if:

1. Your child has been treated by another doctor or clinic.
Your doctor needs to know about what problems your child was having and what treatment or tests were done. If any medicine, shots or tests were given, your doctor needs to know so that the tests won't be repeated unless needed. Your doctor will give you an Authorization for Release of Medical Information Form because he or she will want a copy of the records so that they can be added to your child's medical record.

See Appendix E for sample form for Authorization for Release of Information.

2. Your child looks or acts in a way that concerns or upsets you.
You know your child better than anyone else. Your doctor may miss things. Help your doctor by sharing things that upset or concern you.

3. Your child has had a major loss, such as 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 doctor may want to watch her more closely.

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

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

6. You are planning to take a trip.
Ask your doctor to give you a letter stating:
- What kind of sickle cell disease your child has
- Her normal hemoglobin level
- The medication she takes
- Other special problems
- Who to call in case of an emergency

Also, ask your doctor or nurse about where you should take your child if she needs medical care on your trip. Take the letter with you when you travel with your child.

See Appendix F for sample travel letter.

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

Health Care At Home

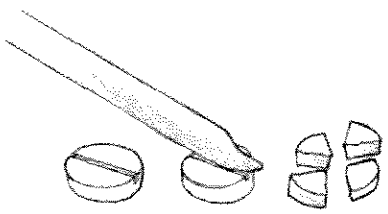
Children with sickle cell disease need some extra health care at home to help them stay well. Giving your child penicillin twice a day (at least until age 5) is one of the key parts of good home care. Extra fluids and a good diet also help to keep your child well. With just a little extra care, your child can be active, go on trips, and do almost all of the things that other children do.

In this chapter, you'll learn about the following areas so that you can help your child stay well:

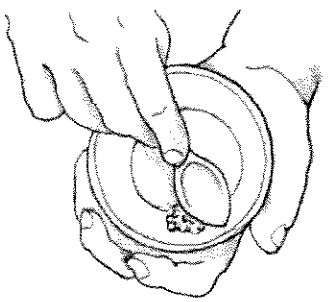
- **Penicillin**
- **Fluids**
- **Nutrition**
- **Active play**
- **Taking a trip**

Penicillin

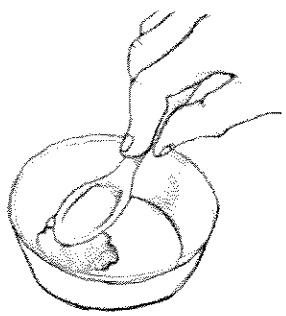
Preparing penicillin



Step 1: Cut the pill into small pieces.



Step 2: Mash the pieces in a bowl with a spoon.



Step 3: Mix the mashed pills with some food.

Your doctor will tell you when to start giving your baby penicillin. He will need it twice a day, once in the morning and once in the evening. Studies have shown that daily doses of penicillin for babies and young children with sickle cell disease greatly reduce the number of infections they get. Your doctor will write a prescription for your child. Making sure your child gets his penicillin is one of the most important things you can do.

Your child needs to take a dose of 125 milligrams (mg), twice a day, until he is three years old. At three years of age, the dose will double to 250 mg, twice a day. If your child is allergic to penicillin, erythromycin can be given instead.

Keep giving your child penicillin until your doctor or nurse practitioner tells you to stop. Don't stop because your child feels well or sick, unless your doctor tells you to stop. Most doctors recommend taking penicillin until at least age five.

Pills

It is better to give penicillin in pills because they last for several years. Liquid penicillin only lasts for two weeks after you get it from the pharmacy, and it must be refrigerated.

Pills can be crushed and mixed with a teaspoon of applesauce, ice cream or flavored yogurt. It is not good to put them in formula or juice because the baby may not drink all of it.

Liquid

Liquid can be given by spoon or dropper. Liquid penicillin should not be put in a baby's bottle because the baby might not drink all of it.

Shots

If your child is having problems taking penicillin at home, penicillin shots can be given once every three weeks at a doctor's office.

Amounts of penicillin	
Age	**Dose**
2 months - 3 years	**125 mg.** 2 times a day
Over 3 years	**250 mg.** 2 times a day

Helpful hints

It can help to give the penicillin at the same time each day. Involve your child as he gets older. Make it a game or put your child in charge of how he takes it. Even a four year old can help remind you that he needs to take his pills.

Fluids

Giving your breastfed baby extra fluids when he is sick.

Children with sickle cell disease need more fluids than other children. They usually get thirsty more often than other children. Give your child fluids whenever he is thirsty. Fluids can include any clear liquids like water, juices or soft drinks. Keep enough fluids on hand so that your child can have as much as he wants.

Special times when your child needs to drink more

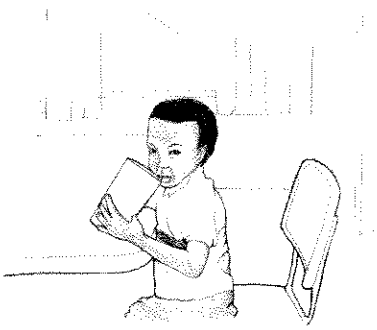
Your child needs more fluids when:

1. He has a fever.
2. He has pain.
3. It's hot outside.
4. He is very active.
5. He is traveling.

Your child may not want to drink a lot of fluids at these times, but he still needs them. You may have to push your child to drink more clear fluids. Try ice chips, popsicles, jello, milk or soup as well as water, juices and sodas.

Use the chart below to figure out how much fluid your child needs during these special times.

A baby who is breastfeeding or on infant formula only needs extra fluids during special times. At these times, you need to encourage him to take all of his breastmilk or formula, unless he is vomiting.



Drinking a big glass of juice on a hot day.

Amount of Clear Fluids Your Child Needs Each Day During Special Times

Child's Weight	Number of 8 oz. Cups Per Day
10 lb	2 cups
15 lb	3 cups
20 lb	4 cups
25 lb	5 cups
30 lb	5-6 cups
35 lb	6-7 cups
40 lb	7 cups
50 lb	8 cups
60 lb	9 cups
More than 60 lb	10 or more cups

Nutrition

Like everyone, children with sickle cell disease need to eat a well-balanced diet. Because their red blood cells break down faster, they need to have good food sources of protein, vitamins and minerals every day. They also need to have more calories in their diet to make new cells.

Vitamins

When your child is about a year old he may be given a vitamin supplement called folic acid, 1 mg a day. This can be crushed and mixed with milk, juice or food. Folic acid helps the body make new red blood cells. Some children don't need extra folic acid.

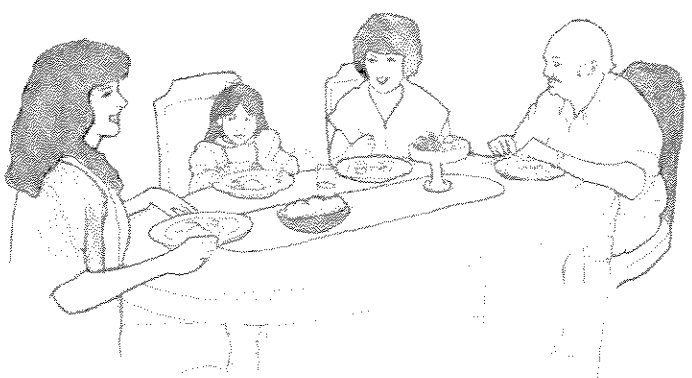
In addition to folic acid, some children with sickle cell disease may need to take other vitamins and minerals such as zinc, iron and vitamin E. Your doctor will prescribe these when needed.

You can also give your child a multiple vitamin without iron if you want, but it isn't necessary.

Height and weight

Your child may be smaller or thinner than his brothers and sisters, even with a good diet. That is because he has to use more energy to make new red blood cells. As he grows older, he will usually catch up.

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



Active Play

Sickle cell disease won't keep your child from doing most kinds of activity that he enjoys as long as he takes care of himself. Taking care of himself means:

1. Resting when he feels tired.
2. Drinking extra fluids when he is active.
3. Dressing for the weather.

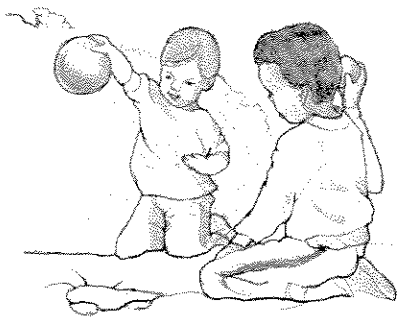
Tell your child to rest as often as he needs to. Then let him find his own level of activity and enjoy it.

Putting on a jacket when it's cold.

A few activities can sometimes cause problems related to sickle cell disease and should mostly be avoided.

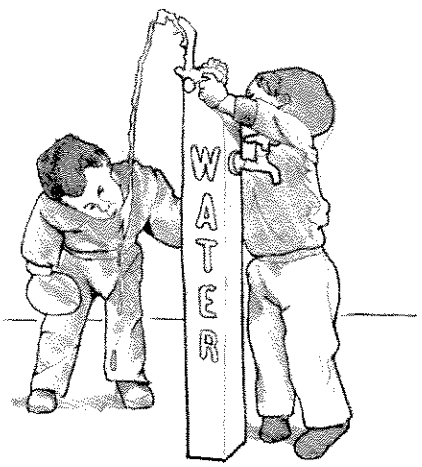
1. Ones that expose him to cold temperatures, such as swimming in cold water.
2. Ones at high altitudes, such as backpacking, hiking, or skiing.

If your child wants to do any of these, talk to your doctor first.



Playing with his friends.

All preschool and day care playground activities and most elementary PE class activities are fine for your child. If he is in a program where there is a leader or a teacher, tell them about your child's need to drink extra fluids and to rest when he is tired. They can help your child take good care of himself.



Taking time out to drink.

Taking a Trip

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

1. Fly only in a pressurized plane.
This should not pose a problem for most trips because almost all commercial planes are pressurized.

2. Drink plenty of fluids when traveling.
This is important when your child is:

- ❑ Flying in an airplane
- ❑ Riding in a car
- ❑ Visiting an area that is very dry
- ❑ Riding or walking at high altitudes

3. Be careful at high altitudes (above 5,000 feet).
If your child is riding in a car or walking above 5,000 feet, he needs to rest often and drink plenty of fluids. If he starts to feel sick, take him to a lower elevation.

Let your doctor know if you plan to take your child on a trip and ask him or her for a "Travel Letter."

See Appendix F for a sample Travel Letter.

Make sure that you take along the penicillin your child needs as well as any other medicines he is taking. Talk about your plans with your doctor to see if any other special care needs to be taken.

Travel Checklist

- ___ Talked to doctor
- ___ Travel Letter filled out by doctor
- ___ Filled prescription for penicillin
- ___ Made arrangements for other medicines
- ___ Packed extra fluids
- ___ Names and addresses of doctor or center to contact if needed.
- ___ Thermometer

See Appendix M for a sample Travel

Packing up your car with extra fluids for a long trip.

Fever, Pain and When to Get Help

4

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

This chapter will teach you how to find problems early so that you can get help. It will also give you ways to deal with the pain that sometimes is a part of sickle cell disease.

The following topics will be covered:

- **Fever**
- **Easing pain at home**
- **When to call the doctor**

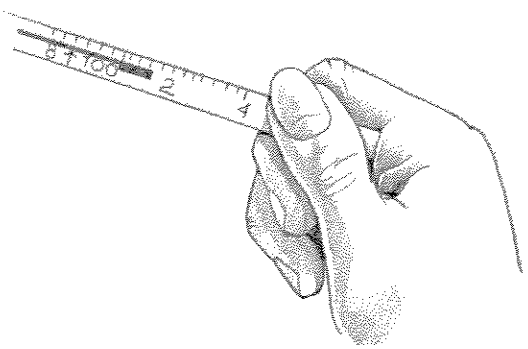
Fevers

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 her body is fighting an infection. Infections can be very serious in children with sickle cell disease. Catching an infection early can let you take actions to prevent it from getting worse.

If you think your child might have a fever, take her temperature. Your child has a fever if her temperature stays over:

- ❑ 100°-101°Rectal (in the rectum)
- ❑ 100° Oral (in the mouth)
- ❑ 99° Axillary (in the armpit)

The normal oral temperature is 98.6°. Rectal temperatures are about 1 degree hotter than oral temperatures. Armpit temperatures are about 1 degree cooler.



Time to call the doctor.

When to call the doctor

If your child's temperature is 101°F or higher, call your doctor or nurse right away. A child can have a fever of less than 101° with a cold. But a fever of 101° or more may mean a serious illness which the doctor needs to know about. (This is 38.4° for the less common Celsius markings).

When you talk to your doctor or nurse, describe where you took the temperature: in the mouth, rectum or armpit. Since the body's temperature is different in each of these places, it is important for the medical staff to know where you took it.

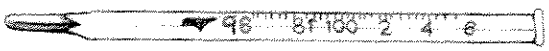
Before you give your child any medicine for a fever, call and speak with your doctor or nurse. If the fever is less than 101°, they will probably tell you to give your child **acetaminophen** (e.g., Tylenol, Tempra or Panadol). They will also tell you how much of this medicine to give, depending on your child's weight. Aspirin should not be given to children because it can cause a serious disease called Reye's Syndrome.

See Appendix G for list of brand names and doses for acetaminophen.

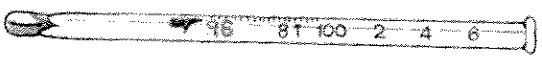
You don't need to take your child's temperature every day if your child is well. It is not needed and can be upsetting to your child.

Oral and rectal thermometers

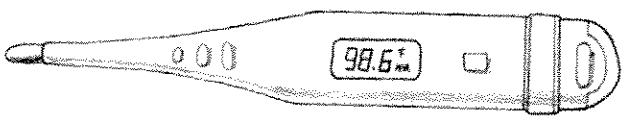
There are two main types of fever thermometers —oral and rectal. Oral and rectal thermometers look different. Oral thermometers have a long, thin end for fast, accurate readings. Rectal thermometers have a short, stubby end so they won't break in the rectum or cut the skin. An oral thermometer should never be used in the rectum.



Oral thermometer



Rectal thermometer



Digital thermometer

Digital thermometers are now available. These can be used in the mouth, rectum or armpit. Many parents like to use them because they are fast and easy to read.

All three kinds usually show temperature in Fahrenheit (F). It's best to use one that has markings in F, not C (Celsius).

If you have a thermometer with Celsius markings, use the chart in Appendix H to convert the reading to F.

Oral and rectal thermometers have an arrow pointing to 98.6° F, the normal oral temperature. However, the normal temperature in the rectum is 1 degree higher (99.6° F).

If you don't have a thermometer, tell your doctor or nurse. They may have a sample to give you or can tell you the best kind to buy.

Ways to take a temperature

There are three ways to take your child's temperature:

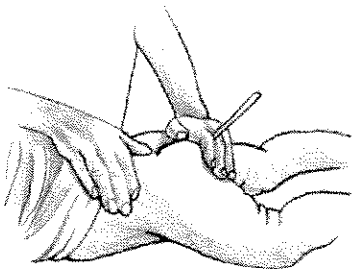
1. In the rectum, using a rectal thermometer. Do not use an oral thermometer.
2. Under the armpit, using any kind of thermometer.
3. In the mouth, using an oral or digital thermometer. Do not use a rectal thermometer.

When to take a rectal temperature

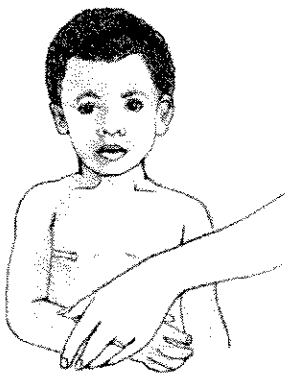
Take a rectal temperature with a baby or a young child who can't hold a thermometer in her mouth for 2-3 minutes or with a child who is congested and can't breathe through her nose. Use K-Y jelly on the thermometer, not vaseline, to make it go in smoothly.

When to take an oral temperature

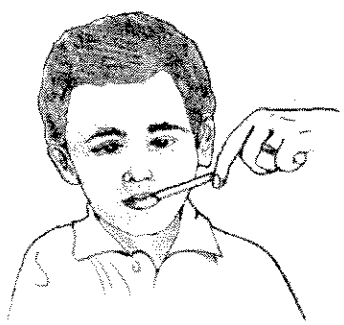
When your child is old enough to hold a thermometer in her mouth and keep her mouth tightly closed, switch to an oral temperature. Place the thermometer under the tongue.



Rectal



Armpit



Oral

Taking a temperature

Before you take your child's temperature with an oral or rectal thermometer, shake the thermometer to get the silver or colored bar to below 95°. Then place it in your child's mouth, armpit or rectum for 2-3 minutes.

You don't need to shake a digital thermometer. Just place it in your child's mouth, armpit or rectum for 1 minute.

- ❑ If your child is sick, take her temperature early in the morning and late in the afternoon. If your child seems very sick, check her temperature more often.
- ❑ When your child is sick for several days, try to take her temperature at the same time each day.
- ❑ Don't give your child anything hot or cold to drink or eat for a half hour before taking an oral temperature. Food or drink can change the reading by warming or cooling the child's mouth.
- ❑ Stay with your child to be sure she stays still while her temperature is being taken.

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.

Reading the thermometer

To read the temperature, take the thermometer out of your child's mouth, armpit or rectum and follow these steps:

1. Turn the thermometer until you can see the colored bar.

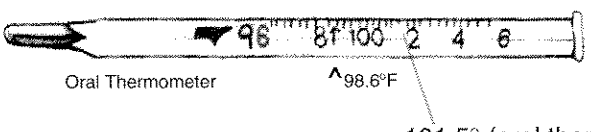
2. Line up the end of the colored bar with the degree mark.

3. Read the mark. Each mark usually stands for two-tenths (2/10ths) of a degree.

No matter what kind of thermometer you use, your child's temperature is found by reading the amount written at the end of the colored bar.

After reading the temperature, wash the thermometer with cold water and soap. Then wipe it with alcohol and put it back into its case.

REMEMBER:
If your child's temperature is 101° or higher, call your doctor or nurse right away. Make sure to say where you took the temperature.



Oral Thermometer

Gently massaging his sore leg.

5. 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.

6. Tylenol (acetaminophen)

Tylenol can provide a lot of relief for the pain caused by sickle cell disease. Make sure to give your child the right dose for her weight. Never give your child aspirin, unless ordered by your doctor, because it can cause a serious disease called Reye's Syndrome.

Helpful hints

Your child will feel less pain if she is involved in something she enjoys. Some children like to be alone when they feel pain. Others want to be busy. Learn what works best for your child. If she likes to be busy, keep her busy with games, stories and other fun things. Read her books, talk to her or let her watch movies or TV.

See Appendix G for list of brand names and doses for acetominophen.

Pain that could be serious

Call your doctor right away if your child has any of these symptoms:

1. **Chest pain or shortness of breath**
2. **Abdominal pain**
3. **Pain along with fever or swelling and redness**
4. **Pain which isn't relieved by home remedies**
5. **Severe headache**

If your child has to go to the hospital to deal with 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.

Easing Pain at Home

Your child may have pain at times from her sickle cell disease. Usually, the pain is mild enough to treat at home. Infants and toddlers may show pain by crying, refusing to walk, 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

1. More fluids

Extra fluids can help keep the sickle cells from clogging up small blood vessels. Since this is a major cause of pain, extra fluids can do a lot to ease the pain. Give your child up to double the amount of fluids she usually drinks. (See the chart on page 23 for more information.)

Playing quietly and drinking a lot of fluids makes her feel better.

2. Quiet play

Cutting back on physical activity can be helpful. Complete bedrest may not be needed, just less active play. Find things for your child to do quietly inside for awhile. See if quiet play will help her feel better.

3. Warm baths

Let your child soak in a warm bath for awhile. When it cools off, you can add more hot water or she can get out. Sometimes it feels good to do mild exercises in the warm water.

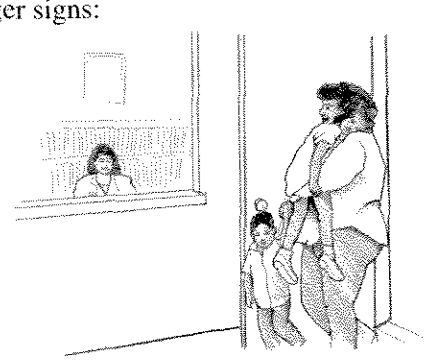
4. Heating pad or warm, moist towels

Apply either 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, then wring them out. You can apply these as often as it helps.

Taking a warm bath.

Call to be Seen Right Away

Call your doctor or nurse **immediately** to find out where you should bring your child to be seen if your child has any one of these danger signs:



FEVER	101° F or higher
HEAD	Severe headache or dizziness
CHEST	Pain or trouble breathing
STOMACH	Severe pain or swelling
COLOR	Very pale
PENIS	Painful erection
BEHAVIOR	Seizures
	Weakness or paralysis (can't move arm or leg)
	Can't wake up

If you think something is wrong, call your doctor. Trust your own judgment.

If you can't reach your doctor, go to the emergency room. These symptoms could be a sign of serious problems that need medical attention right away.

Call for Advice

Call your doctor or nurse for advice if your child does or has any of these problems:

STOMACH	Vomits more than once
	Has diarrhea more than once
COLOR	Jaundiced (eyes or skin look yellow)
ARMS, LEGS OR BACK	Pain with no other symptoms
CHEST	Coughs without fever or chest pain
NOSE	Runny or stuffed nose
BEHAVIOR	Isn't acting right
	Refuses to take penicillin
	Is less active than usual
	Refuses to eat or drink

Again, if you think something is wrong or your child just doesn't look right, call your doctor.

Many times, you can handle problems at home after talking with your doctor or nurse. You may be asked to call in each day for several days to be sure your child is getting better.

A copy of this list of danger signs is in Appendix A. Cut it out, write down your doctor's telephone number, and post in a place where it can be found.

Medical Problems in Early Childhood - Birth to Five Years Old

5

This section describes the most common problems your child may have because of sickle cell disease. Signs and treatment approaches are included to help you understand what might happen to your child.

These are the most common problems:

- **General infections**
- **Pneumonias**
- **Problems with the spleen**
- **Anemia (low blood)**
- **Hand-foot Syndrome**
- **Pain**
- **Problems with the kidneys and urine**
- **Gallstones**
- **Delayed growth**
- **Less common problems - stroke and priapism**

If your child gets one of these problems, use this section to learn about it.

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

General Infections

Taking penicillin twice a day helps to keep your child well.

Checking for a fever.

Infection has always been the leading cause of death in young children with sickle cell disease. Serious infections can occur in these areas:

1. Blood
2. Lungs (pneumonia)
3. Spinal fluid (meningitis)
4. Kidneys or bladder (urinary tract infections, pyelonephritis).

These infections can be caused by many different kinds of bacteria and viruses.

Prevention

To help protect young children from infections, they are given penicillin pills twice a day. They also receive a pneumococcal vaccination shot. This greatly reduces the chance of infection by pneumococcus. This is the most common cause of serious illness or death in sickle cell disease. A major study of young children with sickle cell disease from all over the country was done recently. It showed that taking penicillin twice each day prevented these infections and did not cause harmful side effects.

Signs

The most important sign of infection is fever. If your child has a fever over 101° F, take her to the doctor right away. Call first so your child can be seen more quickly.

Treatment

When you get to the doctor's office or clinic, the doctor will examine your child, take samples of her blood and urine, and do a chest x-ray. If her fever is very high, if she is very young or if she looks very sick, a spinal tap to check the spinal fluid will also be done. The samples of blood, urine and spinal fluid will be sent to the laboratory to find out what is causing the infection so that your child will get the proper treatment.

While waiting for the lab results, your doctor may decide to put your child into the hospital so that she can get antibiotics through an IV. Your child will have to stay in the hospital on the antibiotics until the final lab test results are ready. This can take up to three days.

If the tests don't show a bacterial germ, it means the fever may have been caused by a virus. If she doesn't have a fever and seems well, she will be able to go home.

If the tests do show that a bacterial germ is causing the infection, your child will have to stay on the IV medicine for up to two more weeks. Sometimes the doctor will also tell you to give your child antibiotics at home for another week or so. Then your child will need to be checked to make sure she is well. It is important for you to give your child all the medicine the doctor ordered, even if she seems well. This will help prevent the infection from coming back.

Pneumonia

Pneumonia (lung infection) is very common in children and adults with sickle cell disease. In fact, children with sickle cell disease are about 300 times more likely to catch pneumonia than other children. This is one of the most dangerous infections your child can get. Prompt treatment is critical.

Signs

Signs of pneumonia include some or all of these:

1. Fever
2. Frequent coughing
3. Rapid breathing
4. Shortness of breath
5. Difficulty breathing or "grunting"
6. Tiredness
7. Chest pain

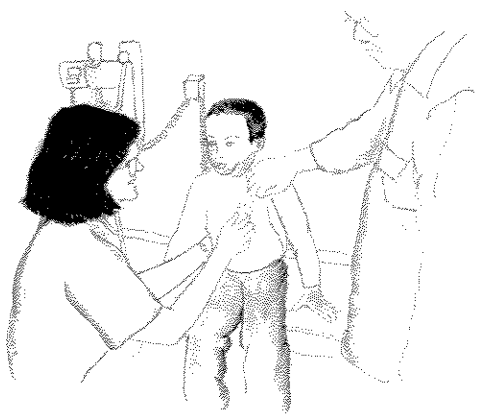
If your child has a cough that won't go away or has a cold and seems more tired than usual, have your doctor check for pneumonia. In a child with sickle cell disease, infections which might cause only a cold in another child can turn into pneumonia. The doctor will order a blood culture for infection and a chest x-ray. Pneumonia can usually be diagnosed from the x-ray.

Treatment

The treatment for pneumonia is the same as the treatment for other infections. Your child will be given IV antibiotics in the hospital for several days.

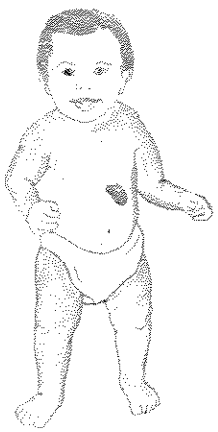
If the pneumonia is severe, your child's blood oxygen level may fall. This causes sickling in the lung known as "lung infarction." This condition can cause severe chest pain and make it hard for her to breathe. If her blood oxygen level gets too low, she may need to get extra oxygen through a mask or a tube under the nose. A transfusion may also be needed.

A few children get pneumonia often. They may have one infection after another.

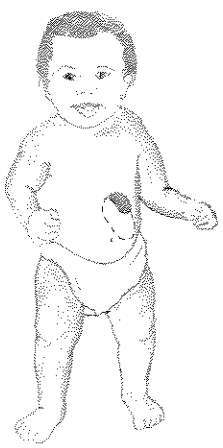


Listening to your child's chest for a sign of pneumonia.

Problems With the Spleen



A normal spleen.



An enlarged 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 why it is important for your child to see a doctor often is to have the size of the spleen checked when she is healthy. Then, if your child becomes ill, the doctor will know whether the spleen is larger than usual for her.

Different types of sickle cell disease affect the spleen differently. In most children with SS disease, the spleen stays enlarged for several years. By age six, their spleen becomes small again due to scarring from sickling. Children with SC disease and S beta thal 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, the scarring keeps the spleen from working well. This is one of the reasons that children with sickle cell disease get so many infections.

Splenic sequestration

In some children, the spleen may become larger very quickly and begin to trap lots of blood cells. This "bleeding into the spleen" can cause the blood count to drop quickly. This can lead to heart failure and death if not treated promptly with a blood transfusion. Rapid spleen enlargement with dropping blood count is known as a "splenic sequestration crisis."

Signs

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

Treatment

If your child has one splenic sequestration, she will be more likely to have another. If your child is at least two years old, her spleen may be taken out so that this won't happen again. If your child is under two, she will probably get blood transfusions to protect her until she is old enough to have her spleen taken out.

Your child does not need her spleen to live. However, if your child has her spleen taken out, she should keep taking penicillin twice a day until she is an adult to help prevent infections.

Anemia (Low Blood)

People who have sickle cell disease have fewer red blood cells than normal. Since red blood cells carry oxygen to the muscles, they often become tired more quickly than people with normal blood counts. In general, people with SS disease have the most severe anemia.

There are times when your child's blood 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 quicker 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.

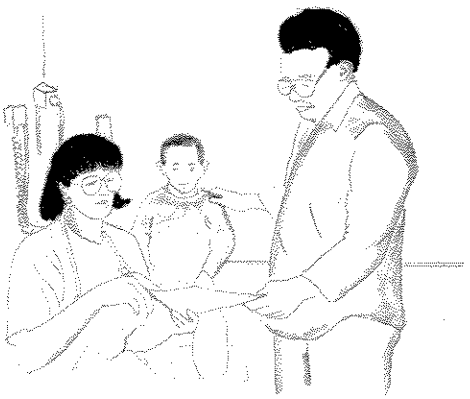
Signs

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

1. More tiredness
2. Pale color
3. Loss of appetite
4. Yellow eyes or skin
5. Dark urine

Treatment

If your child's blood count falls very low, a blood transfusion may be needed. An extremely low blood 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 doctor will usually check it daily when your child is in the hospital.



Going over the blood count results.

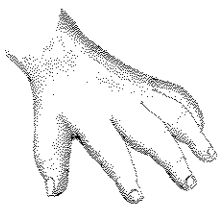
Hand-Foot Syndrome

About a third of children with sickle cell disease who are less than three years old may get a painful swelling of their hands and feet. This is known as the "hand-foot syndrome." It is caused by the sickle cells blocking the tiny blood vessels in the bones of their hands and feet.

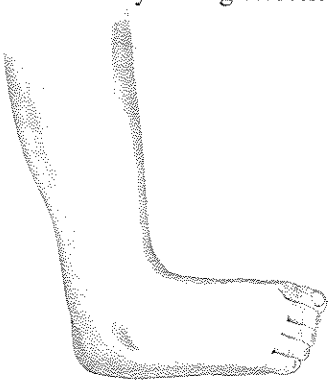
Treatment

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

If your child has a fever or if the pain is very severe, call your doctor. 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, the hand-foot syndrome will go away without any lasting effects.



A swollen hand.



A swollen foot.

Gallstones

About a third of children with sickle cell disease have gallstones by the age of seven. Gallstones are formed from the waste products of broken down red blood cells. These cells collect in the gallbladder and form thick sludge or stones. Gallstones are not harmful. But if they get stuck in the gallbladder duct, they can cause a serious infection. Emergency surgery is then needed to remove the gallbladder.

Signs and treatment

Eyes that are very yellow can be a sign that gallstones are stuck in the duct. Often, there is a warning before the stones get stuck. When the stones pass through the duct, they can cause pain in the right side of the abdomen. If the gallbladder is taken out after this warning, serious problems can be avoided.

Taking out the gallbladder doesn't cause severe problems. Without a gallbladder, people may have trouble eating a lot of fatty foods at one time.

Pain

Asking your child where it hurts so you can help.

Pain from sickle cell disease is not that common in very young children. Pain may also be caused by something other than the disease. All children have minor injuries and complain of aches and pains at one time or another. If the pain is very mild or goes away quickly, there is no reason why your child can't go on with her normal activities.

Treatment

Most pain that is caused by sickle cell disease is mild and can be treated at home. Your child will usually get relief from some "home remedies":

1. More fluids
2. Quiet play
3. Warm baths
4. Heating pad or warm, moist towels
5. Massage
6. Tylenol (acetaminophen)
7. Distraction (games, reading, TV)

See Chapter 4 for more details on ways to relieve pain.

If these don't help, your doctor may prescribe codeine. Codeine is a mild narcotic which eases pain. Sometimes it also causes constipation. If this happens, give your child extra fruits and other foods high in fiber.

If the pain is too severe to manage at home, your child may need to go to the hospital to receive stronger pain medicine (narcotics) and extra fluids through an IV.

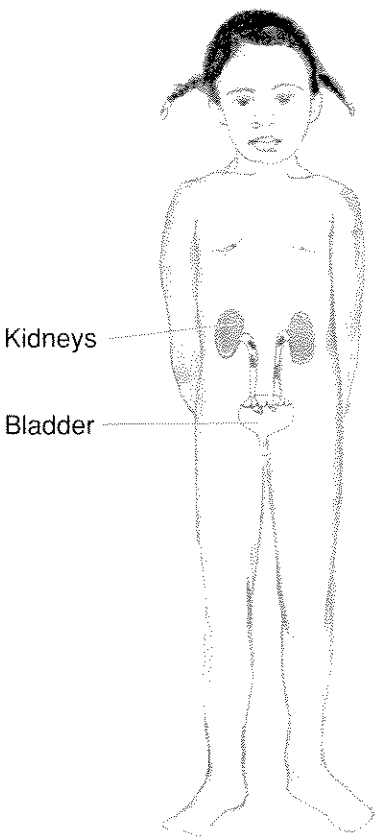
Certain kinds of pain may be a sign of something more serious. Call your doctor right away if your child has any of the following symptoms:

1. Chest pain or shortness of breath
2. Abdominal pain
3. Pain with a fever, swelling or redness
4. Severe headache
5. Pain which isn't relieved by home remedies

When your child feels mild pain in her arms or legs, treat her with sympathy, in a calm, matter-of-fact way. If you become nervous and overly concerned every time your child complains of pain, you may make it harder for her to deal with pain. Your child may become frightened, thinking that the pain means she will become very sick or die. Or she might learn to use pain complaints to get attention or special treats.

If you and your child are having trouble dealing with her pain, ask your doctor for help. He or she may suggest that you talk to a social worker or psychologist.

Problems With Kidneys and Urine



Dehydration

The kidneys help the body retain fluid. In sickle cell disease, the sickle cells damage the kidneys so that even young children have trouble holding onto their urine. Children with sickle cell disease drink more fluids and pass urine more often than other children. When a child becomes sick and drinks less than usual or loses fluid by vomiting, diarrhea or fever, she can get dehydrated.

Signs of dehydration:

1. Tiredness
2. Urinating much less (e.g., fewer wet diapers)
3. Dry, sticky mouth and lips
4. Sunken eyes or softspot (on a baby's head)

If you notice any of these signs, give your child extra fluids to drink.

Bedwetting

Frequent urination can also cause bedwetting. This is common in most children with sickle cell disease. Bedwetting is not caused by a psychological problem.

There are several ways you can help your child stop wetting her bed. You can limit the amount of fluids she drinks in the evening if she has had large amounts to drink during the day. It may also help to wake your child to urinate twice during the night. This could be just before you go to bed and one other time. When she is old enough, you can set an alarm clock to go off in the middle of the night so your child can go to the bathroom. Your doctor or nurse may have other helpful ideas.

Helping your child with a urine test.

Kidney and bladder infections

With sickle cell disease, infections in the kidneys and bladder can occur. If they are not treated promptly, the infection can move from the bladder up to the kidneys and cause kidney damage.

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

1. An increase in the number of times your child goes to the bathroom
2. Bedwetting after your child has stopped wetting the bed
3. Being unable to hold the urine
4. Foul smelling or cloudy urine
5. Fever
6. Burning and pain when urinating
7. Abdominal or back pain

If your child has a bladder infection, she will need to have her urine tested again from time to time to make sure that the infection has not returned. If she gets repeated bladder infections, she will need to take pills every day so that the infections won't come back.

Blood in the urine

Another problem caused by sickle cell disease is blood in the urine due to 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. Usually, the blood disappears from the urine within hours, but in some cases, the bleeding can go on for days and become a serious problem.

Always call your doctor right away if you notice blood in your child's urine or diaper so that tests can be done to find out what is causing the bleeding. In most kinds of kidney bleeding, it is very important to get plenty of fluids, sometimes through an IV in the hospital, and to rest in bed.

Delayed Growth

Most children with sickle cell disease grow normally when they are young babies. After their first birthday, they may start to grow more slowly. Throughout the rest of their childhood, some children with sickle cell disease are shorter and thinner than other children their age. Their height and weight is more like that of children several years younger. 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 that would be expected from the size of their parents.

If your child is concerned about being small, let her know that she will grow bigger like her friends. It will just take her a little longer.

Eating extra food

If your child is very thin and much smaller than normal for her age, she may gain weight and grow better if she eats extra food between meals or before bed at night. Your doctor may also want to give her special 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, sodas, and other "junk foods."

Your child may be smaller than her younger sister or brother who doesn't have sickle cell disease, but she'll catch up.

Less Common Problems — Strokes and Priapism

Strokes

Strokes are a very serious but fairly rare problem caused by sickle cell disease. They are caused by sickle cells blocking blood vessels in the brain. They happen to less than 1 in 20 children who have SS disease and even fewer children who have SC disease and S beta thal disease.

These can be a sign of a stroke:

1. Fainting
2. Sudden weakness of an arm or leg or the whole body
3. A difference in movement of one side of the face from the other
4. An eye that suddenly turns in or out
5. Severe headache
6. Seizure
7. Difficulty speaking

If you see any of these signs, call your doctor and bring your child to the hospital right away. The sooner the child is seen at the hospital, the better. Early treatment can keep a stroke from getting worse.

Children who have had one stroke are usually transfused with red blood cells every month for three to five years to prevent another stroke. At the end of that time, special studies are done to tell whether it is safe to stop the transfusions.

Priapism

A serious problem of sickle cell disease in boys can occur if sickle cells block the blood vessels in the penis. When this happens, the penis becomes erect, hard and painful. This problem is called "priapism," and it can occur at any age.

Sometimes the penis may become hard and painful for brief periods of time and then become soft again without any treatment. If this happens more than once, talk about it with your doctor. These short episodes may precede a longer one.

If the priapism doesn't go away in a few hours, your child needs be treated quickly with blood transfusions to stop the sickling so the blood can flow out of the penis. In rare cases, surgery must be done to flush out these blood vessels.

The Hospital

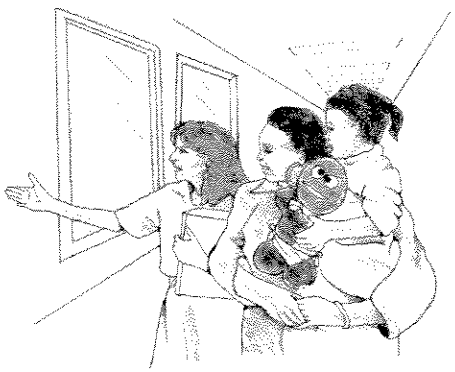
Hospital stays can be difficult times for your child and your family. Your child's medical team will do whatever they can to keep you and your child as comfortable and informed as possible. They want to answer your questions and help you in every way they can.

This section will describe what you can expect and things that can help with each part of a hospital stay. We have also included information about some of the procedures which may be done to your child in the hospital.

- ❑ **Getting ready**
- ❑ **The emergency room**
- ❑ **Getting admitted**
- ❑ **In the hospital**
- ❑ **Medicines and IV's**
- ❑ **Transfusions**
- ❑ **Surgery**
- ❑ **Going home**

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

Getting Ready For The Hospital

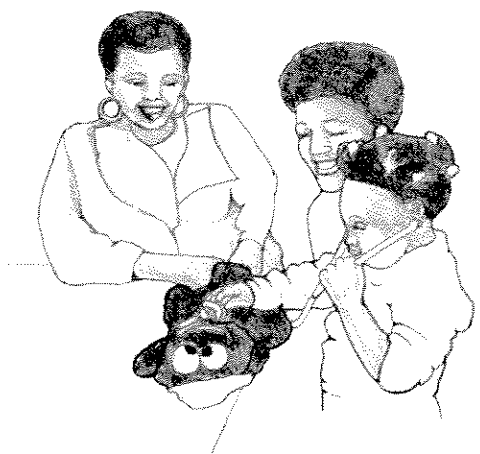


Touring the children's floor of the hospital.

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

Often, your hospital will have information which can help you and your child. They may have a brochure that explains the resources they have to help children and their families. Some hospitals have child life specialists who use play to help children with their feelings about surgery or being in the hospital. 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.



Dealing with fears through play.

You can use play, too, to help your child understand what may happen to him 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 and correct any wrong ideas he might have about what will be done to him 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 if he goes into the hospital. For children up to age six, try to have a parent spend the night. At night, your voice or touch can comfort your child. During the day, if a parent can't be there, the hosptial may have volunteers who can sit with your child for short visits to keep him busy.

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.

Planning for hospital stays

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 need help with these plans.

About the Emergency Room

About the emergency room (ER)

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 page 33, your doctor or nurse may tell you to go to the ER. Always try to call your doctor or nurse before going to the ER. If you can't reach your doctor, then call the ER so that they can get ready to see him.

Your doctor may give your child an ID card which describes his type of sickle cell disease. If you don't have one, ask your doctor. Don't forget to bring this card with you and show it to the ER staff. It is also wise to keep trying to call your doctor. You can ask the ER staff to call your doctor as well.

Getting your child admitted.

PATIENT NAME _____

BIRTHDATE _____ MEDICAL RECORD # _____

DIAGNOSIS _____ BASELINE HEMOGLOBIN _____

MEDICATIONS _____

_____

COMPLICATIONS _____

_____

_____

PLEASE CALL HEMATOLOGIST FOR ANY VISIT TO AN EMERGENCY ROOM OR ANY NEW COMPLICATION

Your child should have an ID card.

Starting with the emergency room

If your child is admitted through the emergency room, several people will help you with the admission process. First, you will usually meet someone who asks questions about you and your child. This is the Admission Clerk. It is very important that you let him or her know that your child has sickle cell disease and describe any medications your child is taking. Also, tell the Admission Clerk the name of your child's doctor. In a serious emergency, your child will be seen right away, and the paper work will be done later.

Next, your child's name will be called, and an ER Nurse will take you and your child to a treatment room. She or he 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 will then decide whether to admit your child into the hospital or to treat him there. Make sure that the ER staff have tried to reach your child's doctor so that they have information about your child's condition (for example, his usual hemoglobin level).

Think about going to the hospital in an emergency

- ❑ Do you know where the nearest emergency room is?
- ❑ Do you have someone to take your child?
- ❑ Do you have someone to watch your other children if you need to go?

Getting Admitted to the Hospital

If your child is admitted directly to the hospital on orders from his doctor's office or clinic, an Admission Clerk will register your child. The doctor will tell the hospital why your child is being admitted.

Getting settled

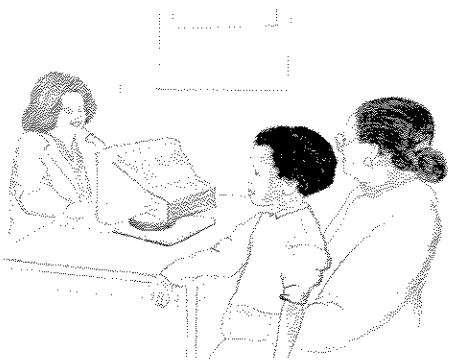
In the hospital, you will meet the Admitting Doctors. These doctors will examine your child and arrange for your child to go to the right floor or unit. When you get to your child's room, you will meet a nurse who will examine your child and help both of you prepare for his hospital stay.

Sharing your worries and concerns

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 to the hospital staff who are helping you. 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.

Learning the rules

In most hospitals, children share a room with other children. Because of this, each floor or unit has rules of conduct they wish 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 your child admitted.

In the Hospital

Getting checked by the doctor during one of your visits.

While your child is in the hospital, many people will be involved in caring for him. However, there are three key groups of people that you should get to know.

The doctor who is assigned to him

In addition to your regular doctor, your child may be assigned a doctor who will see him everyday and be in charge of his care. This doctor will usually check your child once each morning and again later if needed. He or she will give the orders for tests or medicines. Since this doctor will have the most contact with your child, it is important for you to get to know him or her.

The floor or unit nurses

Your child will also be assigned to a floor or unit nurse. This nurse will change when the shift changes (every eight or twelve hours). She or he is in charge of giving your child medicines and exams and doing other routine medical procedures. This nurse is also in charge of making sure all orders for tests and medicines are followed. If your child needs to do any special activities, the nurse will make sure he does them.

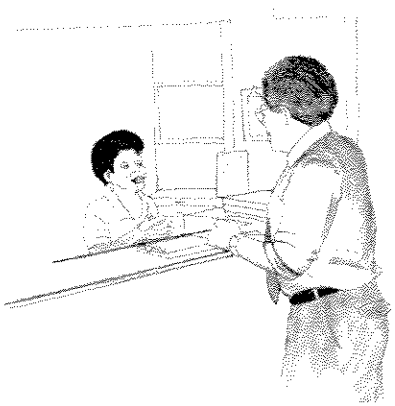
Keep in close contact with your child's nurses. A good way of doing this is to set up a schedule for phone calls and visits so that the nurse knows when to expect you. She or he can then give you an update on your child's progress and make sure that your child is available when you come. In most hospitals, parents can visit their child any time they want, so you can get reports from each shift nurse.

Other specialists

Your child may also see other specialists, depending on the reasons he is in the hospital. These people will visit your child throughout the day. Some will see your child only once. Others will see him many times, depending on how serious his problem is. It is important that you talk with these specialists, too.

Getting help from the nurse.

Lots of toys to play with.



Stopping at the nurses' station to share your concerns.

Activities for your child

There are often special activities in the hospital which can help make your child's stay more pleasant. These can include games and other preschool activities like playing with playdough, trucks and dolls 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.

If the hospital has a playroom, you won't need to bring too many things for your child to play with from home. This way your child won't lose a favorite toy. It is nice to pick out a special "hospital toy" that can go back and forth to the hospital if your child needs repeated visits. Having a toy from home can be calming at hard moments.

Dealing with behavior changes

In the hospital, you may notice some changes in the way your child behaves. He may not seem happy to see you when you visit, or he may cry a lot when you leave. You may notice him eating things in the hospital that he doesn't eat at home or the other way around. Or he may need diapers in the hospital even though he is toilet-trained at home. It can be less confusing for your child if you expect the same behavior at the hospital as you do at home. Of course, you can be a little more patient when he isn't feeling well.

Talk with the staff about any concerns you have about how your child is acting. 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 his "usual self" much more quickly when he leaves the hospital.

Talking about your child's needs or concerns

Your child may have things that he needs or is afraid of when he is in the hospital. If he is old enough to make requests or share his feelings, let him know that it is good to tell the staff so they can help him. If he is not able to talk to the staff himself, do it for him. Let the staff know about his needs or fears. They 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 him get the best care.

Medicines and IV's

Medicines

The most common medicines your child will receive in the hospital are:

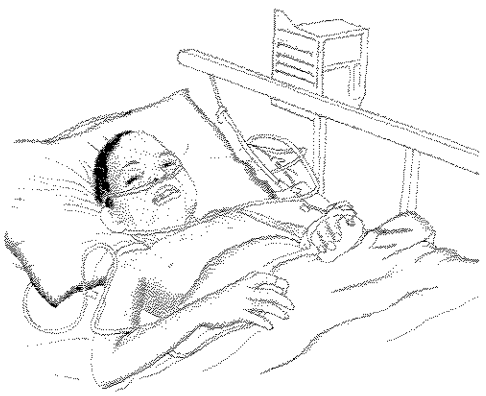
1. **Antibiotics** to fight infections
2. **Narcotics** to relieve pain
3. **Acetaminophen** to reduce fever and pain

Common Medicines
Antibiotics
Ceclor
Ceftriaxone
Penicillin
Ampicillin
Narcotics
Morphine
Demerol
Codeine
Acetaminophen
Tylenol
Tempra
Panadol

IV's

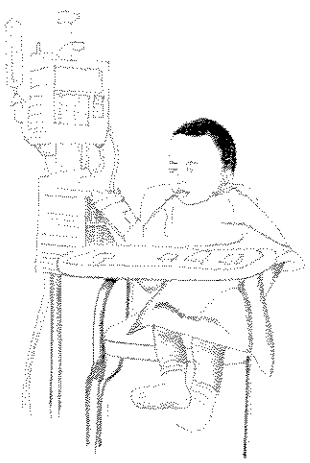
When your child is first admitted to the hospital, he may be given fluids by an IV. "IV" stands for "intravenous" which means "into the vein." The IV will be left in his vein so that he can get the fluids, food and medicine he needs. It may take a few trys to get the IV in the right place. But, once in place, your child won't have to get stuck again. If your child is in the hospital for surgery, he will need an IV before, during and after surgery to give him fluids.

The IV "pump" makes sure that the fluid and medicine go into the vein at the right speed.



Keeping an IV in.

Transfusions



Getting a simple transfusion.

Children with sickle cell disease sometimes need more or different blood. It is called a transfusion when they get this blood. While there are many reasons why your child may need blood, this is not the best treatment for all sickle cell complications. Some problems which may require a transfusion include:

1. **Severe anemia** (blood count drops to a very low level).

   This anemia can be caused by:

   a. **Splenic sequestration** — the spleen enlarges and traps a lot of blood cells in it.

   b. **Aplastic episode** — the body stops making new blood cells, such as when there is an infection like an ear infection, cold, chicken pox or pneumonia.

2. **Life threatening problems**, like a stroke or severe pneumonia.

3. **Prolonged, painful erection of the penis** (Priapism)

4. **Surgery**

Types of transfusions

There are two types of transfusions your child may get: simple and exchange. Simple transfusions are the most common. They involve giving your child a set amount of blood through an IV. The medical team will always try to tell you before they give your child blood. With serious illnesses, transfusion will be discussed as a possible treatment.

An exchange transfusion involves giving your child a set amount of blood at the same time that the same amount of blood is taken out of his body. This can be the best 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 for this procedure so he can be more closely watched.

Each time your child is transfused, he will be typed and cross-matched. This means that a sample of his blood will be taken to determine his blood type (such as "A" positive or "B" negative). Then the sample will be mixed with the blood he will receive to be sure the match is good.

Complications

Every effort will be made to give your child blood only when it is necessary because of possible complications. These are rare, but they can be serious. They include:

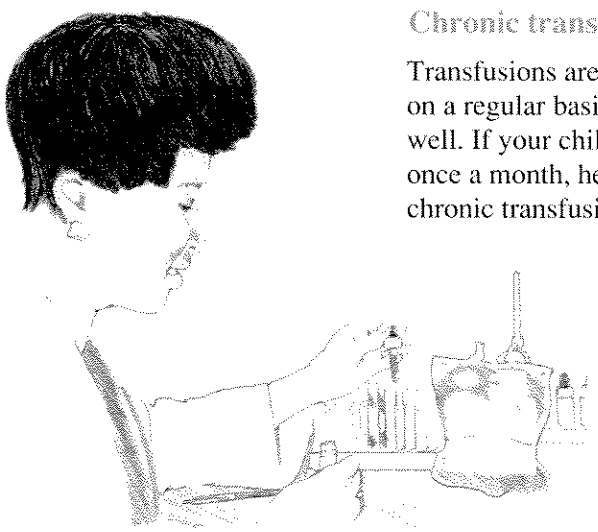
Infection
All blood products are thoroughly screened for AIDS, hepatitis and syphilis. But rarely, infections are transmitted by transfusions.

Allo-immunization
Your child may develop antibodies that destroy the blood he has been transfused with.

Allergic reactions
These can cause rashes, hives, itching, or, rarely, breathing problems.

Chronic transfusion programs

Transfusions are sometimes needed on a regular basis to keep your child well. If your child needs transfusions once a month, he is said to be on a chronic transfusion program.



Screening all blood products thoroughly.

On a chronic transfusion program, a child can get iron overload from the break down of the extra blood cells he receives. This extra iron may build up in his heart, liver or kidneys and damage these organs. He will need to take a special medicine (desferoximine) by needle under the skin at home every day to get rid of the extra iron. This medicine is given at night for eight hours using an infusion pump. He will also need to take the same medicine by IV when he comes to the hospital for transfusions. This treatment is called chelation. Your doctor will arrange for supplies and teach you how to use them.

Designated donors

Friends or family members who don't have sickle cell trait or sickle cell disease can donate blood for your child, if their blood matches. This is called being a "designated donor."

Designated donors can't be used for an emergency. It usually takes at least two days to get the blood from the place where it is donated to the hospital. If your child is going to have a planned operation or needs to be on a chronic transfusion program, ask your doctor or nurse for more information about this way of getting blood.

Surgery

Surgery is either elective or emergency. Elective surgery means that it can be planned ahead. Emergency surgery means that it needs to be done right away.

If your child has to have emergency surgery, he may be given blood to quickly reduce the amount of sickle hemoglobin in his body. With an operation that can be planned ahead (elective surgery), your child will probably be transfused once or twice during the weeks before to reduce the amount of sickle hemoglobin in his blood for the operation.

For elective surgery, your child will usually be admitted to the hosptial the day before so lab tests and other preparations can be done. The surgeon will talk with you about the surgery and possible problems. Another doctor will explain anesthesia, recovery time and any special care your child will need after the operation.

All surgery requires your consent. You must sign an "informed consent form" before the procedure can be done.

Going Home

The discharge meeting

When your child is ready to go home, the nurse, doctor or social worker will usually want to talk with you about important issues. Try to allow time to speak to them when your child is discharged. This is also a good time to discuss any questions or concerns you have about taking care of your child at home.

You will usually be given medicines for your child to take at home. Make sure that you understand how much to give him and how often. If you have questions when you are at home, call the doctor or nurse. **It is important that your child get the medicine as often and as long as the doctor says it is needed.**

Talking over your concerns with the social worker.

Sometimes children don't return to their old patterns when they come home. They may have some of these problems:

1. Trouble sleeping at night.
2. Wanting more attention.
3. Acting more unruly than before.
4. Wetting the bed or themselves when they had already stopped doing that at home.
5. Asking for a bottle when they didn't use one at home.

Keep in mind that these problems can be managed. If they don't last long or are mild, **consistency in your approach before and after the hospital stay is the key.** Your child needs to have the same expectations as any child his age who doesn't have sickle cell disease. For example, a four year old child should be sleeping in his own bed, not with you. However, your child who was hospitalized could 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 so severe as to disrupt your whole household, talk about them with your child's doctor, social worker or a member of the sickle cell team.

Taking Charge

7

Now that you know more about sickle cell disease, you can begin to take charge to help your child. Taking charge involves learning more about yourself and your child. It also means dealing effectively with those who are close to your child.

These are the areas in which you can take charge:

- ◆ Your first feelings
- ◆ Learning about the disease
- ◆ Helping your child at different ages
- ◆ Brothers and sisters
- ◆ Child care
- ◆ The medical staff
- ◆ Getting support
- ◆ Taking care of yourself

It's hard to be a parent, whether your child has a chronic disease or not. Use this chapter to help you learn better ways to help your child and your family.

Your First Feelings

Because sickle cell disease is a chronic, inherited disease, parents may feel a lot of different feelings when they find out that their baby has the disease. You may feel sad about the loss of the wished for "perfect child." You may feel guilty about the fact that the disease is caused by you and your partner's genes. 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.

Notice how you feel

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 deal with them. Instead, your feelings will control you. So try to notice what you are feeling.

Ask yourself:

- ❑ 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 still see your friends and family or 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. They may see things that you can't see. Also, just talking with others will help you learn more about how you feel.

Express your feelings

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 or other people you love.

- ❑ Share your feelings with your partner, a close friend or your clergy.
- ❑ Write about your feelings in a poem or a letter.
- ❑ Find a place where you can be alone and say whatever is on your mind.
- ❑ Get help from a social worker 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.

Learning About the 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 bad as you 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 his future. You can also teach other people how to help him.

Reading about sickle cell disease.

Helping Your Child at Different Ages

Getting to know your baby.

Soothing your baby.

Your infant

During their first year babies grow and change rapidly. Many babies are able to smile at others by two months. At three months you may notice your baby making happy sounds. By three months they begin to recognize their mothers and close family members. By four months babies may begin to be shy with strangers until they get to know them. At seven months they can respond playfully to other persons and by ten months they can wave bye-bye.

For the first months of your baby's life, he is protected from the disease. Most babies don't have sickle cell complications until they are two or three months old. 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 that there is little you can do to prevent some infections or pain.

A baby has few ways to let you know if something doesn't feel right. He may cry, be fussy, eat less or be less active. Give him 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 make him feel better.

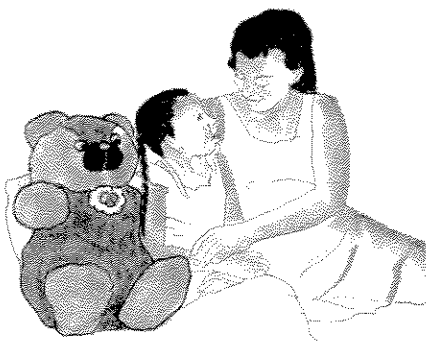
Your toddler

Toddlers are learning 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 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 him with someone else.

Your toddler needs to feel secure in your love. Play with him, comfort him, include him in family activities and let him learn things on his own.



Your child still needs your comfort.

Your 2-4 year old

Children from the ages of two to four want to be independent. They say "no," and want to do things themselves. Let your child begin to make decisions for himself. Even in the hospital, he can choose his own meals or TV programs or walk to the playroom when he feels better. At home, he can help you remember when to take penicillin and remind you to give him drinks. Use your judgment about things he can try for himself and avoid doing most things for him.

Even though your child is more independent, he still needs you. Comfort him, respond to his fears, answer his questions and help him learn more about his world.

Your 4-6 year old

Between the ages of four and six, children use imaginations to understand their world, including their disease. For example, a child may believe that the pain is a punishment for something he did. Or he may believe he caught sickle cell disease from something he ate. Pay attention to what your child may be saying about his illness. Clear up any wrong ideas. Don't worry if you have to go over the same ideas more than once.

Let your child ask any questions he may have about sickle cell disease. Answer these questions clearly, in words that suit your child's age. You may want to use stories, stuffed animals or puppets to help you explain what will happen to him.

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

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

Watching your child's play to learn her ideas about his disease.

Brothers and Sisters

Spending time with your other children.

Letting your older daughter help with the baby.

Whenever a new child comes into a family, each family member may have different reactions and feelings. For some brothers and sisters, the joy that is felt by their parents is shared by them. Others may be afraid that the new baby will threaten the share of love and affection they get. This feeling can be even stronger if the new child has special needs.

Your healthy child may feel afraid of becoming ill himself. He may be afraid he caused his brother's or sister's illness or feel angry about the extra attention the child with sickle cell disease is getting. 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

Many parents have found that by sharing the care of their child with sickle cell disease with their other children, these children have felt more loving and accepting of their younger brother or sister. 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.

Teach them about the disease

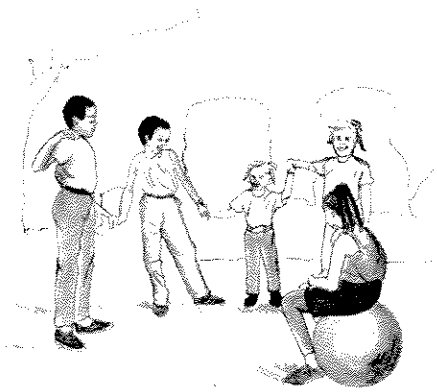
As you learn more about sickle cell disease, you can share what you learn with your other children. You can also let them ask the doctor or the medical staff questions, especially ones that you don't know how to answer. This will help them feel included.

Avoid "special treatment"

Brothers and sisters of children with special needs 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. 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.

Disciplining your child with sickle cell disease like you do your other children.

Child Care



Playing with other children at day care.

Child care 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 ill. It is often helpful to tell your child's caregiver the following about your child:

1. Treat your child like other children. Except for having sickle cell disease, he is just like other children!

2. Watch for signs of infections and respond. Go over a list of signs of infection and other problems. Make sure your caregiver knows how to contact you or another responsible adult who can pick up your child and take him to the doctor if needed. The caregiver should also have your doctor's name and phone number so they can call the doctor if they can't reach you. Be sure your caregiver has an Authorization for Medical Treatment Form from you so that he or she has permission to get emergency medical care for your child.

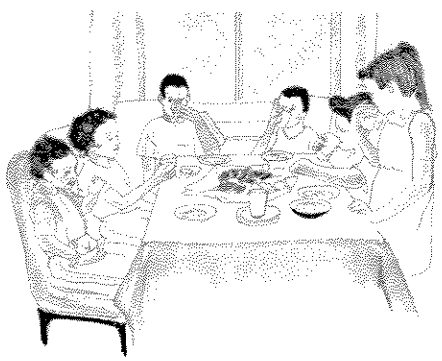
Going over what to do if your child needs help.

3. Take care of his special needs. While your child will have few extra needs, the ones he has are very important. These needs include drinking more fluids, resting when he is 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 written materials, like this handbook, which will provide them with more information. If they have more questions, you can refer them to your child's doctor or medical staff.

Sharing all of this information with these 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 Appendix I for a sample Authorization for Medical Treatment Form to give to all caregivers, including babysitters.



Drinking more fluids at snack time.

Getting 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, church members and community groups are a great source of love and support. Help them to 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 what they can do to be helpful.

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 or to introduce you to another family with a child with sickle cell disease.

See Appendix J for a list of 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 strange ideas about sickle cell disease. With the help of your medical staff and your own knowledge of the disease, you can teach others to provide the support both you and your child need.

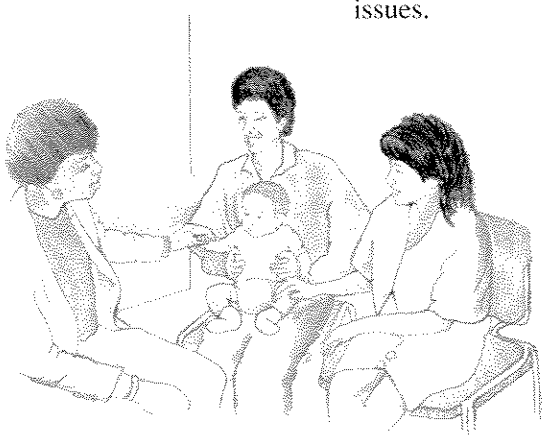
Getting support from another mother who understands.

The Medical Staff

You will probably be the first to notice if something is wrong with your child. Because you observe and deal with your child's everyday needs, you know how he functions from day-to-day. When you share this information with the medical staff, they can better figure out how your child's illness is affecting him. Let the medical staff know about your concerns. Also, teach your child and others caring for him to do the same.

Sharing information goes both ways. The medical staff has a lot of knowledge and experience with sickle cell disease. They want to answer your questions and provide you with some of the support that you will need during the early years of your child's life.

You may find that health care providers sometimes disagree on how to handle a problem that your child is having. Differing opinions are common in any type of medical care. Talk to the staff if this happens so that you can understand the issues.



Working together with your doctor.

It is also okay for you to have a different opinion than your doctor. If this happens, share your thoughts and reasons as well as what you plan to do. If a conflict arises between you and your doctor, talk about it so that your child can receive the best care.

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

1. Make an appointment to talk to the doctor about your concerns or for more information about your child's health. When you call for the appointment, tell the receptionist it is for a consultation.

2. Write down your concerns as they come up. Use it as a reminder to bring with you to the appointment.

3. Get to know all of the medical staff and their roles in helping you and your child. Think about who could be most helpful to you in dealing with each specific problem.

4. Get tips from other parents about ways to express your concerns to the medical staff. New families can learn a lot from the experience of other families who have worked with the same staff.

Take Care of Yourself

It can be very 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. It won't help him, and it won't help you. Your needs are also important. Find time for your other children, for your partner and for your friends. Also, 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 chronic diseases. Most states have special programs to help pay medical costs, like Crippled Children's services or Medicaid. In California, the California Children's Services pays many of these costs. Talk to your doctor or a social worker about your concerns.

Be sure to get help if you need it. Reach out to medical staff, friends, family, clergy, 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 psychologist. Don't wait too long before you ask for help.

For more information on parenting classes or resources that might be helpful to you, check with your local church, YMCA, YWCA, community college, March of Dimes chapter, adult school, or city recreation program.

Planning Your Family

Pregnancy can be a time filled with joy and with stress. There are so many questions in your mind. Will the baby be a girl or boy? Short or tall? What will our child be like?

If there is a chance that you might have a child with a genetic disorder such as sickle cell disease, your questions may be more worrisome. Even though all pregnant women have a 3-5% chance of having a child with a birth defect or genetic disease, this chance is remote to most people.

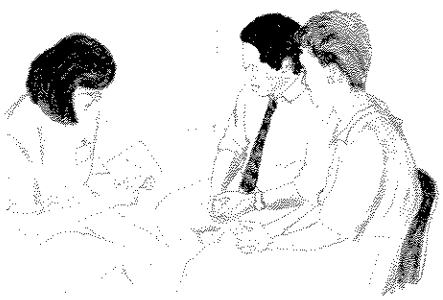
If you already have a child with sickle cell disease you may even be more concerned. You may remember what you felt after you heard that your other child had sickle cell disease. You may also be concerned about caring for another child with special needs.

This section will answer many questions about planning your family. It will cover:

- **Genetic counseling**
- **Testing your baby before it is born**
- **Chances of passing on sickle cell genes**

With this information, you will be able to make informed choices about what is best for you and your family.

Genetic Counseling



Getting information from a genetic counselor.

Genetic counseling can help

Because pregnancy brings with it these extra worries and questions, genetic counseling can be very helpful. Genetic counselors are experts in hereditary disorders and counseling. They give information and support to families with concerns like yours. They can help you figure out your chances of having future children with sickle cell disease and let you know about options for family planning and prenatal testing.

Even if you are already sure that you want to have another baby, it can be helpful to talk about your concerns with a genetic counselor **before you get pregnant**. In fact, many people have found that talking about their concerns before they are pregnant has made the pregnancy more joyous and less stressful.

Common questions

Whether you are pregnant or planning for the future, genetic counseling can address many common questions:

Q: What are my chances of having a child with sickle cell disease?

A: The chances depend on the hemoglobin type of both parents. If both parents have a hemoglobin trait and one of them is a sickle cell trait, there is a 1 in 4 or 25% chance in each pregnancy that the baby will have sickle cell disease.

Q: If I have a different partner than I had when I got pregnant with my child with sickle cell disease, am I still at risk?

A: Again, it depends on your new partner's hemoglobin type.

Q: How can my partner be tested?

A: Testing can be done by a simple blood test called hemoglobin electrophoresis with a complete blood count (CBC). This test can be ordered by your doctor.

Q: My partner was tested years ago and was told he did not have sickle cell trait. Does he need to be tested again?

A: Yes. Some sickle cell trait screening tests aren't very accurate. Also, many don't test for all the hemoglobin traits. Hemoglobin electrophoresis with a CBC gives the best results.

Q: If we are both carriers of sickle cell trait, is there any way to test our baby before it is born?

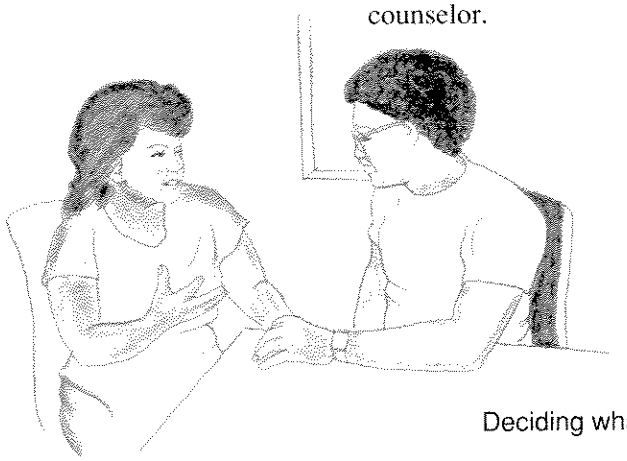
A: Yes. Prenatal testing can be done using one of two methods, chorionic villus sampling or amniocentesis.

Testing Your Baby Before it is Born

There are two types of tests which are usually used to test an unborn baby for sickle cell disease. These tests are:

1. **Chorionic Villus Sampling (CVS)**
   This test is done by putting a tiny tube through the vagina or abdomen into the uterus. A very small amount of the outer placenta is then gently removed. This test is usually done between the 9th and 12th week after the woman's last menstrual period. With this test, you can find out if your baby has sickle cell disease when you are only three months pregnant.
2. **Amniocentesis (amnio)**
   An amnio is done by putting a needle through the abdomen into the uterus. A small sample of amniotic fluid is then removed. This test is done between the 15th and 19th week of pregnancy.

Talk about these tests with your doctor or nurse and a genetic counselor.



Deciding what tests to take.

The test results

These tests can make you feel better if the results show that your unborn baby doesn't have sickle cell disease. But, if the results show that your unborn baby does have the disease, it can be very upsetting. You will then be faced with the decision of whether or not to continue the pregnancy. This decision can be very hard to make. A genetic counselor will give you the facts about the disease and what you can expect. Only you and your family can make the decision, and the counselor will support whatever decision you make.

Should you have the test?

You choose whether you want to have your unborn baby tested for sickle cell disease. A genetic counselor can help you look at whether testing would be helpful. For example, some families want to know whether their baby will have sickle cell disease so they can prepare for the birth. The decision is yours. Only you can decide what is best for you and your family.

Chances of Passing on Sickle Cell Genes

Sickle cell disease is inherited. Each parent gives their child one gene for hemoglobin. If either gene a child receives is for the usual hemoglobin A, the child will not have sickle cell disease.

To have a child with sickle cell disease, both the mother and father must pass a gene for sickle cell to their child. The child must receive the Hemoglobin S (sickle) gene from one parent and a Hemoglobin S, C, beta thal (B), or another different gene from the other parent. Both parents must have a **trait**.

For example, each time two people who have sickle cell trait get pregnant:

1. There is a 25% chance that their child will have sickle cell disease. (1 out of 4).
2. There is a 25% chance that their child will get only regular hemoglobin. (1 out of 4).
3. There is a 50% chance that their child will have sickle cell trait. (2 out of 4).

The chances are the same with each child

These chances are the same for every pregnancy with the same partner. This means that if you already have a child with sickle cell disease, your next child's chances of having sickle cell disease are the same—25%.

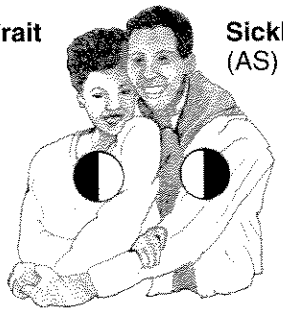
For example, if you have four children, you may not have one child with sickle cell disease. You may have two or three children with sickle cell disease, or none at all. It just depends on chance.

See Appendix K for charts to fill in your and your partner's hemoglobin types. Your genetic counselor or doctor can explain how this applies to your family.

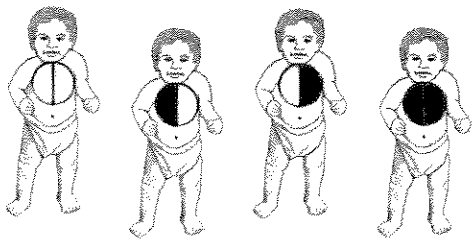
Chances of Having a Baby With Sickle Cell Disease

Both Parents Have Sickle Cell **Trait**

Sickle Cell Trait (AS)



Sickle Cell Trait (AS)



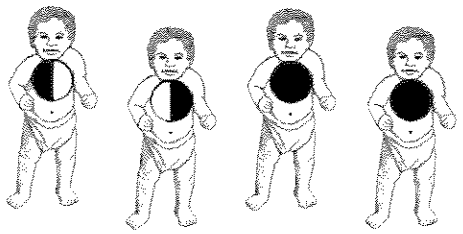
No Trait (AA) | Sickle Cell Trait (AS) | Sickle Cell Trait (AS) | Sickle Cell Disease (SS)

When both parents have sickle cell trait, they have a 25% chance (1 out of 4) of having a baby with sickle cell disease. Each time they get pregnant, they have the same chance.

One Parent Has Sickle Cell **Disease** and **One** Parent Has Sickle Cell **Trait**

Sickle Cell Disease (SS)

Sickle Cell Trait (AS)

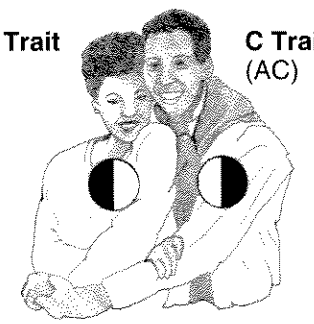


Sickle Cell Trait (AS) | Sickle Cell Trait (AS) | Sickle Cell Disease (SS) | Sickle Cell Disease (SS)

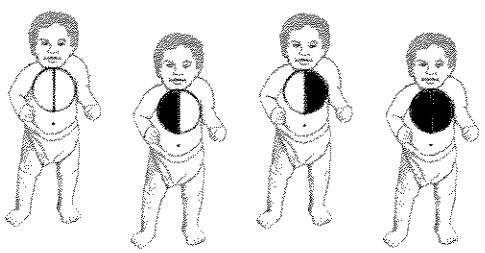
When one parent has sickle cell disease and the other has a trait, they have a 50% chance (or 1 out of 2) of having a baby with sickle cell disease. Each time they get pregnant, they have the same chance.

One Parent Has Sickle Cell **Trait** and **One** Parent Has **C Trait.**

Sickle Cell Trait (AS)



C Trait (AC)



No Trait | Sickle Cell | C Trait | Hemoglobin SC

Research and Treatment

9

There is no cure for sickle cell disease. Now, for the first time, treatments are being developed which could greatly improve the lives of people with sickle cell disease. These treatments are still being studied, so very few people can receive them. However, some of these treatments are now being used on small groups of patients.

Treatments are described in this section which hopefully will:

- **Increase fetal hemoglobin.**
- **Decrease sickle cell stickiness.**
- **Transplant bone marrow.**
- **Increase the water in sickle cells.**
- **Change the hemoglobin gene.**

Increasing Fetal Hemoglobin

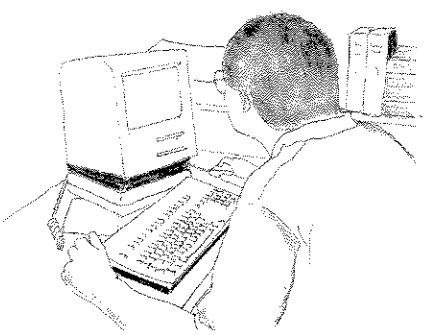
At birth, newborns with sickle cell disease do not have any symptoms of the disease. They are not anemic and do not have pain. This is because newborns have a very high level of fetal hemoglobin. Fetal hemoglobin does not sickle, so babies don't have problems. However, as the baby gets older, sickle hemoglobin replaces fetal hemoglobin.

Since this fact has been known for a number of years, research has looked for ways to increase the fetal hemoglobin level in people with sickle cell disease. In the last few years, experimental treatments have been found which increased fetal hemoglobin levels in a few people. Unfortunately, these treatments have had many side effects. The treatments must be taken every day, and the patient must be watched closely for side effects.

Newer treatments to increase fetal hemoglobin levels are now being developed. They include the drug hydroxyurea and a hormone called erythropoietin (EPO). This hormone is responsible for the body's production of red blood cells. It is found in all people. Drug companies are now able to make enough of this hormone to use as a treatment. There is hope that this hormone, which does not have some of the side effects of the other drugs, can correct the anemia in sickle cell disease and increase fetal hemoglobin levels.

Decreasing Sickle Cell Stickiness

Treatments are also being developed to decrease the stickiness of sickle red cells. Sickle cells are very sticky. It is this quality that makes them stick to the walls of the blood vessels and plug up the vessels. New research techniques can measure how sticky a person's cells are. In a test tube, they can also measure whether certain drugs are decreasing the stickiness.



Transplanting Bone Marrow

Bone marrow transplantation has been used to treat other blood diseases. It is a way to change the bone marrow, the place where blood is made.

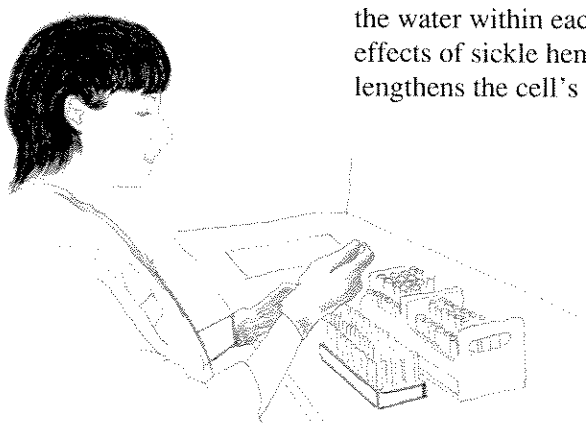
Briefly, this technique involves giving a patient a dose of radiation or drug therapy to destroy the body's bone marrow. After the bone marrow is destroyed, bone marrow is taken from a brother or sister and given to the patient. The new bone marrow then begins to grow in place of the patient's bone marrow and produce new cells. In many cases, the patient is cured.

This technique has worked with sickle cell disease. A few cases of people with sickle cell disease who also have leukemia have had their leukemia as well as their sickle cell disease cured following a bone marrow transplantation. A few people have also had the procedure just for sickle cell disease. However, this technique is very toxic and patients can die from it. Until the procedure becomes safer, it is not suggested for people just with sickle cell disease.

Increasing the Water in Sickle Cells

Another approach to reducing sickling is to increase the amount of water within sickle cells. Increasing the water within each cell dilutes the effects of sickle hemoglobin and lengthens the cell's life.

One way used to increase the water content is to make it easier for water to flow through the red cell membrane. This has been done using a drug called Cetiedil. Early research suggests that this drug may help reduce the problems of sickle cell disease.

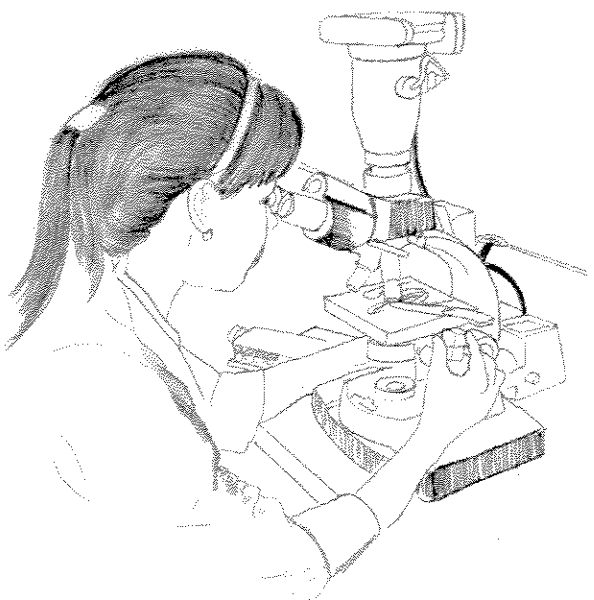


Changing Genes

In the future, genetic disorders may be fixed by putting a normal gene into the patient's tissue. Before this can be done in humans, we have to find a way to get the gene into the right place, where it will work correctly.

The whole area of changing genes raises many ethical and safety questions. Nonetheless, someday we may be able to get rid of sickle cell disease and many other genetic disorders by gene therapy.

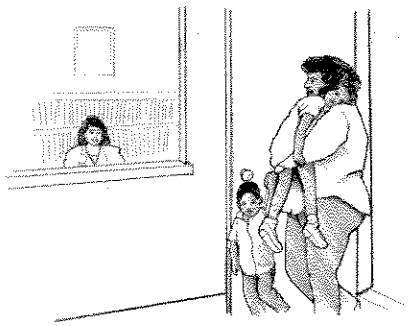
Other new treatments are being developed which may be available in the future. If you want more information about the ones described in this section or others that are not listed, ask your doctor or contact a sickle cell center.



Appendices

Call to be Seen Right Away

Call your doctor or nurse **immediately** to find out where you should bring your child to be seen if your child has any one of these danger signs:



FEVER	101° F or higher
HEAD	Severe headache or dizziness
CHEST	Pain or trouble breathing
STOMACH	Severe pain or swelling
COLOR	Very pale
PENIS	Painful erection
BEHAVIOR	Seizures
	Weakness or paralysis (can't move arm or leg)
	Can't wake up

If you think something is wrong, call your doctor. Trust your own judgment.

If you can't reach your doctor, go to the emergency room. These symptoms could be a sign of serious problems that need medical attention right away.

Call for Advice

Call your doctor or nurse for advice if your child does or has any of these problems:

STOMACH	Vomits more than once
	Has diarrhea more than once
COLOR	Jaundiced (eyes or skin look yellow)
ARMS, LEGS OR BACK	Pain with no other symptoms
CHEST	Coughs without fever or chest pain
NOSE	Runny or stuffed nose
BEHAVIOR	Isn't acting right
	Refuses to take penicillin
	Is less active than usual
	Refuses to eat or drink

Again, if you think something is wrong or your child just doesn't look right, call your doctor.

Many times, you can handle problems at home after talking with your doctor or nurse. You may be asked to call in each day for several days to be sure your child is getting better.

Cut this copy out and put it somewhere you can easily find it, such as on the refrigerator door or by your phone.

Comprehensive Sickle Cell Disease Care Plan:

Birth to 6 years of age

Evaluation	Interval
General Physical Exam	
Under 6 months	Once a month
6 months-1 year	Every 2 months
1-6 years	Every 3-4 months
Immunizations & TB Tests	See Appendix D
Comprehensive Social Worker Evaluation	
Interview	Every 2 years
Home visit	Once a year
School Assessment	Once a year
Genetic Counseling Services	
Family Studies	First visit
Counseling and Education	1-3 times a year
Hematology (red blood cell) Evaluation	Every 3 months to once a year
Liver Studies	Once a year
Gallbladder Evaluation	Every year or when needed
Renal (Kidney tests)	Once a year or when needed
Cardiac (Heart tests)	Every 2 years
Pulmonary (Lung tests)	When needed
Dental Evaluation	Once a year, starting at age 3
Psychological/Family Therapy Consultation	Once a year
Physical Therapy Assessment	When needed
Developmental Screen	Once a year or when needed
Formal Nutrition Assessment	Every 2 years or as needed

Note: This is the Care Plan recommended by Children's Hospital — Oakland Sickle Cell Center. If you have any questions, ask your doctor or sickle cell center.

Baby Shots & TB Test Schedule for Children With Sickle Cell Disease

Age	Type of shot
2 months	DTP, OPV (Polio), HIB
4 months	DTP, OPV, HIB
6 months	DTP, HIB, (Add OPV in areas where polio is more common)
9 months	Pneumococcal vaccine /option
12 months	TB skin test
15 months	MMR, HIB, Polio, DTP
18 months	DTP, OPV boosters
24 months	Pneumococcal vaccine booster
24 months	TB skin test
3 years	TB skin test
4 years	TB skin test
5 years	TB skin test
5 years*	DTP, OPV boosters (school shots)
5 years	Pneumococcal vaccine booster
Variable**	Hepatitis B vaccine
Variable**	Flu shot
Variable**	MMR - 2nd dose

Source: Children's Hospital Oakland, Sickle Cell Center

* Before entry to kindergarten, actual age of child will vary
**Talk to your doctor about the need for these shots.

Keep a record of the dates your child receives shots and TB tests.
Carry the record with you to show your doctor

SAMPLE

AUTHORIZATION FOR RELEASE OF INFORMATION

I, the parent of _____
(your child's name)

hereby authorize _____
(doctor or hospital)

to give any and all information in _____ record
(your child's name)

to _____
(requesting doctor)

Signed _____

Address _____

Witnessed _____

Date _____

NOTE: Your doctor may have his or her own form for you to use.

TRAVEL LETTER

Re: _____
MR# _____
DOB: _____

SAMPLE

To Whom it may concern: _____

is a _____ month old child with hemoglobin _____ disease who is followed at

_____ under the care of _____ .

In order to decrease the morbidity from their disease, our patients and their families are educated to recognize the symptoms and seek immediate treatment of the following emergencies seen commonly in sickle cell disease. We would appreciate your cooperation in the event that any of these patients come to you for treatment.

1. **Fever greater than 101°F:** Aggressive evaluation for the source of such a fever in the young child with sickle cell disease is very important. This evaluation should include CBC, reticulocyte count, blood culture, chest x-ray and urine culture. If the patient is younger than five years, he or she should be started on IV parenteral antibiotics (Cefamandol or Zincef) pending blood culture results. If the patient is over five years of age and nontoxic, oral antibiotics (Ceclor) can be used.

2. **Acute chest pain or difficulty breathing:** The patient should have a chest x-ray, CBC, reticulocyte count, and consider blood gas studies if there is any evidence of acute respiratory distress. If the patient is febrile, antibiotics should be started. In patients with severe chest pain hospitalization is mandatory.

3. **Acute pain not relieved by acetaminophen (Tylenol®), fluids, bedrest:** An aggressive evaluation for the source of the pain is mandatory. CBC, reticulocyte count, and other appropriate laboratory tests are also recommended.

4. **Marked lethargy or tiredness:** Physical examination documenting the size of the spleen, CBC, reticulocyte count and observation are required.

5. **Vomiting, dehydration:** Generally, these patients should be hydrated with saline containing solution. CBC and reticulocyte count should be done, and electrolytes are selectively indicated.

6. **Neurologic symptoms (seizures, weakness in the arms or legs, severe headaches, marked dizziness or visual changes):** The patient should undergo an extensive neurological examination. All patients with neurological symptoms should be admitted to the hospital. The possibility of a cerebrovascular accident should always be considered. Febrile patients demand a spinal tap. Exchange transfusion should be considered, and we should be notified of such a situation immediately.

Telephone number _____ .

Dr. _____ is available at telephone number _____ during regular business hours to provide further information about individual patients, to answer any questions, and to screen calls for appropriate physicians. During other times, or if you need to speak to

the on-call hematologist, the _____

hospital switchboard can page one of our physicians 24 hours a day at _____ .

Suggested Acetaminophen Dose Chart*

*Brand Names: *Tylenol®, Tempra®, Panadol®*

	Drops		Liquid	Chewable Tablets
Age	80mg Dropperful	0.8ml Dropperful	160mg 5ml	80mg tablets
0-3 months	1/2 dropper	0.4ml	-	-
4-11 months	1 dropper	0.8ml	1/2 teaspoon	1 tablet
12-23 months	1 1/2 droppers	1.2ml	3/4 teaspoon	1 1/2 tablets
2-3 years	2 droppers	1.6ml	1 teaspoon	2 tablets
4-5 years	-	-	1 1/2 teaspoons	3 tablets
6-8 years	-	-	2 teaspoons	4 tablets

Give your child the right dose 4 or 5 times a day or as ordered by your doctor. Don't give your child more than 5 doses in 24 hours.

Temperature Conversion Chart *Celsius (°C) to Farenheit (°F)*

	°C	°F
	36.0	96.8
	36.2	97.2
Normal armpit temperature	**36.4**	**97.5**
	36.6	97.9
	36.8	98.2
Normal temperature by mouth	**37.0**	**98.6**
	37.2	99.0
	37.4	99.3
Normal temperature by rectum	**37.6**	**99.7**
	37.8	100.0
	38.0	100.4
	38.2	100.8
Call your doctor if your child has a fever over 101°	**38.4**	**101.1**
	38.6	101.5
	38.8	101.8
	39.0	102.2
	39.2	102.6
	39.4	102.9
	39.6	103.3
	39.8	103.6
	40.0	104.0
	40.2	104.4
	40.4	104.8
	40.6	105.1
	40.8	105.4

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

SAMPLES

AUTHORIZATION FOR MEDICAL TREATMENT

We, the parents of _____
(your child's name)

authorize _____
(caregiver's name)

to take our child to _____
(your doctor's name or local hospital name)

for medical evaluation and treatment in the event we cannot be reached when _____
(your child's name)

has symptoms for which he/she must see a physician.

Parent signature _____

Parent signature _____

NOTE: Your doctor may have his or her own form for you to use.

Parent Support Groups

Alabama

Family Enrichment and Support Team
1601 12th Avenue South
Birmingham, AL 35205
(205) 933-8704
Rosemary Ashford, Program Coordinator *

Patient Assistance Organization
P.O.Box 1094
Mobile, AL 36601
(205) 432-0301
Rose Peterson, Executive Director *
Susan Andrews, Client Support Coordinator *

SC Patient/Parent Support Group
P.O.Box 9278, Gaston Avenue
Montgomery, AL 36108
(205) 263-9278
Willie T. Owens, Executive Director*

Family Support Group
P.O.Box 1079
Tuskegee Institute, AL 36088
(205) 826-1168
Rosa Storrs *

Arizona

Phoenix Metropolitan SC Support Group
7398 W. Topeka Drive
Glendale, AZ 85308
(602) 561-0630
Ellouise Coyne, Vice-President *

California

Alberta Sickle Cell Foundation San Joaquin Valley
4928 East Clinton Street, Suite 108
Fresno, CA 93727
(209) 252-6844
Wesley Forbes, Ph.D., Executive Director *

Greater Modesto Sickle Cell Foundation of Stanislaus County
P.O.Box 4753
Modesto, CA 95352
(209) 526-6889
Timothy A. Daniels, Director *

Parent Support Group
225 Dickenson Street, H918
San Diego, CA 92013
(619) 543-2851
Selma Johnson, RN *

Parents Helping Parents
535 Race Street, Suite 140
San Jose, CA 95126
(408) 288-5010
Florence M. Poyadue, RN, Director*

Sickle Cell Anemia Education & Information Center
P.O.Box 2641
National City, CA 92050
(619) 271-7105
Peola Early, Director *

Sickle Cell Disease Research Foundation of Los Angeles
4401 Crenshaw Blvd.
Los Angeles, CA 90043
(213) 299-3600
Peter Grams, MSW *

Sickle Cell Parent Group Children's Hospital Oakland
747 52nd Street
Oakland, CA 94609
(510) 428-3372
Fran Merriweather, MSW *

The Sca' Shan
P.O.Box 474
San Francisco, CA 94101
(415) 239-5162
Ozella Fuller, President *

National Association for Sickle Cell Disease, Inc.
3345 Wilshire Blvd., Suite 1106
Los Angeles, CA 90010-1880
(213) 736-5455
(800) 421-8453

Colorado

Family Support Group
2823 Fairfax
Denver, CO 80206
(303) 270-5977
Marva Houston, President, Board of Directors *

Connecticut

Adult Peer Support Group

Adolescent Support Group

Parent Support Group

114 Woodland St., #204
Hartford, CT 06105
(203) 548-5513
Rose Thornton, Social Worker Coordinator *

District of Columbia

SC Support Association
Howard University Center for Sickle Cell Disease
2121 Georgia Avenue, NW
Washington, DC 20059
(202) 806-7930
Angela Stephens, Group Facilitator*

Florida

Adult SC Support Group

Adolescent SC Support Group

Parent Support Group

655 West 8th Street
Jacksonville, FL 32209
(904) 350-6899, ext. 3842

SC Support Group

SC Parent Support Group

Dade County SC Foundation
794 NW 18th Street
Miami, FL 33136
(305) 547-6924/6872
Betty Bennett, MSW, Clinical Social Worker *

SC Support Group
SC Foundation of West Palm Beach, Inc.
P.O.Box 2402
West Palm Beach, FL 33402
(407) 833-3113/684-5176
Esther Sherman, Chairman, Program Services

Georgia

Parents' Support Group

Grady Memorial Hospital
Sickle Cell Center
80 Butler Street
Atlanta, GA 30335
(404) 589-4395
Yvonne P. Aldridge, Facilitator *

Teen Group
Grady Memorial Hospital
69 Butler Street
Atlanta, GA 30303
(404) 589-3661
Iris Buchanan, M.D., Hematologist/ Pediatrician *

SC Patient/Parent Group
1147 Maryland Circle, SW
Atlanta, GA 30310
(404) 589-3661
Berrutha Harper, President *

Parents' Group
Comprehensive Sickle Cell Ctr. Medical College of Georgia
Augusta, GA 30912
(404) 721-3091
Arthur Stewart, Research Assistant *

Illinois

Support Group SC Clinic
Univ. of Illinois at Chicago
840 South Wood
Room 1420 CSB, M/C787
Chicago, Il 60612
(312) 996-5680
Pam Moore, Data Coordinator *

Midwest SC Association
65 E. Wackler Dr., Suite 2200
Chicago, IL 60601
(312) 663-5700

Indiana

NW Indiana SC Parents' Auxiliary
4801 West Fifth Avenue
Gary, IN 4606
(219) 949-5310
Carrie Perkins, President *

Martin Center, Inc., SC Program Support Group
3549 N. College Avenue
Indianapolis, IN 46204
(317) 927-5156
Robert Treadwell, Counselor

Iowa

National SC Society of Des Moines, Iowa
1145 14th Place
Des Moines, IA 50314
(515) 243-3446
Charles Watkins, Director *

Kentucky

SC Self-Help Group
932 Darley Drive
Lexington, KY 40505
(606) 253-3765/243-3452
Patricia Crooks *

Citizens Concerned About SCD, Inc.
P.O. Box 11331
Louisville, KY 40211
(502) 637-4336
Kenneth M. Morton *

Louisiana

Patient / Family / Friend
2301 North Boulevard
Baton Rouge, LA 70806
(504) 346-8434
Patricia S. Williams, District Coordinator *

Parent Support Group
NW Louisiana SCA Foundation
2200 Milam Street, Room 9
Shreveport, LA 71103
(318) 226-8975
Deloris Baker, Resource Coordinator*

Maryland

Adult Support Group

Parent Support Group

NASCD, Maryland Chapter
828 East Baltimore Street
Baltimore, MD 21202
(301) 837-3053
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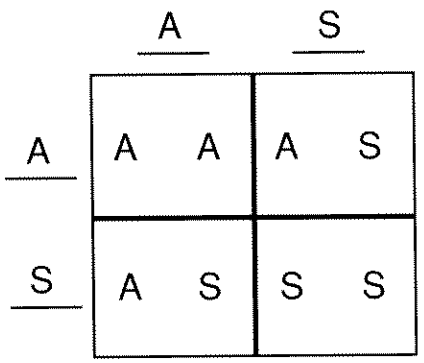
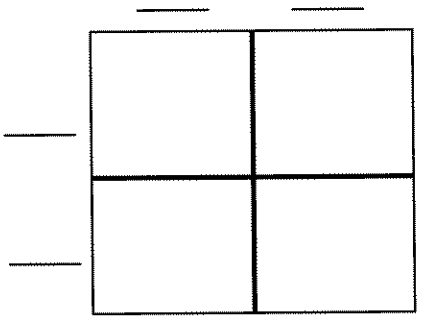
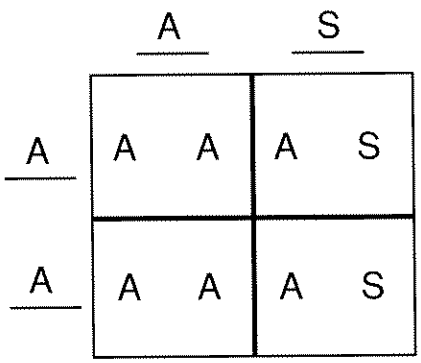
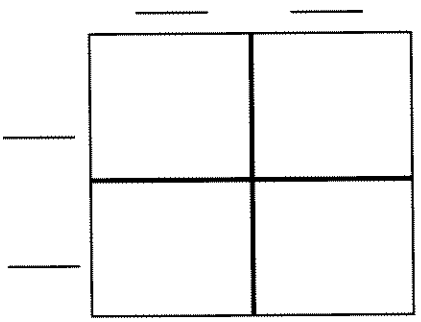
Diagrams of Inheritance

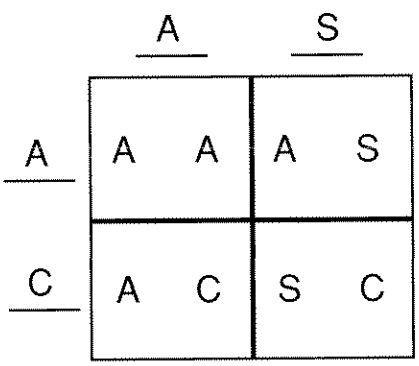
(Punnett Squares)

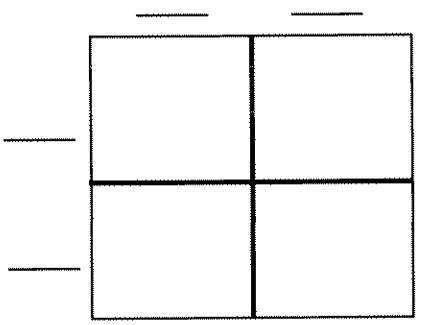












Health Care Providers

Audiologist

The person who tests your child's hearing if any problems come up.

Cardiologist (heart specialist)

The doctor who does EKGs and checks your child's heart.

Child Life Specialist

A person who uses play with your child to lower anxiety and promote understanding and healthy patterns of development during hospital stays. The child life specialist supervises programs for groups of children in the playroom and for the individual child at bedside.

Dentist

The doctor who helps your child keep his teeth healthy and clean.

Family Guidance Services

A service which provides counseling for families and children. Family Guidance therapists are trained to help children and families deal with a chronic illness. They also help with other concerns, such as school problems, behavior problems, and depression. They can include psychologists, psychiatrists, and social workers.

Family Practice Physician

A doctor who provides medical care for people of all ages, from babies to older adults. A family practice physician can give your child routine health care, referring to other specialists as needed.

Genetic Counselor

A person who explains how your child inherited sickle cell disease and your chances of passing it on to future children. By taking a detailed family and pregnancy history, this counselor gives you a chance to discuss your concerns about your child having sickle cell disease. They also can tell you if there are any other hereditary disorders that might be in your family. The genetic counselor teaches you and your child about the disease and how to deal with it.

Hematologist (blood specialist)

The doctor who treats your child's sickle cell disease.

Nephrologist (kidney specialist)

The doctor who treats your child's kidneys if they are damaged.

Nutritionist

The person who gives you advice on the best foods for your child and helps you find ways to get your child to eat what is good for him.

Occupational Therapist

The person who takes your child to "O.T." (Occupational Therapy) when he is in the hospital. These therapists help your child do activities that are useful as well as interesting. These activities can include dressing, cooking, sewing, computer games, etc. These activities can help your child focus on other things besides being sick or in pain.

Ophthalmologist (eye specialist)

The doctor who checks your child's eyes for any sickle cell damage.

Orthopedist (bone specialist)

The doctor who treats damaged bones or joints.

Pediatric Nurse Practitioner

A nurse who has special training to provide health care to children. He or she may see your child instead of a doctor at routine medical visits. A pediatric nurse practitioner can coordinate care between your pediatrician and hematologist and tries to make sure that things go smoothly if your child is in the hospital.

Pediatrician

A doctor who provides medical care for children. A pediatrician can give your child routine care, referring your child to other specialists as needed.

Physical Therapist

The person who takes your child to "P.T." (Physical Therapy) when he is in the hospital. These therapists also bring hot packs to the clinic if your child is being treated for pain or provide activities designed to help relieve pain, such as mild exercise or whirlpool (hot tub) treatments.

Psychiatrist, Psychologist (see Family Guidance Services)

Social Worker

The person who helps children and families cope better with sickle cell disease. A social worker can help you with billing and health insurance, getting medical care in and out of the hospital, and finding support for your emotional needs. You and your child can also talk to a social worker about any of your problems or feelings.

*Child Development Chart**

Activity	Language	Personal/Social
BIRTH TO 6 MONTHS		
1. Signals with hands, feet, other body movements his wants and needs. 2. Follows things in front of him. By 3 months, able to track objects. 3. Able to grasp toy but not hold. By 3 months, able to recognize and grasp objects and bring to mouth.	1. Coos and gurgles when playing and happy, cries when irritated or anxious. 2. Responds to sounds and imitates by making noises. 3. Quiet when satisfied.	1. Responds to noise and sights around him. 2. By first month, responds to a familiar face. 3. Smiles and gurgles appropriately. 5. Able to engage in play with own hands and fingers. 6. Able to play with toys and own clothes, etc.
6 MONTHS TO 12 MONTHS		
1. Able to sit up briefly. By 9 months, able to sit up as long as he wants. 2. Able to support own weight briefly when standing. 3. Can use chair to stand up. 4. Rolls over and crawls.	1. Jabbers when crying. 2. Can say "mama" and "dada." 3. Can say 1 or 2 other words.	1. Understands 3 or 4 words. 2. Begins to explore objects and her body (e.g., sucking fingers). 3. Responds playfully to other persons. 4. Learns through the senses (touch, taste, smell).
12 MONTHS TO 18 MONTHS		
1. Can stand alone for a short time. 2. Walks when hand is held. 3. Can pull up to stand and walk with the support of furniture. 4. Able to play by imitation.	1. Communicates by crude body language (e.g., shaking head for "yes" or "no"). 2. Can say 2 other words clearly besides "mama" and "dada."	1. Can follow some commands. 2. Can play with others by choice. 3. Cooperates in dressing.

*Within each age group the items are in sequential order.

Many items are taken from the Denver Developmental Screening Test.

Activity	Language	Personal/Social
18 MONTHS TO TWO YEARS		
1. Toddles by self with very little falling. 2. Seats self in small chair and climbs into adult chair. 3. Picks up and throws toys (balls) while standing. 4. Crawls up stairs by self. 5. Capable of toilet training. 6. Feeds self with some spilling.	1. Develops own baby talk (special words for objects, people, etc.). 2. Speaks clearly 4 to 10 words, including names.	1. Follows simple commands. 2. Recognizes and names some faces or pictures. 3. Says "thank you" or shows appreciation. 4. Points to or states wants. 5. Likes to play with cuddly toys.
TWO YEARS TO THREE-AND-ONE-HALF YEARS		
1. Runs well, doesn't fall. 2. Walks up and down stairs alone. 3. Walks backwards. 4. Engages in increased physical activity. 5. Feeds self well. 6. Able to dress self with little help. 7. Throws ball overhand. 8. Able to help put toys away. 9. Rides tricycle using pedals.	1. Names some drawings (i.e. horse, shoe, ball or dog). 2. Refers to self as "I." 3. Knows full name. 4. Uses pronouns. 5. Uses 3 and 4 word sentences. 6. Uses phrases.	1. Able to see others as helpers in getting what he wants. 2. Able to state toileting needs. 3. Learns by imitation. 4. Able to recognize differences between boys and girls. 5. Interested in fantasy play (make believe).
THREE-AND-ONE-HALF YEARS TO SIX YEARS		
1. Does broad jump. 2. Washes and dries face and hands, brushes teeth by self. 3. Hops on one foot. 4. Able to tell front of clothes from back. By age 5, able to dress and undress self. 5. Skips, alternating feet.	1. Names one or more colors correctly. 2. By age 4, names penny, nickel, dime. 3. Can make descriptive comments on pictures. 4. By age 6, defines words by their function (e.g., house is to live in).	1. Asks meaning of words. 2. Able to engage in play with others. 3. Begins to explore his living environment more. By age 6, his large increase in vocabulary allows him greater interaction with family and other persons. 4. By age 5, can do tasks by self. 5. By age 6, knows morning from nighttime. 6. By age 6, knows right from left. 7. By age 6, in effort to understand himself, becomes more like his parents. 8. By age 6 shows problem solving skills, may develop a hobby, and becomes more competitive with others.

Resources

Sickle Cell Disease Branch
National Heart, Lung, Blood Institute
National Institutes of Health
7550 Wisconsin Avenue
Bethesda, MD 20892
(301) 496-6931

National Center for Education in Maternal and Child Health
38th and R Streets, N.W.
Washington, D.C. 20057
(202) 625-8400

Publication: *Sickle Cell: A Selected Resource Bibliography*

National Association for Sickle Cell Disease, Inc.
3345 Wilshire Blvd., Suite 1106
Los Angeles, CA 90010-1880
(213) 736-5455 / (800) 421-8453

For information on Sickle Cell Associations and medical services;
Publication: *HELP!: A Guide to Sickle Cell Disease Programs and Services*,
United States, Bahamas, Puerto Rico and the Virgin Islands

Education Programs Associates
1 West Campbell Ave., Bldg. D
Campbell, CA 95008
(408) 374-3720

California residents may obtain a computer print-out of sickle cell and other genetic educational materials, sample copies, and newsletters.

For more information on child growth and development:

The First Three Years of Life
Burton White, Avon Books, New York, 1975

Toddlers and Parents
Berry T. Brazelton, Dell Publishing Company, New York, 1974

Denver Developmental Screening Test
(DDST) Ask your doctor.

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This list was developed by the Council of Regional Networks for Genetic Services (CORN). 1991.

Glossary

Amniocentesis (amnio)

A test done between the 15th and 19th weeks of pregnancy. It is used to find out if an unborn baby has sickle cell disease and certain other disorders. The test is done by putting a needle through the abdomen into the womb. A small amount of amniotic fluid is taken out and tested.

Anemia (low blood)

A condition in which there is less hemoglobin in the blood than usual so that the blood can't carry as much oxygen.

Aplastic Episode

An 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.

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.

Chorionic Villus Sampling (CVS)

A test done between the 9th and 11th weeks of pregnancy. It is used to find out if an unborn baby has sickle cell disease and certain other disorders. The test is done by putting a needle through the abdomen or a thin tube through the vagina into the womb. A small amount of the placenta is taken out and tested.

Chromosome

Structures containing the genes in the body. Most people have 46 chromosomes. Prenatal testing can be done to study an unborn baby's chromosomes.

Complete Blood Count (CBC)

A blood test which 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.

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 determine different types of sickle cell disease.

Fetal Hemoglobin

The most common type of hemoglobin in a fetus (unborn baby). It is later replaced by adult hemoglobin (although small amounts are produced throughout life).

Fetal Blood Sampling

A test done between the 19th and 21st weeks of pregnancy. With the help of ultrasound, a narrow needle is inserted through the abdomen and uterus into an artery in the umbilical cord. A small amount of blood is drawn for testing. In addition to testing chromosomes, other studies can be done on fetal blood.

Gene

The basic unit of heredity. Genes are passed on by a mother in the egg and by a father in the sperm. People have about 100,000 genes which determine many characteristics, including hemoglobin type.

Hand-Foot Syndrome

Painful swelling in hands and/or feet in young children with sickle cell disease. It is caused by blockage of tiny blood vessels with sickle cells.

Hemoglobin

The substance which carries oxygen in red blood cells. People with sickle cell disease often have lower hemoglobin levels.

Hemolytic Anemia

Low blood count due to increased breakdown of the red blood cells.

Hyperhemolytic Episode

A rapid breakdown of red blood cells which causes severe anemia in people with sickle cell disease. It is associated with a quick increase in the size of the spleen.

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 coloring material from red blood cell breakdown.

Leg Ulcer

A breakage in the skin that begins as a small sore on the lower leg—above, over and around the ankle. It can be caused by injury and decreased blood flow.

Malaria

A disease carried by a certain type of mosquito in tropical areas of the world. Malaria causes fever, serious illness, and often death. People who have sickle cell trait are better able to survive malaria than those who only have Hemoglobin A.

Prenatal Diagnosis

Testing for genetic disorders and some birth defects which is usually done before the 20th week of pregnancy. A variety of techniques, such as amniocentesis, Chorionic Villus Sampling, ultrasound and fetal blood sampling, are used.

Priapism

A persistent, painful, unwanted erection of the penis caused by sickling.

Prophylactic Penicillin

Penicillin which is given in order to reduce the number and severity of infections in children with sickle cell disease.

Sickle Cell Anemia

Another name for SS disease, the most common type of sickle cell disease.

Sickle Cell Disease

A term which refers to all types of sickle hemoglobin disorders, such as SS disease, SC disease and S beta thal 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 from the blood. This organ does not work well in sickle cell disease. It can trap blood and become enlarged.

Splenic Sequestration

One type of episode that can occur in patients with sickle cell disease which can be life threatening. It is caused by blood being trapped in the spleen.

Trait (see Carrier)

Transfusion

Blood given to someone because of a very low blood count, to prepare for surgery or to treat certain complications of sickle cell disease.

Ultrasound in Pregnancy

An instrument which uses sound waves to show a picture of an unborn baby on a television screen. Measurements can be used to figure out the baby's due date. Sonagram can also show the baby's position, the number of fetuses, and can help check the baby's growth. More detailed studies can detect certain types of birth defects. Ultrasound is also used to assist in amnio, CVS and fetal blood sampling tests.

Vaso-Occlusive Episode

Occurs when sickle cells block the flow of blood. This causes pain and, if severe, tissue damage.

White Blood Cells

Infection-fighting cells in the blood