

A Parent's Guide to Sickle Cell Disease



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Chapter 1:

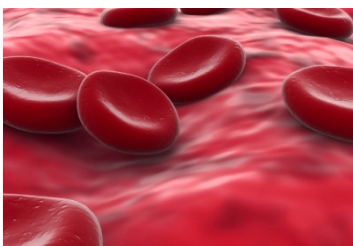
About Sickle Cell Disease

Chapter 1

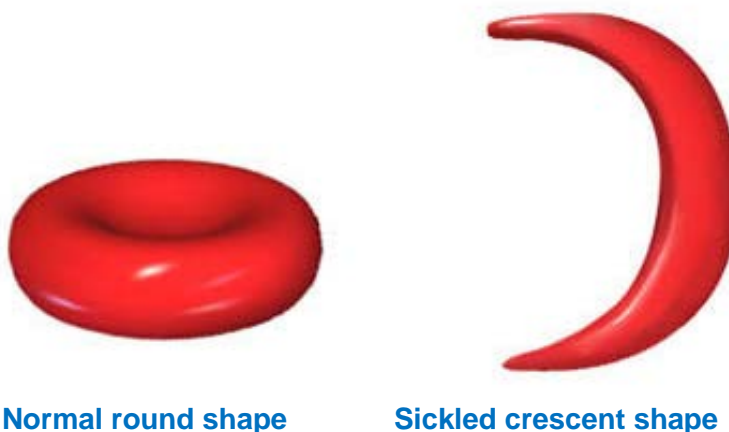
About Sickle Cell Disease

Sickle Cell Disease

Sickle Cell Disease is an inherited disorder that affects the hemoglobin, a molecule in red blood cells, which helps to deliver oxygen throughout the body. These are normal red blood cells with normal hemoglobin (AA hemoglobin). These cells appear round and are very flexible. These normal cells live for about 120 days.



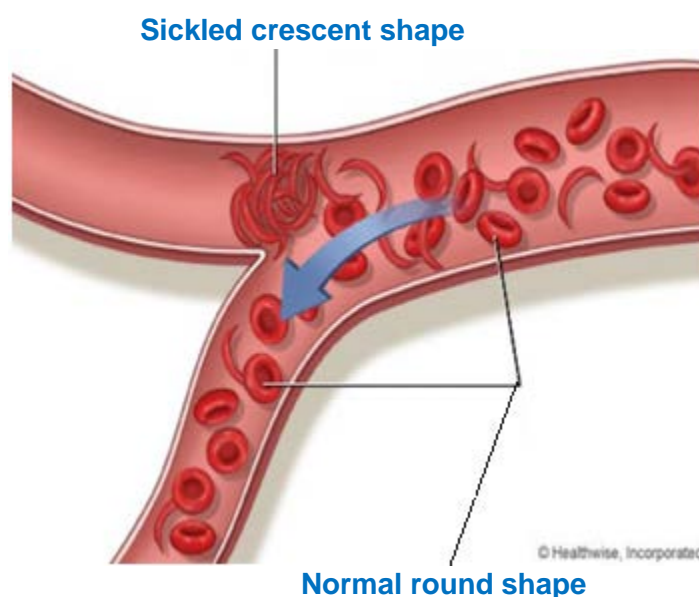
These are red blood cells from a person who has Sickle Cell Disease. Their red blood cells can lose their normal round shape and change to a sickle or crescent shape under certain conditions such as low oxygen. If oxygen does not get to the body's tissues, it will cause pain and begin to weaken and deteriorate the tissues. Sickled cells cannot hold onto oxygen as well as normal red blood cells. These sickled cells live for approximately 14 to 21 days instead of the normal 120 days.



Normal round shape

Sickled crescent shape

These are normal shaped red blood cells and sickled red blood cells in the blood vessels. Normal shaped blood cells move easily through the blood vessels. It is very difficult for sickled blood cells to pass through blood vessels because of their shape. Sickled blood cells can cause blockages in the vessels leading to many of the problems seen in Sickle Cell Disease.



Sickled crescent shape

Normal round shape

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Red Blood Cells & Hemoglobin

Red blood cells are found in our blood and contain hemoglobin. Hemoglobin carries oxygen throughout our body. Most people's red blood cells contain **hemoglobin A**.

This is a red blood cell from a person with normal **hemoglobin A**.



This is a red blood cell from a person with **Sickle Cell Trait**. It contains both **hemoglobin A** and **hemoglobin S**, but it is also a healthy normal cell. Parents and family members can have a simple blood test to determine if they have **Sickle Cell Trait**.



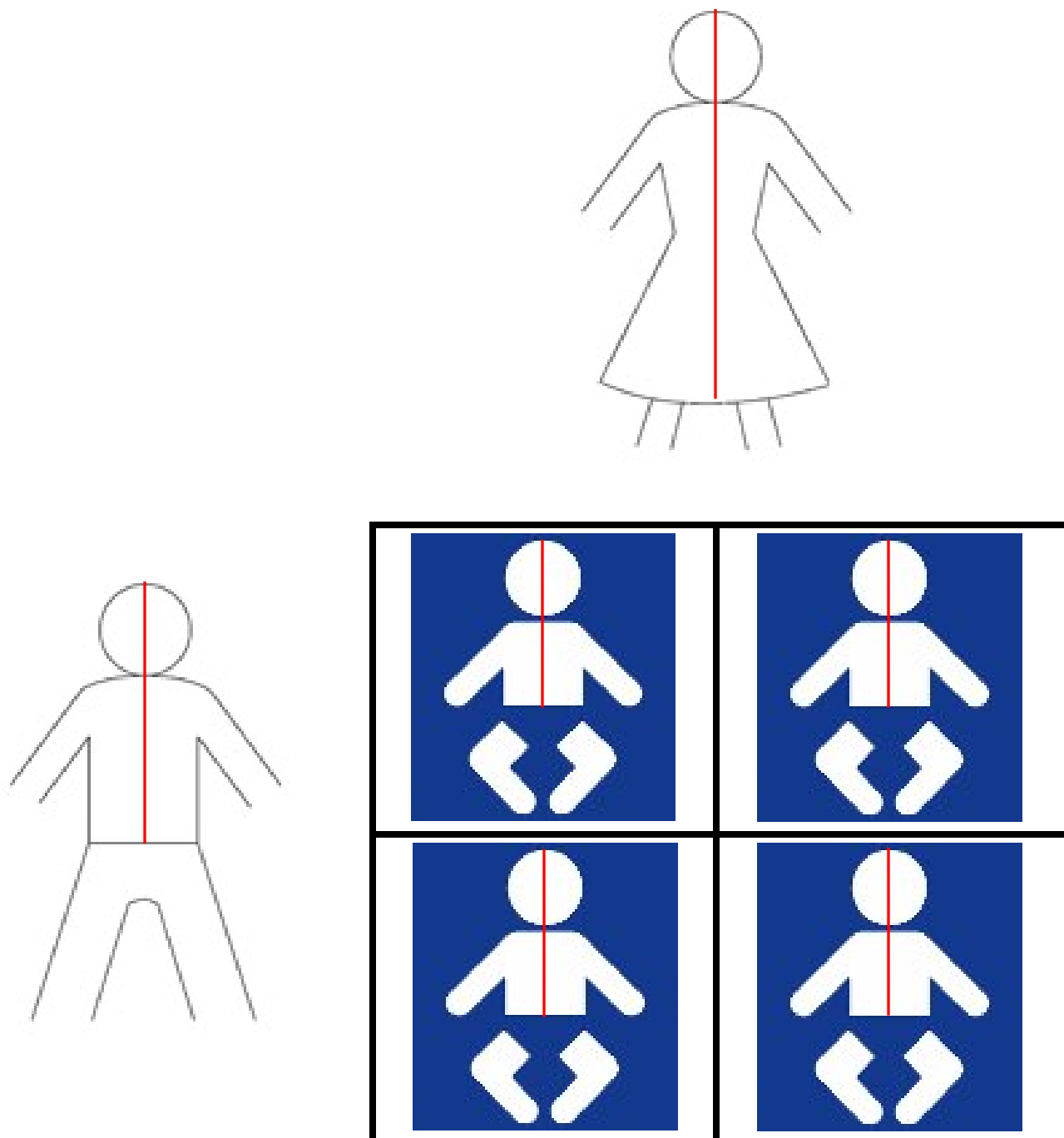
This is a red blood cell from a person with **Sickle Cell Disease**. It contains only **hemoglobin S**. The red blood cells of a person with **Sickle Cell Disease** can lose their round shape and change to a sickle or crescent. The sickled red blood cell does not work like a normal red blood cell. Red blood cells can also contain Hemoglobin F, which may help to reduce the chance of sickling.



Genes, Heredity & Hemoglobin

Genes determine eye color, height, skin color and our **hemoglobin** type. Heredity is the passing of genes from parent to child. Everyone has 2 genes for **hemoglobin**, one from our mother and one from our father.

Hemoglobin possibilities for a pregnancy:



Chapter 2:

Living Well with Sickle Cell Disease

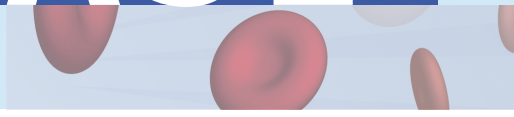
Chapter 2

Living Well with Sickle Cell Disease



SICKLE CELL DISEASE

Tips for Healthy Living.



Living Well With Sickle Cell Disease

People with sickle cell disease can live full lives and enjoy most of the activities that other people do. The following tips will help you, or someone you know with sickle cell disease, stay as healthy as possible.

- **Find good medical care.** Sickle cell disease is a complex disease. Good quality medical care from doctors and nurses who know a lot about the disease can help prevent some serious problems. Often the best choice is a hematologist (a doctor who specializes in blood diseases) working with a team of specialists.
- **Get regular checkups.** Regular health checkups with a primary care doctor can help prevent some serious problems.
 - Babies from birth to 1 year of age should see a doctor every 2 to 3 months.
 - Children from 1 to 2 years of age should see a doctor at least every 3 months.
 - Children and adults from 2 years of age or older should see a doctor at least once every year.
- **Prevent infections.** Common illnesses, like the flu, can quickly become dangerous for a child with sickle cell disease. The best defense is to take simple steps to help prevent infections.
- **Learn healthy habits.** People with sickle cell disease should drink 8 to 10 glasses of water every day and eat healthy food. They also should try not to get too hot, too cold, or too tired.
 - Children can, and should, participate in physical activity to help stay healthy. However, it's important that they don't overdo it, rest when tired, and drink plenty of water.
- **Look for clinical studies.** New clinical research studies are happening all the time to find better treatments and, hopefully, a cure for sickle cell disease. People who participate in these studies might have access to new medicines and treatment options.
- **Get support.** Find a patient support group or community-based organization that can provide information, assistance, and support.

Helping Your Child to Stay Well

Advice for Parents of Young Children with Sickle Cell Disease

► Get Good Providers and Hospitals

Find doctors who:

- Know about sickle cell disease or who are willing to learn.
- Can admit your child to a hospital that has experience in treating children with sickle cell disease.

► Keep Appointments with Your Child's Primary Doctor

- Make sure that your baby gets all his or her baby shots (immunizations) on time.
- Talk about your child's growth and development.
- Have your child's hearing and vision checked.

► Keep Appointments with Your Child's Hematologist

- Take your baby to the hematologist even when your child is well. **Don't skip these appointments.**
- This helps doctors know what your child is like when he or she is well. It can help them to make better decisions about what to do when your child is sick.

► Give Your Child Penicillin – Twice A Day, Every Day

- Penicillin helps prevent certain kinds of dangerous infections.
- Children with sickle cell disease cannot fight off infections as well as other children. They can become very sick, very quickly.
- The infections can be serious and can even cause death.
- **Giving penicillin every day can truly make the difference between life and death.**

► Make Sure Your Child Gets His or Her Baby Shots (Immunizations) on Time

- Be sure to keep all of your child's immunization appointments.
- Your child will also get a flu shot every year.
- Bring your immunization card with you to keep track of your child's baby shots (immunizations).

► Make Sure Your Baby Stays on Breast Milk or Formula for the First Year

- Breast milk or formula is important for your baby's growth and development.
- Give your baby the amount of formula your doctor recommends. Do not give other types of fluids instead of formula or breast milk.

► Let Your Child Drink Enough Fluids

Give your child extra fluids:

- when your child has a fever or pain episode
- when your child is active
- when it is a hot, humid day
- when he or she is traveling



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► Give your child vitamins

- Even a healthy diet can benefit from extra vitamins.
- Your child's doctor may prescribe daily **follic acid** to help your child make new red blood cells.
- Children with sickle cell disease should take regular daily vitamins, especially if they do not eat well.
- More children with sickle cell disease have low zinc, Vitamin D, and Vitamin A. Check with your child's doctor to find out whether your child needs more of these.

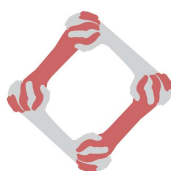
► Avoid Letting Your Child Get Chilled

- Dress your child warmly when the weather is cool, or when the child is going to be in an air conditioned room for a long time.
- Don't let your child swim in very cold water, even when the weather is warm.
- Dry your child off quickly after a swim and be sure to wrap him or her in a dry towel to keep warm.

► Avoid Temperature Extremes

Your child may be more likely to have a pain episode when it is very hot and humid (over 80 degrees) or very cold (less than 32 degrees) outside. Sudden changes in weather conditions can cause the child to develop a pain episode.

► Let Your Child Rest When Needed



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Eating to be Well with Sickle Cell Disease

With Sickle Cell Disease it is especially important to eat a **balanced diet** to make sure that we give our body the energy, fiber and vitamins and minerals that it needs to keep us healthy.

What is a balanced diet?

A balanced diet is a diet that includes foods from all of the five major food groups and oils. Each of the different food groups provides our bodies with important nutrients. Avoiding any one food group places individuals at risk for less than optimal nutrition. The following includes examples of the different food groups:

Food Groups:	Examples of foods and what they give our bodies:
Grains 	Whole grain breads, rice, pastas, cereals and crackers – <i>aim to make at least half of the grains that you eat “whole” grains</i> <ul style="list-style-type: none"> • <i>Fiber – helps to reduce blood cholesterol and keeps our bowels moving</i> • <i>Folate – (in fortified grains)- helps our body make new red blood cells</i> • <i>B-vitamins – helps our body to use the energy from foods that we eat</i> • <i>Magnesium – important for healthy bones</i> • <i>Selenium – important for healthy immune system</i>
Vegetables 	Spinach, carrots, broccoli, tomato, green beans, salad – <i>aim for orange and dark green leafy vegetables*</i> <ul style="list-style-type: none"> • <i>Fiber – Tip: keep the skin on veggies and fruits for extra fiber</i> • <i>Folate – Tip: choose leafy greens</i> • <i>Vitamin A – helps to keep eyes and skin healthy and helps to protect against infections</i> • <i>Vitamin C – helps heal wounds and keeps teeth and gums healthy</i> • <i>Potassium – may help maintain healthy blood pressure</i>
Fruits 	Oranges, bananas, apples, grapes, melon, berries, kiwi, peach, plums * <ul style="list-style-type: none"> • <i>Provide similar vitamins, minerals and fiber as vegetables, see above.</i> • <i>*Note: individual fruits and vegetables provide different amounts of vitamins, minerals and fiber so it is important to eat a variety of different fruits and vegetables daily to maximize your benefits.</i>
Milk 	Low fat milk, yogurt and cheese <ul style="list-style-type: none"> • <i>Calcium and Vitamin D – for strong, healthy bones</i> • <i>Potassium – especially in milk and yogurt</i>
Meats & Beans 	Lean chicken, pork, fish and beef, nuts, seeds, peas and beans <ul style="list-style-type: none"> • <i>Protein and zinc – for energy and growth</i> • <i>Iron – helps to carry oxygen in our blood</i> • <i>Omega 3 fatty acids –in certain fish, nuts and seeds–help to reduce inflammation and decrease risk of chronic diseases, like heart disease</i>
Oils	Liquid vegetable oils like olive, canola, corn oil; nuts <ul style="list-style-type: none"> • <i>Vitamin E – works as an antioxidant to help to prevent disease</i> • <i>Omega 3 fatty acids –in flax, canola, soybean and walnut oils, and walnuts, flaxseeds and pumpkin seeds</i>

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.

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.

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.



Putting on a jacket when it's cold.



Playing with his friends.



Taking time out to drink.



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 areas in which you can take charge:

- ◆ **Your feelings**
- ◆ **Helping your child at different ages**
- ◆ **Brothers and sisters**
- ◆ **Getting support**

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

Your Feelings after Learning Your Child has Sickle Cell Disease

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 baby”. You may feel guilty about the fact that the disease is caused by you and your partner’s genes. You may be angry that you or your partner did not know that you or your partner carried the trait. You may feel angry that your baby’s and family’s future will be changed in an unknown way. You may also feel afraid and helpless in facing your baby’s future.

No matter how you feel, it is okay to feel that way. Feelings aren’t right or wrong. What matters is how you deal with your feelings.

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.

Try to notice what you are feeling:

- 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 want to 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 sickle cell disease

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

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 fevers, 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.



Soothing your baby.

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 imagination 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.

Because your child has sickle cell disease, you may feel he should be treated differently than other children (special treats, toys, attention, less discipline). This is not really best for the child, however. Children with sickle cell disease need to feel that they are the same as other children. If your child has brothers and sisters, they should all have the same rules to follow and your expectations for their behavior should be the same.

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.

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.

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. This can be true even if the attention your child with sickle cell disease is getting is not fun, like having IV's, shots, and seeing doctors and nurses. 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.

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."

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



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

Information You Should Share with Your Child's Babysitter or Childcare Provider

Childcare Providers

- You may have a babysitter or other childcare provider that helps you take care of your child.
- You can help your childcare providers learn what they need to know about taking care of a baby or young child with sickle cell disease.
 - Make sure your childcare providers learn how to tell if your child is getting sick.
 - Also make sure that they know how to contact you and what to do if your child has a fever or is sick.

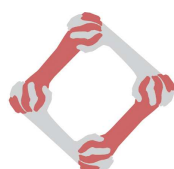
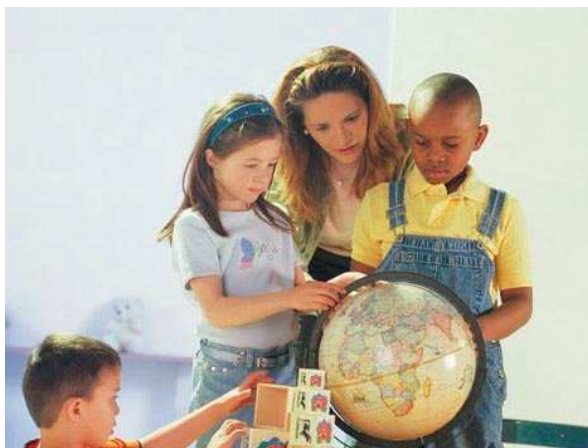


Teachers

- As your child gets older, they may go to pre-school or start kindergarten.
- You can also help your child's teachers learn what they need to know about sickle cell disease

On the next pages you will find information that you can share with your child's babysitter or childcare providers

- You should make sure to give a copy of this information to people that take care of your child.



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Information for Babysitters and Childcare Providers

My child has **Sickle Cell Disease**. This is a condition that affects red blood cells and causes anemia.

You should call me immediately if you notice any of the following symptoms in my child. These are all times when my child needs to see a doctor RIGHT AWAY.

- Fever of **101° or higher (Never ignore this!)**
 - Severe headache or dizziness
 - Severe pain or swelling in the belly
 - Rapid breathing, or coughing with chest pain
 - Very pale skin or palms or inner eye lids
 - Cannot move hands, arms or legs
 - Limps without pain
 - Cannot wake up
 - Slurred speech or drooling
- **If you cannot reach me, you should call 911.**
 - **When you call the doctor or 911, make sure to tell the person that answers that my child has Sickle Cell Disease.**

There are things that I need to know about right away so that I can call my child's doctor for advice. You should call me if my child:

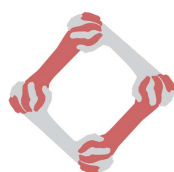
- Vomits or has diarrhea
- Keeps coughing
- Has pain
- Is not acting like they usually do:
 - Refuses to take penicillin
 - Is less active than usual
 - Refuses to eat or drink

Here are some other things you need to know about my child. I will talk with you about these things:

- My child needs to drink plenty of fluids and have healthy meals and snacks.
- My child needs to get enough rest.
- My child needs to avoid temperature extremes (too cold or too hot and humid). It is important to dress my child in warm clothes when the weather is cold.

Please provide your babysitters, childcare providers and family members with this important information.

Extra copies of this form can be located at the end of "A Parent's Guide to Sickle Cell Disease"



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CONTACT INFORMATION

You can reach me at _____

If you cannot reach me, call _____ (Name)

_____ (Phone Number)

My child's doctor is _____ (Name)

_____ (Number)

Other notes

Credits



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Sickle Cell Disease and
Newborn Screening Program



SCDAA National Headquarters 2008

Uade available through grant number U38MC00217-07-03
from the Genetic Services Branch of the Maternal and Child Health Bureau

Chapter 3:

Health Issues in Early Childhood

Chapter 3

Health Issues in Early Childhood

Be Aware of Some Problems That Can Arise In Your Child with Sickle Cell Disease

Anemia

What you need to KNOW

- Sickled red blood cells don't stay in the blood as long as normal red blood cells.
- Because sickled cells don't last as long, a person with sickle cell disease may not have as many red blood cells in their blood as people who don't have sickle cell disease. This is called **anemia**.
- Babies with anemia may become fussy or not want to eat. They may also feel weak or tired.
- Children with anemia feel weak, too.
 - They may feel tired more easily than other children and should rest when they feel tired.
 - They may have a tough time concentrating and this can affect how they do in school.
 - Children with sickle cell disease may need some extra help from you or their teacher.
- Sometimes, children with sickle cell disease need a blood transfusion because of their anemia.

What you need to DO

- Talk with your child's doctor about signs of anemia in your child.
- Learn to recognize if your child's anemia is getting worse.
- Know your child's baseline hemoglobin level.
- Do not give your child extra iron unless your doctor tells you to do this. Too much iron in the body is bad.

Write down your child's baseline hemoglobin level and date of test, and keep it in a safe place.



National Coordinating
and Evaluation Center

Sickle Cell Disease and Newborn Screening Program

**Sickle Cell Disease and Newborn Screening
Program**



**SCDAA National Headquarters
2008**

This fact sheet is made available through grant number U38MC00217-07-03 from the
Genetic Services Branch of the Maternal and Child Health Bureau

PENICILLIN....EVERYDAY!

Penicillin is one of the most important medicines you can give your child. Studies have shown that daily doses of penicillin for babies and young children with Sickle Cell Disease will lessen the number of infections they get.

Your child will take penicillin **twice** a day, once in the morning and once in the evening. Your doctor will write a prescription for the penicillin. You will need to go to your pharmacy to have the prescription filled. Penicillin can be given in two ways:

LIQUID: Liquid can be given by spoon or dropper and given to the child without mixing it with food or liquids. You can also mix liquid penicillin in a small amount of juice or milk (**2 ounces**) and have your child drink **all the mixture**. **Liquid penicillin lasts for only two weeks after the prescription is filled and it must be refrigerated.**

PILLS: Penicillin in pill form lasts a long time. You can get a supply that will last many months. For young children, pills will need to be crushed and added to **1 tablespoon** food. A pill crusher can be purchased at your local pharmacy.

IMPORTANT TIPS:

- **Give your child penicillin until the doctor tells you to stop**
- **Give penicillin at the same time every day, make it a routine!**
- **Give penicillin even if your child is NOT sick.**
- **Your child may be given a different antibiotic for other infections (i.e., ear infection) or during a hospitalization. Restart the penicillin immediately after the other antibiotic ends.**

Be Aware of Some Problems That Can Arise In Your Child with Sickle Cell Disease

Infections

What you need to KNOW

When it comes to fever and infections your child with sickle cell disease is different from other children.

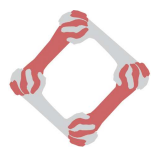
- Children with sickle cell disease cannot fight off infections as well as other babies.
 - They can become very sick, very quickly.
 - The infections can be serious and can even cause death.
- There are things that your doctor will do to help your child stay well:
 - Your doctor will tell you to give your child **penicillin**. Your child must take penicillin every day. This will help protect your child from infections.
 - If your child has an allergy to penicillin your doctor will tell you about other medicine that can be used.
 - Your child should get all of the baby shots (immunizations) on time.
 - Your child will also need special shots against the flu and the dangerous bacteria blood infections.

What you need to DO

- **If your child gets a fever of 101 degrees call or go see your doctor right away.**
 - Learn how to take your child's temperature.
 - Do not give your child fever medications, like Tylenol® or Advil®, until you talk with a doctor who understands sickle cell disease. These medications can hide the signs and symptoms of infection but not treat it.
 - **Make sure your doctor knows that your child has sickle cell disease.**
 - Give your child his or her medicine every day.
 - Take your child to the doctor to get all the shots.



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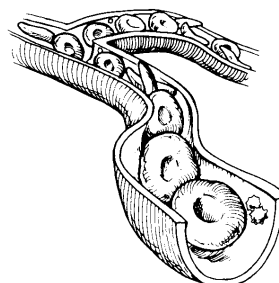


Fever and Infection

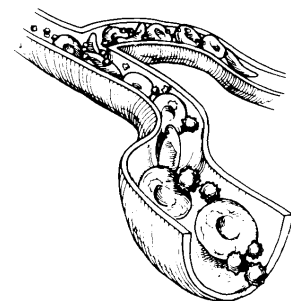
PNEUMOCOCCAL INFECTION

Infection is the major cause of death in children with sickle cell anemia. Infections cause deaths more rapidly, and are more difficult to get rid of in patients with sickle cell anemia than in normal persons.

An especially serious germ is the *pneumococcal* bacteria. It causes pneumonia, meningitis (infection of the brain) and septicemia (blood poisoning). This germ is responsible for most of the deaths in children with sickle cell anemia under 3 years of age.



Infection



Septicemia (blood poisoning)

It is estimated that the children with sickle cell anemia are 600 times more likely to get a *pneumococcal* infection than the normal population. Most of the infections occur before the age of 3 years. 35% of the children with sickle cell who get a *pneumococcal* infection die.

The spleen in a normal person has two functions to help fight infection. It filters or removes germs from the blood stream and makes antibodies that help fight infection outside of the spleen. In a child with sickle cell anemia, the sickled cells block the blood vessels in the spleen so blood can't move through it to be filtered. It also can't make the antibodies that fight infection. Thus, the bacteria can grow in the blood stream and cause blood poisoning (septicemia).

SYMPTOMS OF PNEUMOCOCCAL INFECTION

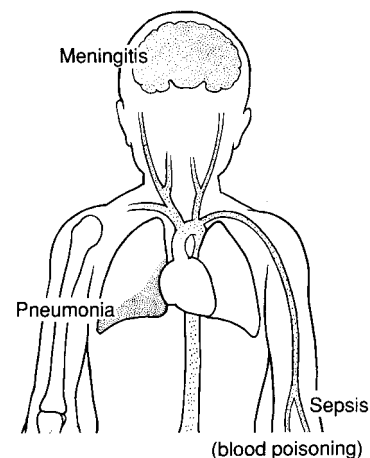
- Fever 101°F or 38°C or higher
- Cranky
- Unusual sleepiness
- Vomiting
- Diarrhea
- Rapid breathing
- Cough
- Pale color
- Trouble breathing

**** A FEVER MAY BE THE ONLY SYMPTOM AT FIRST**

If your child has any of these symptoms, even if over 5 years of age and/or on penicillin, the child should be seen by either your private doctor or a doctor in the emergency room as *soon as possible*.

Your child will be examined and may have lab work and x-rays to find the cause of the fever. Your child will be given an IV antibiotic and may be sent home on a strong oral antibiotic. Depending on your child's symptoms, he/she may be admitted to the hospital to be watched closely.

The *pneumococcal* infection is treatable and complete recovery is possible if the infection is recognized and treated early enough. However, even with treatment, permanent disabilities and death can result.



PENICILLIN

Penicillin kills the *pneumococcal* bacteria before it can cause blood poisoning in a child with sickle cell. It must be taken every 12 hours. If a dose is missed, the body is not protected against the pneumococcal bacteria and blood poisoning can still occur very rapidly. **IT IS IMPORTANT TO GET YOUR CHILD'S PENICILLIN REFILLED BEFORE IT RUNS OUT.**

It is important to remember that your child can still get blood poisoning even though the penicillin is taken regularly. Some bacteria may be resistant to the penicillin. If your child develops a fever of 101°F or 38°C or higher, even taking penicillin, he/she should see a doctor immediately. Other antibiotics can be used to fight the bacteria resistant to penicillin.

There are also immunizations given to children with sickle cell that will help to prevent infection with pneumococcal bacteria.

Supported in part by Projects # MCJ-481004 and # 2H46 MC00232-02 from the Maternal and Child Health Bureau (Title V, Social Security Act).

Adapted from materials by the Texas Department of Public Health Newborn Screening Program.





SICKLE CELL DISEASE

Tips for Healthy Living.

5 Tips To Help Prevent Infection

Common illnesses, like the flu, can quickly become dangerous for a person with sickle cell disease. The best defense is to take simple steps to help prevent infections.

1. Washing Hands. Washing your hands is one of the best ways to help prevent getting an infection. People with sickle cell disease, their family, and other caretakers should wash their hands with soap and clean water many times each day. If you don't have soap and water, you can use gel hand cleaners with alcohol in them.

Times to wash your hands:

BEFORE

- Making food
- Eating

AFTER

- Using the bathroom
- Blowing your nose, coughing, or sneezing
- Shaking hands
- Touching people or things that can carry germs, such as:
 - Diapers or a child who has used the toilet
 - Food that is not cooked (raw meat, raw eggs, or unwashed vegetables)
 - Animals or animal waste
 - Trash
 - A sick person

2. Food Safety. A bacteria, called salmonella, in some foods can be especially harmful to children with sickle cell disease. How to stay safe when cooking and eating:

- Wash hands, cutting boards, counters, knives, and other utensils after they touch uncooked foods.
- Wash vegetables and fruit well before eating them.
- Cook meat until it's well done. The juices should run clear and there should be no pink inside.
- Do not eat raw or undercooked eggs. Raw eggs might be hiding in homemade hollandaise sauce, caesar and other homemade salad dressings, tiramisu, homemade ice cream, homemade mayonnaise, cookie doughs, and frostings.
- Do not eat raw or unpasteurized milk or other dairy products (cheeses). Make sure these foods have a label that says they are "pasteurized."

3. Avoid Reptiles. A bacteria, called salmonella, that some reptiles have can be especially harmful to children with sickle cell disease. Make sure children stay away from turtles, snakes, and lizards.

4. Vaccines. Vaccines are a great way to prevent many serious infections. Children with sickle cell disease should get all the regular childhood vaccines, plus a few extra.

The extra ones are:

- Flu vaccine every year after 6 months of age.
- A pneumococcal vaccine (called 23-valent pneumococcal vaccine) at 2 and 5 years of age.
- Meningococcal vaccine (for some children).

Adults with sickle cell disease should have the flu vaccine every year, as well as the pneumococcal vaccine and any others recommended by a doctor.

5. Penicillin. Penicillin can help prevent infections. Take penicillin (or other antibiotic prescribed by a doctor) every day until at least 5 years of age.

For more information, please visit www.cdc.gov/ncbddd/sicklecell



Acute Chest Syndrome

WHAT IS ACUTE CHEST SYNDROME?

Acute chest syndrome is a common cause of hospitalization in children with sickle cell disease. It is clinically similar to pneumonia. Acute chest syndrome can be fatal in the child with sickle cell disease.

WHAT CAUSES CHEST SYNDROME?

Although the reasons are not clearly understood, it is believed that "sickled cells" clump together in the small blood vessels either in the lungs or moves there from somewhere else in the body. Sometimes this is triggered by a lung infection like pneumonia. Acute chest syndrome can also develop right before, during, or after an episode of pain in the abdomen or bones. There are no methods available to separate pneumonia from chest syndrome caused by blocked vessels. That is why your child may be treated like he/she has both.



WHAT ARE THE SYMPTOMS OF ACUTE CHEST SYNDROME?

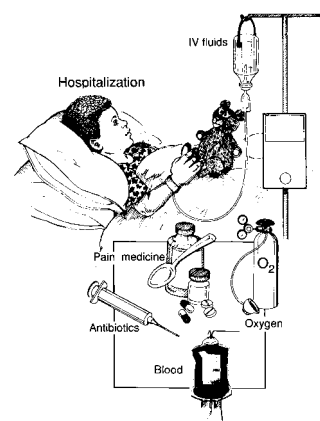
- Sometimes the chest hurts so bad that the pain spreads to the stomach.
- Fever of 101°F or 38.5°C or higher.
- Very congested cough.
- Trouble breathing.
- Fast breathing.
- You may see your child's ribs "suck in" when he/she breathes in.

If you see any of these symptoms in your child, visit your doctor or clinic **IMMEDIATELY!**

TREATMENT

Your child will need to have blood drawn to check the complete blood count (CBC) and any infection in the blood. X-rays of the lungs will also be taken.

Most children with acute chest syndrome are admitted to the hospital so that they can be watched closely. Pain medicine for the chest pain, oxygen and IV fluids may be given to your child. Sometimes a blood transfusion is necessary. He/She will also take a strong antibiotic to fight any infection.



OUTCOME

With proper therapy, children with acute chest syndrome usually do very well. However, some children have repeated episodes. The long-term effect of acute chest syndrome on lung function during adulthood is unknown.

For children with repeated or severe acute chest events, medications or blood transfusions may help to prevent future episodes. Any child with sickle cell and asthma should be taking daily asthma medications to prevent acute chest syndrome.

Supported in part by Projects # MCJ-481004 and # 2H46 MC00232-02 from the Maternal and Child Health Bureau (Title V, Social Security Act).

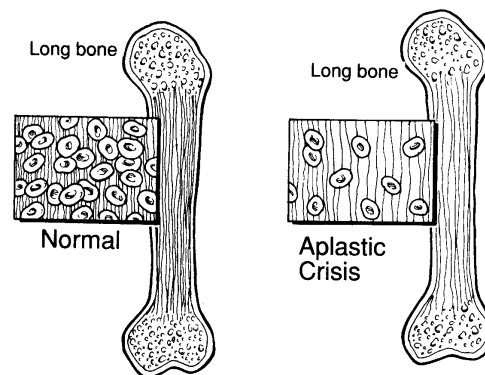
Adapted from materials by the Texas Department of Public Health Newborn Screening Program.



Aplastic Crisis

WHAT IS AN APLASTIC CRISIS?

Red blood cells are made inside bones (bone marrow). An aplastic crisis is an infection caused by parvovirus B19. It causes production of red blood cells (RBCs) in the marrow to be shut down for up to 10 days. This means that RBCs are not being made during this period. Because the RBCs in children with sickle cell anemia live only 10-15 days (compared to 120 days in children who do not have sickle cell anemia), the blood count (hemoglobin & hematocrit) drops rapidly to dangerously low levels during the infection.



WHAT ARE THE SYMPTOMS?

- Paleness
- Lethargy
- "Not feeling good"
- Headache
- Fever
- Anemia (low blood count)
- Recent upper respiratory infection
- Passing out (fainting)



WHO GETS AN APLASTIC CRISIS?

An aplastic crisis usually occurs in children under the age of 16 years. It occurs in the general population, but can only be noticed in those people with chronic anemias (such as sickle cell anemia).

WHAT IS THE TREATMENT?

Most often a blood transfusion is given to raise your child's blood count until the body starts making its own RBCs again. Occasionally it is necessary to hospitalize a child during an aplastic crisis.

CAN AN APLASTIC CRISIS HAPPEN MORE THAN ONCE?

No, recurrences of aplastic crisis are rare. Once affected, immunity to parvovirus is usually lifelong.

WHAT TYPE OF FOLLOW-UP IS REQUIRED?

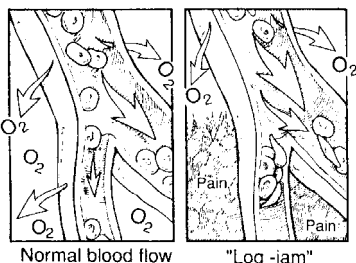
Your child will be given a follow-up appointment to check the blood count to make sure his or her body is producing RBCs again and to make sure the blood count is back to normal. Usually just one or two extra visits are needed. If there is another child in the household with sickle cell anemia, he or she should also have a blood count since they may have an aplastic crisis too. Parvovirus is usually very contagious to those who have never had it.



Painful Episodes

PAINFUL EPISODES IN THE CHILD WITH SICKLE CELL DISEASE

Painful episodes occur in children with sickle cell disease as a complication of the disease. These episodes more commonly occur in older children, but often happen in infants and young children.



WHAT CAUSES THE PAINFUL EPISODES?

The exact cause of the pain is not known. It is thought that the red blood cells (RBCs) become trapped inside a blood vessel and interfere with normal blood flow. If blood flow is reduced in even a small area of the body, it can cause pain.

Sometimes swelling is seen in the area of the pain. In children under 2 years of age the swelling usually occurs in the hands and/or feet. Older children can have swelling in the arms and/or legs. Swelling usually does not mean that something is seriously wrong, but in rare cases, swelling and pain are caused by infection in the bone. A child with swelling other than hands and feet should be seen by a doctor.

Hand-Foot Syndrome (Dactylitis)



WHERE IS THE PAIN?

Most often the pain seems to be in the bone, but it can occur anywhere in the body (chest, stomach, hands and/or feet, back, etc.) These painful episodes are not usually dangerous. They can last for several hours, days or even up to a week or two.

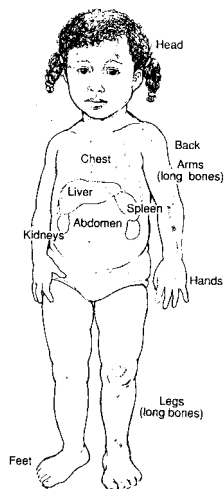
CAN THE PAIN BE PREVENTED?

Not all pain crises can be prevented. The best way to keep pain away is to make sure the child is drinking throughout the day and to avoid getting cold or chilled (cold water swimming, going outside in the winter without warm clothing). Ice packs can cause pain and should never be used for someone with sickle cell.

WHAT CAN BE DONE TO EASE THE PAIN?

Painful episodes can be mild, moderate or severe in terms of how much it hurts to your child. Taking medication for pain and drinking plenty of liquids usually relieves discomfort. A child may refuse to use the part of the body that is painful. If a child will not stand or walk or move a part of his body that is painful, do not force him to do so. As soon as the pain is better, he will be active again. To ease the pain, try giving Ibuprofen (Motrin, Advil) or Acetaminophen (Tylenol) every 4 hours.

If your child is still uncomfortable after receiving the Tylenol, you may need to give him Tylenol with Codeine (liquid or tablets) every 4 hours until the pain is better. Call your doctor or clinic if you do not have enough pain medicine at home.





Splenic Sequestration in Sickle Cell Disease

WHAT IS THE SPLEEN AND WHAT DOES IT DO?

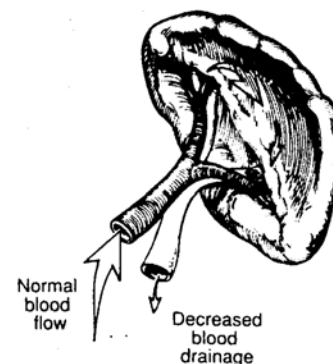
The spleen is normally a small organ located on the upper left side of the abdomen under the rib cage. It functions as part of the body's defense system that fights infection by removing bacteria (germs) from the blood. Basically, it serves as a filter in the bloodstream.

In sickle cell disease the spleen usually does not work normally after 4-6 months of age. It has been damaged by the sickled red blood cells (RBC's) and is not able to remove bacteria from the blood. This means that bacteria can grow in the blood and cause septicemia or blood poisoning (infection of the blood).

Some children with sickle cell anemia have an enlarged (big) spleen. Spleen enlargement is rare after age 5 in children with Sickle Cell Disease (Hemoglobin SS) - by this age, the spleen is usually shriveled up and therefore your doctor is not able to feel it. Children with Hemoglobin Sickle C disease (SC disease - another form of sickle cell disease) may have a big spleen until a much older age. Also, the spleen functions more normally in Sickle C disease.

WHAT IS SPLENIC SEQUESTRATION? (SPLEEN CRISIS)

When sickled cells block the blood vessels in the spleen, blood stays in the spleen instead of flowing through it. This causes the spleen to get bigger. When this happens the blood count (hemoglobin and hematocrit) falls and the spleen gets very large and easy to feel. This is called splenic sequestration crisis (or "spleen crisis"). Splenic sequestration can sometimes be painful.



WHO CAN GET SPLENIC SEQUESTRATION?

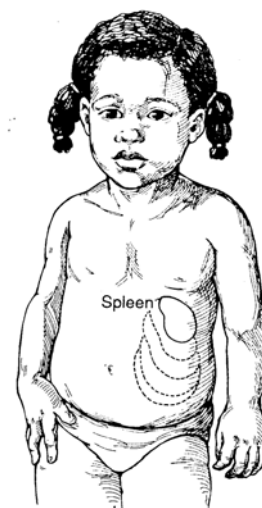
Infants and young children with sickle cell anemia who are between the ages of 2 months and 4 years are at greatest risk of splenic sequestration. Sequestration episodes can occur in older children with SC disease and Sickle Beta-Plus Thalassemia.

WHAT ARE THE SYMPTOMS OF SPLENIC SEQUESTRATION?

Your child may experience any of the following symptoms:

- Weakness
- Irritability
- Unusual sleepiness
- Paleness
- Big spleen
- Fast heart beat
- Pain in the left side of the abdomen

A child can have a seriously low blood count without many symptoms. Sometimes the only symptom is that he/she is not as active as usual.



IS SPLENIC SEQUESTRATION SERIOUS?

Any enlargement of the spleen must be looked at by your doctor. Parents should be taught how to feel for their child's spleen at their regular check-up visit. They need to know how their child's spleen usually feels so that when he/she seems sick, they can feel the spleen to see if it is bigger.

Acute splenic sequestration crisis can be serious and a potentially life-threatening problem if the spleen suddenly enlarges with a large drop in the blood count. The child needs to see a doctor immediately.

When the spleen gradually gets bigger over several weeks, the blood count does not change much and therefore it may not be as serious.

WHAT IS THE TREATMENT FOR SPLENIC SEQUESTRATION?

A blood transfusion is given if the blood count is dangerously low. Minor episodes of splenic sequestration are common. You will notice moderate increases in spleen size associated with a decrease in hemoglobin levels. These minor episodes usually resolve spontaneously, but require monitoring of spleen size and blood counts. If a child experiences several episodes of splenic sequestration, surgery to remove the spleen may be considered.

WHAT CAN BE DONE TO HELP CHILDREN WITH SICKLE CELL ANEMIA FIGHT INFECTION SINCE THE SPLEEN DOES NOT WORK?

Prevention and early treatment of infection is the best defense against serious complications. The child who is sick should be carefully watched for symptoms of serious infection. A fever of 101 degrees or higher should always be considered a symptom of possible septicemia or bacteria in the blood (see fever section).

Penicillin is given twice daily to prevent infection. Pneumococcal vaccines and other vaccines are given to children with Sickle Cell Disease until age 5 in order to boost immunity to harmful infection. If the spleen is surgically removed, vaccination continues for a longer period of time.

CAN SPLENIC SEQUESTRATION HAPPEN MORE THAN ONCE?

Yes. An infant or child that has had one episode of splenic sequestration is likely to have other episodes.





Stroke

WHAT IS A STROKE?

A stroke is a sudden and severe complication of sickle cell anemia. It affects from 6 to 8% of patients with sickle cell anemia, especially between 2 and 10 years of age. A stroke may occur with a painful episode or an infection, but in most cases there are no related illnesses.

Although recovery from the stroke may be complete in some cases, frequently the stroke can cause brain damage, paralysis, seizures, coma and even death.

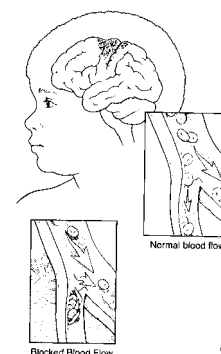
A repeat stroke causes greater brain damage and increases the risk of death. Repeat strokes occur in at least 60% of the children who have already suffered one stroke unless treatment is given. There is a test called Transcranial Doppler Ultrasound (TCD), which can predict strokes in some children with sickle cell.

WHAT CAUSES A STROKE?

The sickled cells in a child with sickle cell anemia have a hard time moving through the blood vessels in the brain. If some cells get "stuck" and can't move, other sickled cells pile up behind and cause a "log jam" that blocks the blood vessels. Oxygen can't get past the block to other parts of the brain, which causes the stroke.

SYMPTOMS OF A STROKE

- Jerking or twitching of the face, legs, arms.
- Convulsions or seizures.
- Strange, abnormal behavior.
- Inability to move an arm and/or a leg.
- Staggering or an unsteady walk when your children walked normally before.
- Stuttered or slurred speech when your child had clear speech before.
- Weakness in the hands, feet or legs.
- Changes in vision.
- Severe headaches that won't go away with Tylenol.
- Severe vomiting.



INITIAL TREATMENT OF A STROKE

Your child will be watched carefully in the hospital for any new or progressing symptoms. He/She may be put on medicine to control or prevent any seizures caused by the stroke. Your child may need special x-ray tests (such as CT and MRI scans) and physical therapy. He/She will probably be transfused with blood that does not have any sickle cells in it.

PREVENTION OF A REPEAT STROKE

To prevent another stroke from happening, your child will probably need to receive blood transfusions every 3 to 4 weeks. Your child will be transfused with blood that does not have any sickled cells in it. Your child's body then won't need to make any new blood with sickle cells. Although blood transfusions may help prevent any more strokes from happening, we do not know how many years your child must continue to be transfused.

Transcranial Doppler Ultrasound (TCD)

What is a TCD?

A TCD is a type of ultrasound done for children with sickle cell disease (SS or S β^0 Thalassemia) to find out their risk of having a stroke.

Why should my child have a TCD?

Children with sickle cell disease (SS or S β^0 Thalassemia) are at high risk of having a stroke, especially in early school age (ages 2-10). Studies have shown that we can identify children who may be at higher risk of having a stroke by performing a TCD. This study does not hurt, can be performed on any child who can stay still for the study (it lasts about 30 minutes) and is done in the outpatient setting.



TCD is:

- Proven, almost always, to predict stroke risk
- Non-invasive
- Painless
- Fast (exam takes about 30 minutes)

<http://www.georgiahealth.edu/neurology/Research/strokesicklecell/sicklepatients.html>

How does a TCD work?

A TCD uses sound waves to check how fast blood is moving through the blood vessels of the brain. The study is not an absolute: that is children may still have strokes even if the study is normal and occasionally, even with an abnormal result, the child may not have a stroke.

How often should my child have a TCD?

The National Heart, Lung, and Blood Institute recommend that all children with sickle cell disease (SS or S β^0 Thalassemia) have a TCD done every year from age 2-16. If the study is normal, it is usually repeated every year. If the TCD is abnormal it will be repeated sooner. Sometimes it is difficult to get a good TCD study, in which case your child's doctor will explain what else might be done to try to understand your child's risk of having a stroke.

If the TCD is abnormal can something be done to try to protect my child from having a stroke?

Yes! The doctor will talk to you about blood transfusions or possibly Hydroxyurea to reduce the risk of stroke.

Please ask your child's doctor if you have any questions about a TCD or the risk of stroke.

Chapter 4:

Organizing Your Child's Care

Chapter 4

Organizing Your Child's Care

Know What Good Health Care Is

Care that Follows a Plan

Check (✓) to make sure these happen:

Right After Birth

___ Baby has a primary care doctor (Pediatrician)

By 2 Months of Age

___ Baby should be referred to a Pediatric Comprehensive Sickle Cell Clinic or to a Pediatric Hematologist (a baby doctor who is a specialist in blood diseases)



- **Primary care doctor will see your child every 2 months for immunizations.**
- **Hematologist may also see your child every 2-3 months.**

Birth to 6 Months

___ The type of sickle cell disease is confirmed.
 ___ Parents are offered testing for themselves.
 ___ Baby starts getting penicillin 2 times a day.
 ___ Baby starts getting baby shots (immunizations).

6 Months to 1 Year

___ Baby keeps on getting penicillin 2 times a day.
 ___ Baby still sees the doctor every 2-3 months.
 ___ Doctor gets a blood count done every 2-3 months to find out what the baby's usual Hemoglobin level is.
 ___ Doctor gets other blood tests done to check the baby's liver and kidney function.
 ___ Doctor checks size of the spleen and teaches family members and other caregivers to do the same.
 ___ Baby keeps on getting baby shots.
 ___ Baby gets a flu shot.

1 to 2 Years

___ Baby keeps on getting penicillin 2 times a day.
 ___ Baby keeps on getting baby shots.
 ___ Baby starts getting a flu shot every year in the Fall.
 ___ Baby gets scheduled (soon after turning two) to have a special study called TCD to see if he or she is at risk for stroke (if your baby has a certain type of sickle cell disease).
 ___ Once a year, baby gets other blood tests to check his or her liver and kidney function.

3 to 5 Years

___ Increase dosage of penicillin and continue to take 2 times a day.
 ___ Continue baby shots (immunizations).
 ___ Continue getting yearly flu shot.
 ___ Continue getting a TCD (for children with certain types of sickle cell disease).
 ___ Continue yearly blood tests to check liver and kidney function.



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Care that is Comprehensive

Check (✓) to make sure this happens.

Baby sees a doctor regularly

- to get checked for symptoms of the disease
- to see if referrals to other specialists are needed
- to see if special treatments are needed

Parents get information, support and advice

- **About the disease**

Parents get information about the disease, what to be aware of and what to do.

- **About parenting**

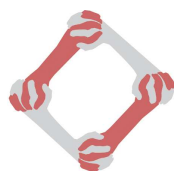
Parents get emotional support and advice on parenting.

- **About other issues of importance**

Parents get support on issues concerning insurance, transportation, housing, child care, when needed.



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Make Sure You Know the Answers to These Questions

- Here is a list of questions that the parents of a baby or child with sickle cell disease may ask.
- By talking with your child's Primary Care Doctor, Hematologist, and the other health care providers that you see, you should **know the answers** to the questions below.

From Birth to 6 Months

You should know the answer to questions about:

The symptoms of sickle cell disease you can expect to see in your baby.

- What you should do if you notice these symptoms.
- How to tell if your baby has a fever.
- How to take your baby's temperature.
- What doctors appointments, including specialists, you should set up for your child.
- If there are any community-based sickle cell programs in your area.
- If they can put you in touch with other families who have children with sickle cell disease.

From 6 Months to 1 Year

You should know the answer to questions about:

- Why it is important to give your child enough to drink.
- What foods, vitamins and minerals are best for your child.
- How to respond to fever as a medical emergency.
- How to know if your baby is having pain.
- How to manage pain at home.
- What to do if your baby's hands and feet are swollen.
- What the spleen is and how to tell if it is enlarged.



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When Your Baby is 1 to 2 Years

You should know the answer to questions about:

- ___ How to manage your child's pain.
- ___ Why eyes get yellow and what to do.
- ___ How to tell if there are problems with your child's lungs (Acute Chest Syndrome).
- ___ How to tell if there are problems with the spleen (Splenic Sequestration).
- ___ The signs and symptoms of a stroke.

When Your Baby is 3 to 5 Years

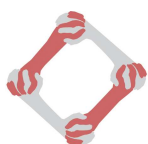
You should know the answer to questions about:

- ___ How to make sure your child is getting plenty of liquids and a proper diet.
- ___ How your child might grow and develop.
- ___ How to manage your child's pain.
- ___ Materials you can use to teach your child's pre-school teacher or staff about sickle cell disease.
- ___ Why it is important to have an educational plan for your child.

Questions?

You can ask your child's doctor, nurse, or other healthcare workers any questions you may have about your baby and sickle cell disease.

Write your questions down here.



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When to Call the Doctor or Go to the Emergency Room Advice for Parents of Young Children with Sickle Cell Disease

When it comes to fever and infections, your child with sickle cell disease is **DIFFERENT** from other children.

- Babies and children with sickle cell disease cannot fight off infections as well as other babies.
- Children with sickle cell disease have died in a matter of hours when they had certain bacteria infections that were not treated quickly. They could not fight off these infections without help.

When to Call to Be Seen **Right Away**

Call your child's doctor or nurse right away if your child has one of these danger signs:

- Fever of **101°** or higher (Never ignore this!)
- Severe headache or dizziness
- Severe pain or swelling in the belly
- Rapid breathing, or coughing with chest pain
- Very pale skin or palms or inner eye lids
- Cannot move hands, arms or legs
- Limps without pain
- Cannot wake up
- Slurred speech or drooling

If you cannot reach your child's doctor, go to the emergency room **right away**. **Make sure to tell the emergency room doctors right away that your child has sickle cell disease.**

When to Call for Advice

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

- | | |
|-------------------------------|--------------------------------------|
| • Vomiting | • Is not acting like they usually do |
| • Diarrhea | • Refuses to take penicillin |
| • Eyes look yellow (jaundice) | • Is less active than usual |
| • Pain | • Refuses to eat or drink |
| • Keeps coughing | |



Have a Plan for Getting Your Child to the Doctor or Emergency Room

- ▶ **Ask your child’s doctor and hematologist which emergency room you should use.**
 - Do this ahead of time.
 - Then you will know what to do if there is an emergency.

- ▶ **Write down the directions from your house to your child’s doctor’s office and hospital emergency room**
 - Make sure anyone who regularly cares for your child has a copy of the directions.
 - Keep a copy of these directions with you at all times.

- ▶ **Keep a list of phone numbers so that you can call a cab or ambulance company if needed**
 - Keep a copy of this list by your phone.

- ▶ **Find out if your Community-based Sickle Cell Program offers transportation services.**

- ▶ **Ask your family, friends and neighbors if they will help you when you need to get your child to the doctor or hospital quickly**
 - Keep a list of phone numbers of people that are willing to help out in an emergency.

- ▶ **Ask your child’s doctor or hematologist for a “travel letter” that you can take with you when traveling.**
 - You can show the letter to other doctors if your child gets sick while you are away.

Credits



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**Sickle Cell Disease and Newborn Screening
Program**



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2008**

Made available through grant number U38MC00217-07-03 from the
Genetic Services Branch of the Maternal and Child Health Bureau

Additional Support & Resources

A diagnosis of sickle cell disease may be overwhelming and extremely stressful at times. The ways people respond to the diagnosis and treatment vary, and many social, emotional, and spiritual concerns may arise. A pediatric hematology social worker is available to provide individual and family counseling that will assist you and your child to help deal with concerns in the following areas:

- Your child's feelings about him/herself
- Reactions to the illness
- School issues
- Financial and insurance concerns
- Providing electric and gas company documentation of medical necessity
- Application for a disabled parking permit



Suggested Reading: Platt, A., Eckman, J. and Hsu, L. Hope and Destiny: The Patient and Parent's Guide to Sickle Cell Disease (Revised Third Edition). Munster, IN. Hilton Publishing, 2011.

Local Organizations

Greater Boston Sickle Cell Disease Association

Telephone: (617) 825-4595

Website: www.gbscda.org

Boston University Center of Excellence in Sickle Cell Disease

Website: www.bu.edu/sicklecell

Massachusetts Department of Public Health Community Support and Care Coordination Programs for Children with Special Health Care Needs

Telephone: (800) 882-1435

Website: www.mass.gov/dph/specialhealthneeds

New England Newborn Screening Program

University of Massachusetts Medical School

305 South Street

Jamaica Plain, MA 02130

Telephone: (617) 983-6300

Website: www.umassmed.edu/nbs

National Organizations

Sickle Cell Disease Association of America

Website: www.sicklecelldisease.org

Provides the latest information on the treatment of sickle cell disease as well as research and news about the disease.

Sickle Cell Information Center

Website: www.SCInfo.org

Provides sickle cell disease patient and professional education, news, research updates, and worldwide sickle cell resources.

American Red Cross

Telephone: 1-800-GIVE-LIFE (1-800-448-3543)

Website: www.newenglandblood.org

National Society of Genetic Counselors

Telephone: (312) 321-6834

Website: www.nsgc.org

Baby's First Test

Website: www.babysfirsttest.org

Provides information about newborn screening

Financial Assistance

Mass Health

Telephone: 1-800-841-2900 (general information)

1-800-332-5545 (to apply for assistance)

Website: www.mass.gov/masshealth

Also known as Medicaid, this is a program of comprehensive medical coverage for low and moderate income residents of Massachusetts. Many kinds of coverage are available. Some programs include prescription drug coverage, as well as transportation assistance to medical appointments.

If you are eligible for Mass Health benefits, you most likely are eligible for Prescription for Public Transportation and WIC (please check with your primary care doctor or social worker)

Prescription for Public Transportation (PT-1 form)

Telephone: 1-800-841-2900

Website: www.mass.gov/masshealth

Provides free door-to-door transportation for your child to medical appointments. The application should be completed by your child's physician. Ask your physician or social worker how to obtain an application

WIC

Telephone: 1-800-942-1007

Website: www.mass.gov/wic

Supports low income women and children up to age five years by providing foods to supplement nutritional needs.

Education

Massachusetts Department of Education (DOE)

Telephone: (781) 338-3000

Website: www.doe.mass.edu/sped

- Bureau of Special Education Appeals

Telephone: (617) 626-7250

- Program Quality Assurance

Telephone: (781) 338-3700

Federation for Children with Special Needs

1135 Tremont Street, Suite 420

Boston, MA 02102

Telephone: (617) 236-7210/1-800-331-0688

Website: www.fcsn.org

Provides information, support and assistance to parents of children with disabilities, their professional partners, and their communities.

Mass. Association of Special Education Parent Advisory Councils (MASSPAC)

Telephone: (617) 236-7210/1-800-331-0688

Website: <http://www.masspac.org/>

Special Kids Special Help – Boston Medical Center

Website: www.specialkidsspecialhelp.org

Web based resource for parents with children with special needs.

Sickle Cell Disease: Information for School Personnel

Website: www.state.nj.us/health/fhs/sicklecell

Provides information designed for school personnel explaining many issues related to sickle cell disease. This is an excellent site to pass on to teachers or childcare providers.

Camps for Children/Teens with Sickle Cell Disease

The Hole in the Wall Gang Camp

Telephone: (860) 429-3444

Website: www.holeinthewallgang.org

Located in Ashford, Connecticut, this camp offers free sessions for children age 7 to 15. Special sessions are available for specific illnesses.

Camp Sunshine

Telephone: (207) 655-3800

Website: www.campsunshine.org

Located in Casco, Maine. Offers free sessions for families. Special sessions available for specific illnesses.

Legal Aid

Patient Advocate Foundation

Telephone: 1-800-532-5274

Website: www.patientadvocate.org

Provides education and legal counseling on managed care, insurance, and financial issues for Sickle Cell patients.

Disability Law Center of Massachusetts

Telephone: 1-800-872-9992

Website: www.dlc-ma.org

Massachusetts Advocates for Children

Telephone: (617) 357-8431

Website: <http://www.massadvocates.org/>

Parent Professional Advocacy League (PPAL)

Telephone: 1-866-815-8122

Website: www.ppal.net

Volunteer Lawyers Project of the Boston Bar Association

Telephone: (617) 423-0648

Website: <http://www.vlpnet.org/>

U.S Office for Civil Rights

Telephone: (617) 565-1340

Website: www.hhs.gov/ocr

Send complaints to:

Peter Chan, Regional Manager

Office for Civil Rights

U.S. Department of Health and Human Services

Government Center

J.F. Kennedy Federal Building – Room 1875

Boston, MA 02203

Government Agencies

Massachusetts Department of Mental Health

Telephone: 1-800-221-0053

Website: www.mass.gov/dmh

Massachusetts Department of Public Health

Telephone: (617) 624-6000

Website: www.mass.gov/dph

Massachusetts Office for Refugees and Immigrants

Telephone: (617) 727-7888

Website: www.mass.gov/ori

Massachusetts Department of Transitional Assistance

Telephone: (617) 348-8500

Website: www.mass.gov/dta

Contact Information

Boston Medical Center:	<p>Monday-Friday: 9am – 5pm (617)-414-5725</p> <p>Emergencies, Evenings and Weekends (617)-638-5795 and enter pager ID number 5731 to page the pediatric hematologist/oncologist “on call”</p> <p>Boston Medical Center Directory: 617-414-5000</p>
Children’s Hospital Boston:	<p>Monday-Friday: 8:30am – 5pm (617)-355-8246</p> <p>Emergencies, Evenings and Weekends (617)-355-6363 and ask for the pediatric hematologist/oncologist “on call” to be paged</p>
Massachusetts General Hospital:	<p>Monday-Friday: 9am – 5pm (617)-726-2737</p> <p>Emergencies, Evenings and Weekends (617)-726-2737 and ask for the pediatric hematologist/oncologist “on call” to be paged</p>
Tufts Medical Center:	<p>Monday-Friday: 8:30am – 5pm (617)-636-5535</p> <p>Emergencies, Evenings and Weekends (617)-636-5114 and ask for the pediatric hematologist/oncologist “on call” to be paged</p>
Baystate Medical Center:	<p>Monday-Friday: 8am – 4:30pm (413)-794-9338</p> <p>Emergencies, Evenings and Weekends (413)-794-9338 and ask for the pediatric hematologist/oncologist “on call” to be paged</p>
UMass Memorial Medical Center:	<p>Monday-Friday: 8:30am – 4:30pm (508)-856-4225</p> <p>Emergencies, Evenings and Weekends (508)-334-1000 and ask for the pediatric hematologist/oncologist “on call” to be paged</p>

Chapter 5:

Special Treatments for Sickle Cell Disease

Chapter 5

Special Treatments for Sickle Cell
Disease

Hydroxyurea for Children with Sickle Cell Disease

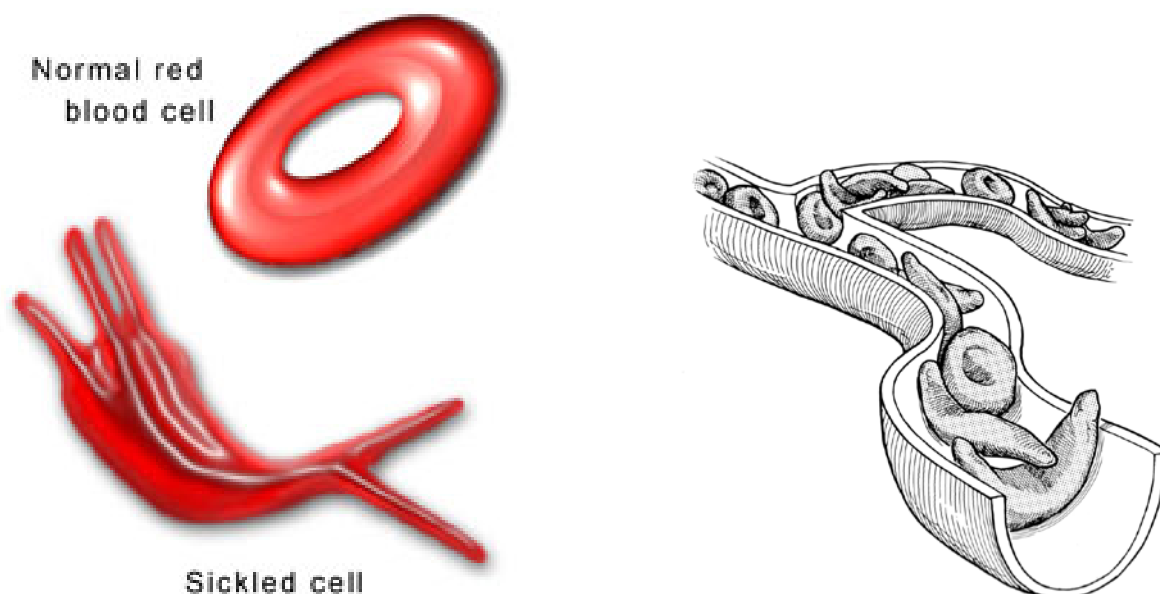
What is hydroxyurea?

- Hydroxyurea is a medicine that has been shown to help adults and children with sickle cell disease.
- Studies have shown that hydroxyurea can decrease the number of pain episodes, acute chest syndrome (pneumonia), blood transfusions and admissions to the hospital.
- Patients take hydroxyurea by mouth daily, either as a capsule or liquid.
- Hydroxyurea must be prescribed by a doctor and is available at most drug stores.
- The U.S. Food & Drug Administration approved hydroxyurea in 1998 for treatment of sickle cell disease in adults, although it has been used since the 1960s for other diseases.
- Hydroxyurea has been used in children with sickle cell disease for over 10 years.

How does hydroxyurea work?

Red blood cells contain hemoglobin, which carries oxygen. In sickle cell patients, abnormal hemoglobin causes the red blood cells to become long, rigid “sickle shaped” and sticky. Sickling causes blockage of blood flow to vital organs, muscles and tissues.

With hydroxyurea red cells have more fetal hemoglobin, become larger, less sticky, and travel more easily through blood vessels. Hydroxyurea also decreases the number of white blood cells which causes less inflammation and sickling.



Who should take hydroxyurea?

Children with sickle cell disease are eligible for hydroxyurea treatment. Children must come regularly to Hematology Clinic visits for blood counts and physical exams. Children with complications of sickle cell disease (frequent pain, pneumonia, very low hemoglobin, etc.) are especially suited for hydroxyurea. Even those with milder disease may benefit since the treatment may prevent sickle complications.

What are the proven benefits of hydroxyurea?

- Fewer pain episodes
- Fewer episodes of pneumonia and acute chest syndrome
- Fewer hospital admissions
- Fewer blood transfusions
- Improved appetite
- Increase in height and weight
- More energy
- Improved self-image
- Fewer missed school days

Are there side effects?

Hydroxyurea is extremely well tolerated. Although hydroxyurea is in the chemotherapy family of medicines, the dose is low enough so as not to cause some of the usual side effects seen with chemotherapy (hair loss, vomiting, etc.). It can cause a decrease in blood counts and must be carefully monitored for this side effect. A monthly blood count is usually required when starting the medication.

Adolescent girls who have their periods must take some form of birth control due to the potential harm hydroxyurea can have on their unborn baby.

The long-term effects of hydroxyurea are still being studied. There is a question that it might cause cancer. However, this has never been shown in individuals with sickle cell disease, including those who have been taking the medicine for more than twenty years.

Is it safe?

Yes. There have been no severe side effects reported in children ranging in age from 6 months to 15 years. Some pediatric patients have been with treated hydroxyurea for more than ten years, and they have no reported serious complications. If taken as directed and if blood counts are checked regularly, it appears to be safe.

How do you take it?

Hydroxyurea comes as a capsule or a liquid and is taken by mouth daily. Treatment will begin on a low dose daily based on weight and increasing every few weeks depending on its effect.

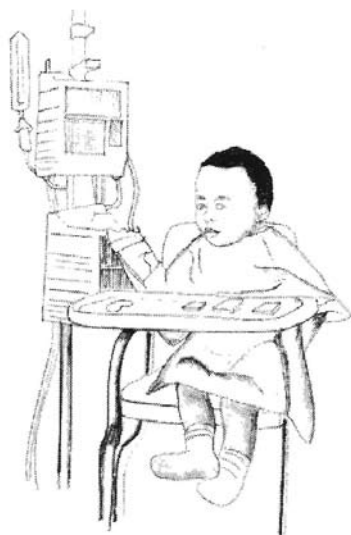
What is the cost of Hydroxyurea?

Hydroxyurea is fully covered by most private drug insurance policies and by MassHealth.

How do I get more information?

Contact your hematologist for more information about whether hydroxyurea is right for you or your child.

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. Usually this is caused by a viral infection, but there are other infections and reasons for your child's blood counts to drop to a dangerous level.
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. This procedure can be done by nurses with syringes or with a machine run by special nurses that can exchange blood very rapidly. Sometimes your child will need a new IV line or a special IV line for these procedures.

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 correct.

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 HIV (AIDS), hepatitis and syphilis. But rarely, infections are transmitted by transfusions. Infection by the viruses that are screened (HIV, hepatitis B and C) are very rare, less than one in two or three million. Infection by bacteria that may be in the blood is also rare, but is more common than viral infections (less than one in thirty or forty thousand).

Allo-immunization

Your child may develop antibodies that destroy the blood he has been transfused with. Most sickle cell patients receive “phenotypically matched” blood that will reduce the chance this will happen.

Allergic reactions

These can cause rashes, hives, itching, or rarely, breathing problems. Rashes are very common; fever or breathing problems are much less common.

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. Your doctor will not start such a program unless you are fully informed about the reason for it. You should understand why this is necessary and agree that this is a reasonable therapy. 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 (desferrioxamine/Desferal) by needle under the skin at home every day to get rid of the extra iron or an oral medication (deferasirox/Exjade). The Desferal is given at night for eight hours using an infusion pump. He may also need to take the same medicine by IV when he comes to the hospital for transfusions. The Exjade is not to be taken with food and must be mixed in water or orange juice. It is taken once a day. This treatment is called chelation therapy. 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 three 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.



Screening all blood products thoroughly.

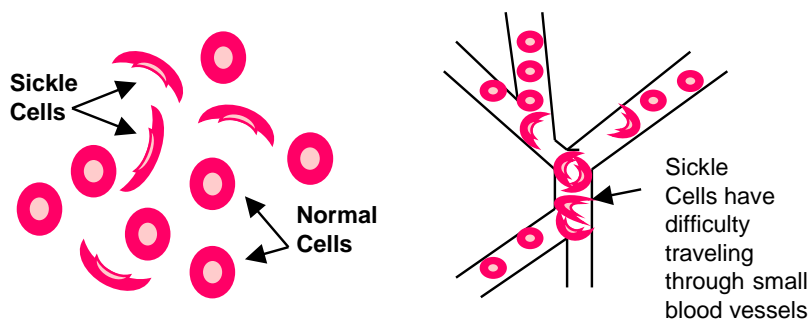
The Role of Stem Cell Transplantation in Pediatric Sickle Cell Disease

- Stem cell transplantation (SCT) is currently the only treatment option offering a chance of cure in sickle cell disease (SCD).
- More than 500 SCD patients have received transplants worldwide.
- SCT has its own risks and benefits, which are best discussed with your hematologist and / or a specialist in transplantation.
- The goal of this information sheet is to introduce you to the role of stem cell transplantation for children with sickle cell disease so you are prepared to ask the right questions to understand SCT and how it might benefit your child.
- There is hope that, in the future, gene therapy will be available and will also provide a curative option for sickle cell disease. However, gene therapy is currently still in investigative phase.

THE BASICS

Sickle Cell Disease (SCD):

You probably already know many details about sickle cell disease through your experience with your child. Sickle cell disease is a condition in which hemoglobin, the molecule that carries oxygen in the blood, has an increased tendency to precipitate (clump) under stressful conditions thereby altering the shape of the red blood cell. As the affected cells travel through small vessels they can get “stuck” and “block” the blood flow. This prevents blood from flowing normally through vital organs and causes pain as well as damage to the organ. Pain crises or ‘episodes’, acute chest syndrome, stroke, and osteonecrosis (thinning of the bone) are some examples of complications from SCD.



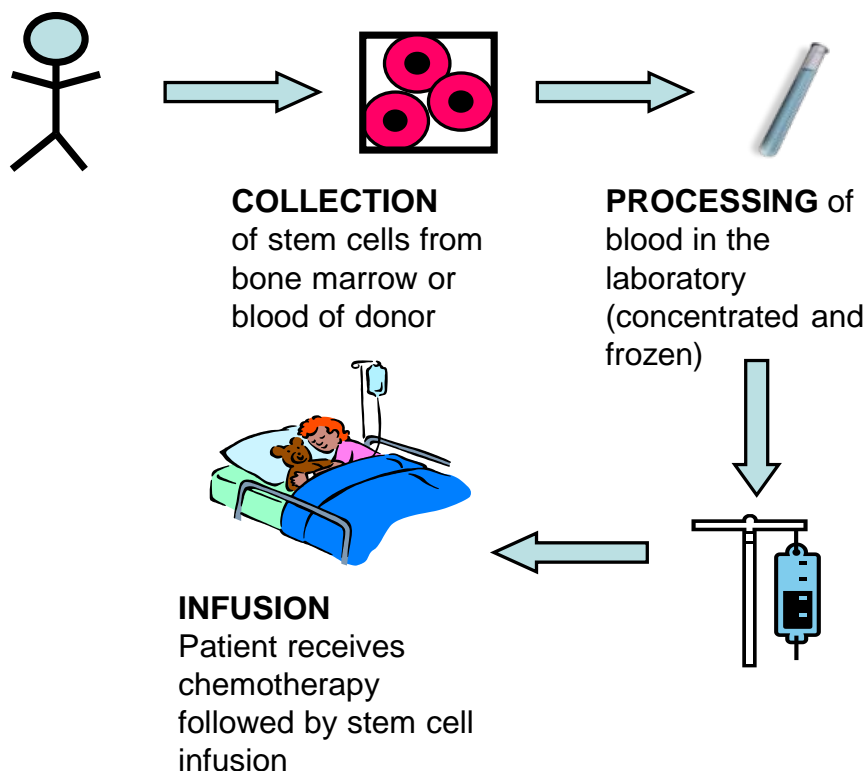
Tools for treatment:

Patients with sickle cell disease are managed with vaccines and may receive antibiotics, pain medications (including narcotics), hydroxyurea, or blood transfusions (both single and ongoing). These therapies have allowed children to live longer than in the past and allow most children to reach adulthood. This management is able to help symptoms and complications of sickle cell disease, but is not able to cure the disease itself.

STEM CELL TRANSPLANT

What is a stem cell transplant?

This is also called a “bone marrow transplant”. The bone marrow is the liquid portion of bones. It serves as a home for the stem cells. Blood stem cells are “mother cells” and have the capacity to create all other blood cells (red cells, white cells, and platelets,). During a stem cell transplant, stem cells are taken from the bone marrow of a healthy individual (donor), or sometimes from the donor’s blood, processed and infused (injected) into the recipient (patient).



The donor often goes into the operating room to donate, but the patient receives the cells similarly to a blood transfusion. From there on, the patient will have a new group of cells creating all of his or her blood cells. This means that red cells with normal hemoglobin will be produced and circulate in the blood stream. The main drawback is that in order for the patient to accept these stem cells, the patient's immune system has to be turned off for a few months through the use of chemotherapy. Eventually, as the stem cells grow, they form a new immune system and protect the patient from infection.

COMMON QUESTIONS

Who can be a donor a for stem cell transplant?

- **Sibling / Relatives:** The preferred donors are siblings who are “full match”. In brief, being a “full match” means that the siblings have inherited a similar set of immune system genes from their parents. The problem is that only 15-25% of children with sickle cell disease will have a sibling that is a “full match”. Parents and other relatives are even less frequently found to be a match.
- **Unrelated volunteer:** A second option is to check the National Registry for a volunteer donor. About 60% of patients with sickle cell disease are found at least one potential donor that is a “full match”.

What is a cord blood transplant?

- Stem cells are also present in the blood of a newborn's umbilical cord. This is blood that normally gets discarded after the baby is born. However, cord blood can be collected upon delivery of the baby and stored for future use. After processing, it also can be used for stem cell transplant. For details on how to have cord cells saved in future pregnancies visit: <http://www.marlow.org> or call 1 (800) 627-7692.

What if I want my child and his/her siblings checked to see if they are a “full match”?

- Some states offer screening (called “HLA-screening”) free of charge for children with sickle cell disease and their siblings. If you are interested, ask your hematologist during your next appointment or visit <http://www.marlow.org> to find more information.

Information for Babysitters and Childcare Providers

My child has **Sickle Cell Disease**. This is a condition that affects red blood cells and causes anemia.

You should call me immediately if you notice any of the following symptoms in my child. These are all times when my child needs to see a doctor RIGHT AWAY.

- Fever of **101° or higher (Never ignore this!)**
 - Severe headache or dizziness
 - Severe pain or swelling in the belly
 - Rapid breathing, or coughing with chest pain
 - Very pale skin or palms or inner eye lids
 - Cannot move hands, arms or legs
 - Limps without pain
 - Cannot wake up
 - Slurred speech or drooling
- **If you cannot reach me, you should call 911.**
 - **When you call the doctor or 911, make sure to tell the person that answers that my child has Sickle Cell Disease.**

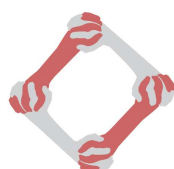
There are things that I need to know about right away so that I can call my child's doctor for advice. You should call me if my child:

- Vomits or has diarrhea
- Keeps coughing
- Has pain
- Is not acting like they usually do:
 - Refuses to take penicillin
 - Is less active than usual
 - Refuses to eat or drink

Here are some other things you need to know about my child. I will talk with you about these things:

- My child needs to drink plenty of fluids and have healthy meals and snacks.
- My child needs to get enough rest.
- My child needs to avoid temperature extremes (too cold or too hot and humid). It is important to dress my child in warm clothes when the weather is cold.

Please provide your babysitters, childcare providers and family members with this important information.



**National Coordinating
and Evaluation Center**

Sickle Cell Disease and Newborn Screening Program

CONTACT INFORMATION

You can reach me at _____

If you cannot reach me, call _____ (Name)

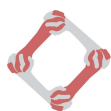
_____ (Phone Number)

My child's doctor is _____ (Name)

_____ (Number)

Other notes

Credits



National Coordinating
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Sickle Cell Disease and Newborn Screening Program

Sickle Cell Disease and
Newborn Screening Program



SCDAA National Headquarters 2008

Uade available through grant number U38MC00217-07-03
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 - Cannot wake up
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- **If you cannot reach me, you should call 911.**
 - **When you call the doctor or 911, make sure to tell the person that answers that my child has Sickle Cell Disease.**

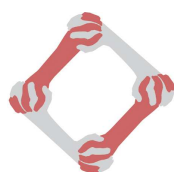
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_____ (Phone Number)

My child's doctor is _____ (Name)

_____ (Number)

Other notes

Credits



National Coordinating
and Evaluation Center
Sickle Cell Disease and Newborn Screening Program

Sickle Cell Disease and
Newborn Screening Program



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Authors' Notes

Living with sickle cell disease is challenging. We encourage you to be proactive and learn about all treatment options for your child. The more prepared you are, the more you can do to prevent complications. **Go ahead, look up information and ask your Hematologist!**

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