A Parents' Handbook For Sickle Cell Disease

PART II

Six to Eighteen Years of Age

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Referral Components For Pediatric Comprehensive Sickle Cell Services In Virginia

Central		Contact Numbers
Virginia Commonwealth University Health System **Department of Pediatrics** Division of Hematology/Oncology Kamar Godder, MD, – Consultant P.O. Box 980121 Richmond, Virginia 23298-0121	Phone: Fax:	P: (804) 828-9605 F: (804) 828-0504
(Professional and parent education) **The Virginia Sickle Cell Awareness Program** Jene Radcliffe-Shipman, Program Manager Division of Women's and Infants' Health Virginia Department of Health 109 Governor Street Richmond, Virginia 23219	Phone: Fax: E-Mail	**Jene R. Shipman, Program Manager** (804) 864-7769 (804) 864-7771 jene.radcliffe-shipman@vdh.virginia.gov
Eastern		
Children's Hospital of the King's Daughters Department of Pediatrics Division of Hematology/Oncology William C. Owen, MD – Consultant 601 Children's Lane Norfolk, Virginia 23507-1971	Phone: Fax:	(757) 668-9783 (757) 668-7811 **Terri J. Forsyth, RN, MSN, LPNP** **Maureen Fountain** 757-668-8260
Northern		
Pediatric Hematology and Oncology of Northern Virginia Jay Greenberg, MD - Consultant 8301 Arlington Boulevard, Suite 209 Fairfax, Virginia 22031	Phone: Fax:	(703) 876-9111 (703) 698-8338 **Janet Parry, RN** Case Manager affiliate with INOVA
INOVA Fairfax Hospital **Women and Children's Center** **Comprehensive Sickle Cell Center** 3299 Woodburn Road – Suite 220 Annandale, Virginia 22003	Phone: Fax: E-Mail	(703) 876-2715 (703) 876-2716 **Janet Parry, RN**
Western		
University of Virginia Medical Center Department of Pediatrics, Hematology/Oncology Peter Waldron, MD - Consultant P.O. Box 800386 HSC, University of Virginia Charlottesville, Virginia 22908	Phone: Fax: E-Mail	(434) 492-8499 (434) 924.5452 **Lindley Banner, RN, CPNP** LOB2Y@hscmail.mcc.virginia.edu **Karen Vest – Clinic Coordinator** Kev5w@virginia.edu
Carilion Roanoke Community Hospital Department of Pediatrics Division of Hematology/Oncology Joan Fisher, MD, Ph.D., Consultant 102 Highland Avenue, Suite 435 Roanoke, Virginia 24029-2946	Phone: Fax: E-Mail	(540) 985-8055 (540) 985-5306 JMFisher@carilion.com

A Parents' Handbook For Sickle Cell Disease

By
Ann Earles, RN, PNP
Shellye Lessing, MS
Elliott Vichinsky, MD
Editors
Children's Hospital - Oakland
Sickle Cell Center

With
Shelley Mann, MPH
Susan Karlins, MPH
Donna Bell Sanders, MPH
Joy DuVaul, MA, CHES
Education Programs Associates

And
Karen Whitney, MS
Kathleen Velazquez, MPH, CHES
California Department of Health Services
Genetic Disease Branch
George C. Cunningham, MD, MPH, Chief

PART II

Six to Eighteen Years of Age

Children's Hospital - Oakland Sickle Cell Center

Authors

Ann Earles, RN, PNP	Coordinator, Clinical Studies
Marsha Treadwell, PhD	Psychologist
Deborah Hurst, MD	Associate Director
Susan Fortune Pinheiro, MS	Genetic Counselor
Shellye Lessing, MS	Genetic Counselor
Joseph Telfair, DrPH, MSW, MPH	Social Worker
Fran Merriweather, MSW	Social Worker
Elliott Vichinsky, MD	Director

Illustration

Sue Parkinson

Design/Typesetting

Susan Stasi

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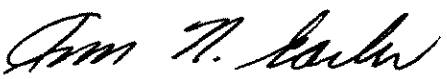
Preface

If you have a child with sickle cell disease who is between 6 and 18 years of age, this handbook is for you! This handbook is the second part of a series for parents of children with sickle cell disease. Part I was written for parents of children from birth to 6 years of age. This book, Part II, focuses on older children and teens.

By using this handbook and working together with your child's health care provider, you can make sure that your child gets the best care. If you have a special concern, use the Table of Contents to find the chapter that covers this concern. For example, if your 8 year-old child is having a problem with gallstones, you will find that Chapter 4 covers health concerns for school-aged children. Or, if you want to know more about school success for your teen, you will find it covered in Chapter 7. Once you find the right chapter, go through the pages until you find your concern.

We hope that you will share this handbook with others who are close to you. This can help them to learn more about sickle cell disease. Relatives, friends, teachers and your family doctor may have questions that this handbook can answer. The more they know, the more they can help you and your child. You don't have to manage all by yourself.



Ann Earles, RN, PNP

Elliott Vichinsky, MD
Editors

Children's Hospital - Oakland
Sickle Cell Center

Note: In some of the chapters in this book, your child will be referred to as male. In others, your child will be referred to as female.
ALL of the information applies to both girls and boys unless it is clearly stated.

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

Note To Health Care Providers

This handbook is the second part of a two-part series to assist parents of children with sickle cell disease. It was developed in response to requests from parents for more information about the care of their children.

"A Parents' Handbook for Sickle Cell Disease, Part II" covers the developmental years from school-age through adolescence. In addition to ongoing medical needs, the handbook addresses other areas at home, school and in the larger community that have been voiced as concerns by parents. The goal of this second volume is two-fold: first, to continue to support parents as partners in the care and the development of their children; and second, to help parents of adolescents begin to shift some of the responsibility of care to their teens.

The role that parents play in the care of their school-age and adolescent children with sickle cell disease is very important. The intent of the authors is that this handbook will be used by parents as a resource guide in conjunction with the care provided by a sickle cell center or a primary care physician treating a child with sickle cell disease.

Parents of children with sickle cell disease were involved in the development of this handbook. The formal field test with parents resulted in many comments and suggestions that were incorporated into the final version.

Table of Contents

Key Guidelines for Parents

Most children with sickle cell disease can look forward to a full life. It is important that they be prepared for adulthood and living on their own. School, friends, hobbies and work are all part of helping them grow up.

The outlook for people with sickle cell disease gets better every year. Research on how to treat and cure sickle cell disease is being done. We hope that we will make even more progress during the coming years. With parents, health care providers and researchers working together, children with this disease will be better able to enjoy a full life.

There are a few key guidelines that are important for all parents of children with sickle cell disease. These guidelines are briefly discussed in this section. They also appear in each of the chapters in this book. We hope that you will use these guidelines to help you be the best possible parent for your child.

Guidelines for Parents

- ◆ Prevent Problems
- ◆ Get Problems Treated Early
- ◆ Help Your Child Take Care of Himself
- ◆ Build Self-Esteem
- ◆ Take Care of Yourself and Your Family
- ◆ Get Expert Help When Needed

Key Guidelines for Parents

See Appendices A and B for answers to the questions "What is Sickle Cell Disease?" and "What Causes Sickle Cell Disease?"

Prevent Problems

The best way to help keep your child healthy is to prevent problems from getting started. There are many different ways to prevent problems. Here are some of the most important ways described in this handbook:

- Keep giving your child penicillin until your doctor says to stop.
- Take your child for routine check-ups even when he is healthy.
- Make sure your child gets all the shots he needs.
- If your child has sickle cell pain, help your child manage it at home.
- Follow your doctor's advice for care at home.

Get Problems Treated Early

Not all problems can be prevented. But most problems can be handled if they are found and treated early. Watch your child for danger signs and call or take your child to see the doctor when needed. Also, take your child to the doctor for routine check-ups.

Help Your Child Take Care of Himself

Children with sickle cell disease need to learn to do things for themselves. Sometimes, it is hard for parents of a child with a chronic illness to allow their child to grow up. They may feel like they need to protect their child all the time.

Start young. Help your child do things himself. As he gets older, allow him to do more things for himself. By the time he reaches the teen years, he will be able to handle most of his own care. Then, when he is an adult, he should be ready to live on his own.

Build Self-Esteem

There are many ways that you can help your child learn to feel good about himself. These ideas will be covered in more detail in this book:

- Listen to your child. Let him know that you value what he says and does.
- Praise him when he does something well.
- Spend time with him.
- Help him learn to do things that he can do well and enjoy.
- Expect success.

Children who have a chronic illness like sickle cell disease sometimes have poor self-esteem. Help your child see himself as a person, not just a disease. Make sure that you and others treat him as a whole person. Expect him to succeed. Help him overcome any problems that get in the way.

Take Care of Yourself and Your Family

You need to help your child with sickle cell disease stay strong and healthy. Your other family members also need you to help make sure that their needs are met. And you need to have a good life, too.

It is often hard to balance each person's needs. It is even harder when your child with sickle cell disease is having problems. Still, this is all part of your challenge. You may need to ask relatives or friends to help out with your family or give you a break.

Get Expert Help When Needed

It is a sign of strength to reach out for help. If you, your child or other family members are having problems, get outside help. There are many sources of help. You can go to relatives, friends or a parent support group. You can also see a social worker or counselor who knows a lot about sickle cell disease.

Set Limits for Your Teen

Groups Who May Offer Help on How to Parent A Teen

- Your local YMCA or YWCA
- Church groups
- Schools
- Local substance abuse prevention programs
- Support groups for parents of teens
- Support groups for parents of children with sickle cell disease
- Parents Anonymous

As the parent, you are still in charge. You have a right to know what your teen is doing and who your teen is with. It is your job to decide how much freedom to allow.

These limits should both protect your teen and support his growth. They tell your teen how you expect him to act. Limits can cover many issues, like curfew, homework, chores and use of the car. The rules should be fair and make sense. Discuss them with your teen. Better yet, see if you and your teen can set the limits together. Make sure the limits are clear to both of you.

If the rules are broken, you should respond as you would with any child. The disease is not an excuse for breaking rules. Your teen needs to be held responsible for his actions.

Sickle Cell Disease and Risk Taking

Like other teens, young people with sickle cell disease sometimes take risks. Many teens have a need to prove that they fit in with their friends. Drugs and alcohol, sex and fast driving are all things that some teens do to prove themselves.

Your teen needs your guidance and attention.

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

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

- Sex without condoms can bring a greater risk of getting a sexually transmitted disease (STD).
- Getting pregnant can be more of a problem for your teen.
- Alcohol can increase sickling because it dehydrates the body.
- Cigarettes can also increase sickling because smoke lowers the oxygen level in the blood.

All of these are risks for any teen. It is just that they can cause more harm to teens with sickle cell disease.

When to Step In

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

He also needs your guidance and attention. Listen to your teen. Find out what is behind his actions. Ask why he is taking risks, and listen to his answers. Tell him how they could hurt him and ask him what he thinks.

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

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

Build Self-Esteem

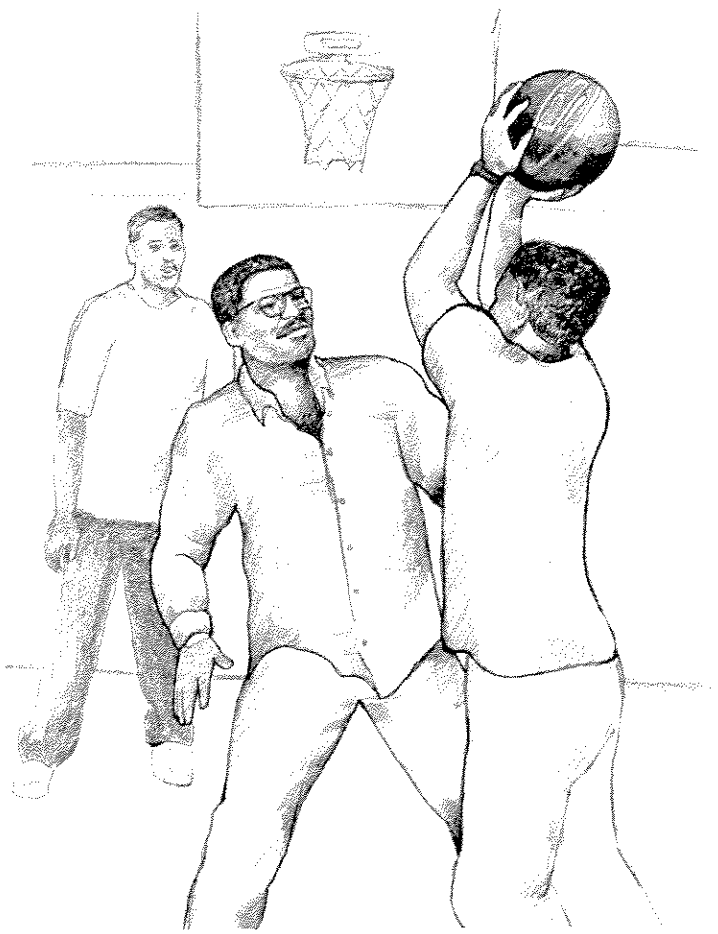
Like all teens, teens with sickle cell disease may sometimes have low self-esteem. There are many things you can do as a parent to help your teen feel better about himself:

- Listen to what your teen says.
- Don't put your teen down.
- Encourage your teen.
- Do things with your teen.
- Help your teen feel better about his body.
- Let him know how good you feel about him.

Listen To What Your Teen Says

Your teen needs to feel that you listen to him when he speaks to you. Even if you don't like what you hear, you can still try to understand what he is saying. A good way to let him know that you have heard him is to repeat what he has said. Once he knows he's been heard, you can then tell him how you think and feel.

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



Spend time together doing something you both enjoy.

Don't Put Down Your Teen

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

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

Do Things With Your Teen

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

Your teen may also want you to be involved with his health care sometimes. Check and see how things are going from time to time. Once you've stopped taking care of all your teen's health care needs, it can still be nice to help him out sometimes.

Help Your Teen Feel Better About His Body

Teens with sickle cell disease often feel ashamed of their bodies. Some look younger than their friends because they mature late. Some feel embarrassed by jaundiced eyes. Some have scars from surgeries and IVs. Besides the things that can be seen, some teens may also have limits on what they can do, like sports or dancing.

There are some things you can do to help your teen feel better about his body. If your teen is small for his age, help him find clothes that are right for his age and look good on him. If he has scars, help him find clothes that cover them up. You can also help him find easy ways to talk about his differences to others. If he can talk about them, he may not feel as ashamed.

Remind your teen that his body will mature, and he will get bigger. It will just take him a few more years than his friends. Also let him know that it is OK to be what ever size he is. Focus on his strengths and help him feel good about them.

Let Him Know How Good You Feel About Him

All of us need praise. Lots of it. What others say means a lot to us. If you tell your teen that he is worthwhile, he'll listen. If you tell him that you know he can succeed, it will give him a boost.

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

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

Encourage Your Teen

Treat your teen like he has something to offer, and he will. Help him get involved in things that he does well. Ask him about school, work or any special projects. Ask him to show you what he is working on. Your attention will show that you believe that what he does has value.

Praise your teen for a job well done.

Help with School and Future Plans

Staying in School

Many teens with sickle cell disease have missed a lot of school because of their illness. Some of these teens have managed to do well in school. Others have major problems. They may be behind their classmates or have poor study habits.

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

Urge your teen to stay in school. Help him keep up his drive to do his best. Encourage him to focus on what he wants to do in the future and to work towards it.

Your teen can arrange to make up school work he misses if he has to be out of school. Also, find out if the school has help for students with special needs. Help your teen get extra help if he needs it.

It may help him to have a model of success. If you know an adult with sickle cell disease whose life he might admire, tell him about what that person is doing. He might even want to meet him or her.

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

Your teen will need to decide whether he wants to go on to college. Many people with sickle cell disease go to college. Some colleges have programs to help students with special needs. If college is right for him, he can make it work.

Encourage your teen to plan for a full life. When he takes some control of his life, his image of himself will improve, and he will have a better chance for success.

Planning for the Future

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

A job can help a teen gain confidence in her skills.

Care for Yourself and Your Family

Other People Can Help You in Many Ways

Friends, relatives and neighbors can:

- Listen to you.
- Take you and your child to the doctor.
- Spend time with your other children when you have to give your child with sickle cell disease special care.
- Include your child with sickle cell disease in parties, outings and other social events.
- Tell you about places to get help or other resources.
- Comfort you when you're feeling down.

Even though you do all that you can to help your child stay well, she may still have problems. And these problems can affect your whole family.

As your child grows up, you may face new problems, like learning problems or pain. You may feel scared and angry again, even though you thought those feelings had left for good.

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

Your Other Children

Your other children need your attention and care, too. Make time to talk to them and be with them. Try not to miss school or sports events that they are involved in because you are focused on your child with sickle cell disease.

It can be helpful to teach all of your children about sickle cell disease. If they have questions that you can't answer, let them ask the staff at the sickle cell center. Knowing more about the disease will help them feel included.

One of the keys to a healthy family is to treat your child with sickle cell disease like her brothers and sisters as much as possible. Try to use the same system of discipline and rewards with all of your children. Special treatment isn't good for any of them.

Ask for Help and Support

Chronic illness raises questions and concerns for all families. It can help to have someone to talk to about your concerns.

Family members can be a big source of help. Share your problems and successes with them. Grandparents are often the most helpful. If your family wants to help you out, let them.

You can also get help from your church, social worker or counselor. Or look for a support group with other parents of children with sickle cell disease.

Don't be shy about asking for help. If you hold on to problems for a long time, they can be harder to solve.

See Appendices K, M, and P for additional resources.

"My son said he wished he had sickle cell disease like his sister. I told him that he was special, too."

Plan Ahead for Illness

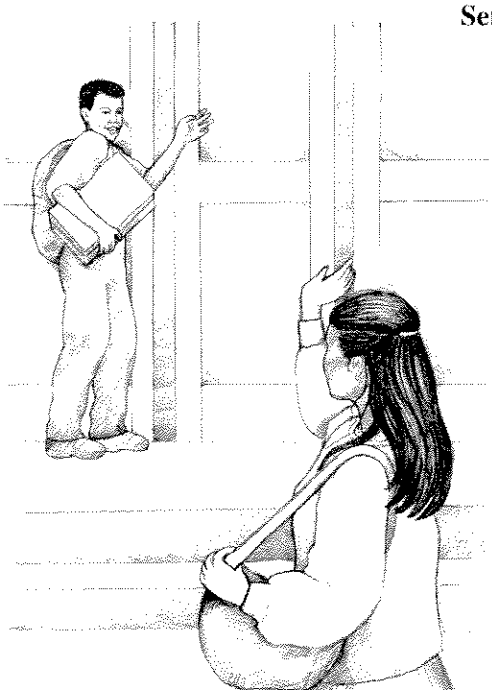
Set up a plan with your child's teacher for your child to do homework or make up the work that she misses if she gets sick. Talking about the plan will give you a sense of what the teacher thinks about your child's disease. It is a good time to give the teacher more facts about sickle cell disease. Most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

Find out what resources your school has for children who have a chronic illness. Ask your child's teacher if there is a hospital tutor program or other community programs that could be helpful if needed.

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

Keep Your Child in School

Send your child to school unless she is sick enough to see a doctor. Don't keep your child home from school if she just has something like a runny nose. She also doesn't need to stay home because of bad weather. Just make sure she wears the right clothes.



The more your child is in school, the better off he'll be.

Keeping your child home from school when she doesn't need special care will cause problems. She may be left out of friendships and have trouble learning social skills. She may also find it harder to do well in school.

If your child does have to spend time in the hospital, have her try to do her homework in the hospital. You can also encourage her to talk or write about what has happened to her in the hospital. If she misses much school, she may need outside help or a tutor so she can keep up.

Most of the time, it's best to send your child back to school as soon as she comes home. The more she is in school, the better off she'll be.

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

Expect the Best

Like other children, children with sickle cell disease can excel in school. Both you and her teachers need to expect the best from your child. When you expect more of your child, she will do better.

Some teachers may not expect enough from your child. Don't let a teacher protect your child from learning what she needs to learn. If your child is not doing well, she should get help.

School Success

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

Be a Partner with Teachers

At the start of a new school year, go to school and meet your child's teacher and school nurse. Bring your child with you. Let your child ask any questions that she may have and help her feel at ease. Tell the teacher about sickle cell disease. Give the teacher things to read so she can learn about the disease.

The teacher needs to know that your child will come to school even when she has minor aches and pains. She should be sent home only if she has a fever or severe pain or if she needs to see a doctor.

Explain your child's special needs. She needs to:

- Get water when she is thirsty.
- Go to the bathroom as soon as she feels the need.
- Make up school work if she has to miss school.
- Rest or slow down if she is tired or sore. For example, during gym class, she may only be able to run 2 laps, not 6.
- Rejoin the class as soon as she is ready.
- Get medicine if she needs it.

Check to see that her teacher gives your child what she needs. Some teachers may protect your child too much while others may ignore her. Talk to the teacher about these things if you are concerned. If you need help or support, talk to your doctor, nurse or social worker.

Stand Up For Your Child's Rights

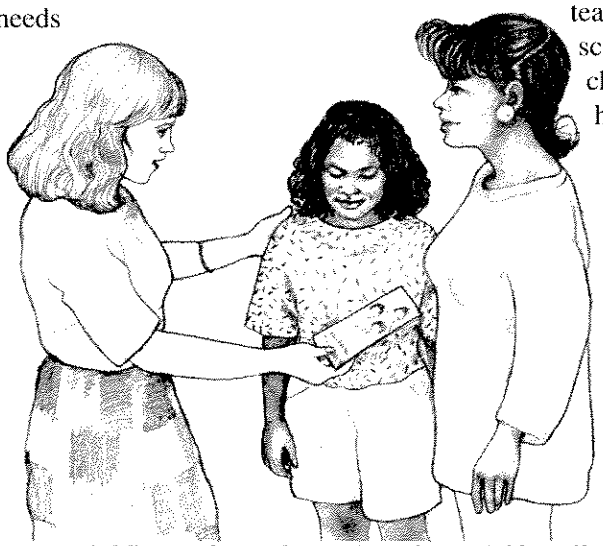
Your child has the right to get an education that meets her needs. There is a law which says that the school has to give it to her. This law (PL94-142) means that the school must provide help if your child needs it.

If your child is not doing well in school, talk with her teachers. You may want to ask that she be tested for learning problems. If she has a learning problem, she should get special help so she can learn better. Ask the school counselor for an "individualized educational plan (IEP)." This plan could include any of these:

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

If a teacher suggests that your child be held back, get an opinion from another learning expert. Often, this is not helpful with learning problems. Make sure that your child is tested for these problems and gets any extra help she needs.

Tell your doctor or a social worker if you don't think your child is getting enough help. They can help you work with the teacher and school so your child gets the help she needs.



Give your child's teacher information about sickle cell disease.

Self-Esteem

School Sports: For Boys and Girls

One of the ways that many children build self-esteem is through sports. If your child wants to play school sports, help her to be realistic about what she can do. Talk to your doctor about sports. Look at her strengths and find a sport that matches these.

If the sports program has a leader or coach, tell them about your child's special needs. Remind your child that she needs to take good care of herself. She needs to:

- Drink when she is thirsty.
- Rest when she feels tired.
- Dress for the weather.

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

- **Praise her when she does well.**

You can't give too much praise. Children thrive when they are told that they are special. They feel good when someone sees something they've done well.

- **Listen to what she has to say.**

Ask her questions. Show her that you care about what she thinks and feels.

- **Help her get involved in things besides her illness.**

She can try some of these things to see if she wants to do them:

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

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

Help your child try new things, like learning how to use a computer.

Accepting Herself

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

Make sure your child knows that she didn't get the disease because she was "bad." Nothing she did gave her the disease. Nothing she can do will get rid of it.

Your child needs to accept the fact that she has sickle cell disease and make the most of her life. Tell her that except for her disease, she is just like other children. Help her learn what she can and cannot do so she can gain more control.

Fitting In

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

Some children feel good about telling their friends about sickle cell disease. Others don't want anyone to know. It is good for children to tell at least one close friend about their disease. If no one knows, it is more likely that they will feel ashamed of their secret. When friends know, they can support your child when she needs it.

If your child doesn't know what to say to her friends, let her practice with you first. Her friends will handle it best if she tells them what the disease means and that they can't catch it from her. Once she has told others, she will probably feel better about herself.

Doing Things for Herself

Your child can learn to take care of herself, even though she has sickle cell disease. She needs to do things on her own. You still have to see that your child gets what she needs. But your job as parent is changing. You need to help your child learn to do things for herself.

By age 6, most children want to start doing things themselves. Your child may want to:

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

Besides the things she wants to do, she can also help out with the house. She can:

- Help with household chores.
- Clean up her room.
- Clear and set the table.
- Do the dishes.

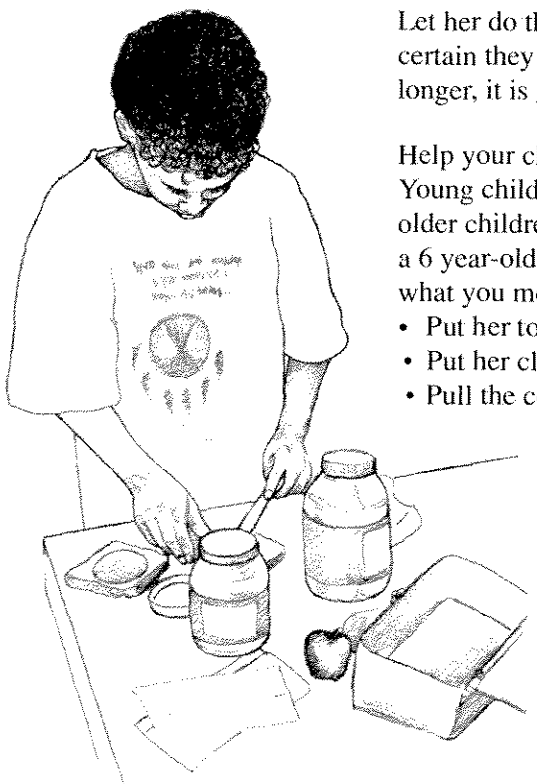
Let her do things herself unless you're certain they are not safe. Even if it takes longer, it is good for her to do them.

Help your child plan what she needs to do. Young children often need more help than older children. For example, when you ask a 6 year-old to clean up her room, explain what you mean. Tell her to:

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

By the time she is older, she should know what it means to "clean up her room."

If your child doesn't ask to do things by herself, you may need to push a little. Parents often find it hard to push a child who has an illness. They feel guilty or afraid. Don't let your child's illness get in the way of helping her to grow up. It's OK to let her fail sometimes. That is part of how we all learn what we can do.



Your child may be old enough to make his own lunch.

No matter what, don't do everything for your child. When she does things for herself, she will feel good about herself.

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

Sickle Cell Summer Camp

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

Sickle cell summer camp provides a safe and fun setting for your child to gain confidence and self-esteem. Spending time away from parents with other children her own age will help your child be more independent.

Ask your doctor or local sickle cell group about where you can get information about the nearest camp.

Guiding your Younger Child

While you need to care for your child's health, you also need to focus on more than her health care needs. When your child starts school, she begins a new stage in her life. She will spend a lot of time away from you, in the care of other adults. She may also spend more time with other children her own age. Both her social life and her school life may raise new questions or problems.

This is a time to help your child learn to do more things for herself. It is also a time to help build your child's sense of herself and to give her a good start with school. These are the same challenges faced by parents of all children, and the same guidelines apply.

This chapter will give you information about how to help your child with these issues:

- Doing Things For Herself
- Self-Esteem
- School Success

It will also help you look at the needs of your whole family and how you can take care of yourself.

Learning Problems

Any child, with or without sickle cell disease, can have learning problems. Learning problems can affect how well children do at school and in their social lives. Most children with learning problems have normal intelligence. They are just as smart as other children.

There are many different kinds of learning problems. Some children have a hard time taking in information. Others have problems understanding or remembering things. Some children have a problem with reading, writing or other skills.

If your child is having trouble at school or socially, work with the school and tell your doctor. If there is a concern, the doctor will refer your child for neuro-psych testing. This testing is done by a psychologist who has special training in learning problems.

Neuro-psych testing will help find out how your child learns.

Neuro-Psych Testing

To do this testing, the psychologist will talk to your child and ask him questions. Sometimes your child will be asked to read questions and write down the answers. Nothing is done to your child's body.

The goal of this testing is to find out how your child learns. The results will help show you and your child's teachers what kind of help he needs.

Help for Learning Problems

A child with learning problems can learn when he is taught in a way that works for him. Often, small changes in how a child is taught can help him do well in school. Also, teachers may be able to show him ways to use his strengths to learn better.

Make sure that you understand your child's problems and how he learns best. Use that information to guide you when you are teaching your child something. Also, stay involved with your child's teachers and school to make sure that they give him whatever help he needs to do well.

Treatment for Strokes

If your child has a stroke, he will need monthly blood transfusions. These transfusions help prevent more strokes. They are usually given for at least five years.

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

- Rehabilitation program, including physical, occupational and speech therapy
- Neuro-psych testing and care
- Desferal instruction

Treatment for minor strokes is often the same as treatment for other strokes. Transfusions are used to help prevent more strokes. If there are learning problems, steps can be taken to help him manage them.

Strokes and other brain problems can cause learning problems in some children with sickle cell disease. To find these learning problems early, all children should be screened at routine exams, starting at age 6.

Transfusions

A transfusion is given when a person needs more blood or a different kind of blood. There are two types of transfusions your child may need. Simple transfusions are the most common. For a simple transfusion, a person is given a set amount of blood through an IV. For an exchange transfusion, a person is given a set amount of blood while the same amount of blood is taken out of the body.

Each time your child needs to be transfused, his blood will be typed and cross-matched so he will get a good match. Complications are rare, but they do happen. This may include infections or a negative reaction to the transfused blood.

Your child may need monthly blood transfusions.

Kidney and Bladder Infections

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

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

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

If your child has a bladder infection, he will need to have his urine tested again from time to time. If the infection returns often, he will need to take pills every day so that the infections won't keep coming back.

Blood in the Urine

Another problem caused by sickle cell disease is blood in the urine due to bleeding from the kidney. When this happens, the urine usually looks bright red or brownish. In more severe cases, there may be back pain and small pink specks or lumps in the urine. The blood usually leaves the urine within hours. In some cases, the bleeding can go on for days and become a serious problem.

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

Strokes and other Brain Problems

One of the worst problems caused by sickle cell disease is strokes. Early treatment can help prevent serious damage.

A stroke occurs when part of the brain doesn't get as much blood as it needs. This is caused by sickle cells blocking a blood vessel in the brain.

Watch for these signs of a stroke:

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

If you see any of these signs, call your doctor and bring your child to the hospital right away. The sooner he gets help, the better.

Minor Strokes and Other Brain Problems

Minor strokes and other brain problems can happen without your child having any signs or symptoms. The only way to know that a small stroke has happened is through some special tests.

Minor strokes often cause learning problems. These learning problems can affect how well your child does at school.

Tests to Learn About The Brain

There are some tests which check whether your child is having problems with his brain due to sickle cells. The results of these tests will tell you if your child needs to take special steps to prevent strokes.

- **MRI (Magnetic Resonance Imaging)**

An MRI is done to look at the brain. Pictures of the inside of the brain are taken by a computer. It shows where an injury is in the brain. It usually takes about an hour and doesn't hurt. Young children may need to be given medication to relax while it is being done since they have to lie still for an hour. No X-rays or needles are used.

- **CAT Scan**

When an MRI is not available, a CAT Scan can be used instead. It uses X- rays to make a computerized picture of the brain.

- **Arteriogram (Angiogram)**

This test shows the blood vessels in the brain. A needle is placed in a large blood vessel, and dye is injected. The dye shows which blood vessels are blocked. Drugs are often given to help people relax during this test.

Problems with Kidneys and Urine

Dehydration

Sickle cell disease can damage the kidneys so that it takes more fluid to get rid of the body's wastes. Children with sickle cell disease drink more fluids and pass urine more often than other children. When a child becomes sick and drinks less than usual or loses more fluid by vomiting, diarrhea or fever, he can get dehydrated.

These are some signs of dehydration:

- Urinating much less than usual
- Difficulty urinating
- Burning when urinating
- Dry, sticky mouth and lips
- Sunken eyes

Sometimes, the skin may feel different. If you pinch it, it doesn't return to normal right away.

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

Your child should drink plenty of fluids.

Bedwetting

The need to urinate often can lead to bedwetting. This is common in many older children with sickle cell disease.

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

Priapism

Priapism happens when sickle cells block the blood vessels in the penis. This makes the penis hard and sore. This is different than a normal erection. It can happen at any age.

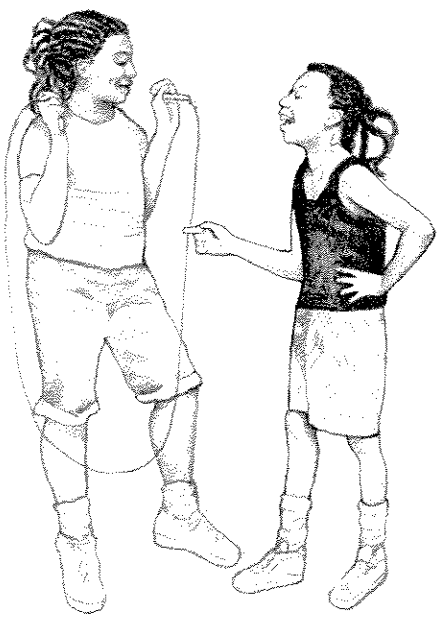
If the pain is mild, treat it like other sickle cell pain. If the pain is severe or it lasts more than a few hours, take your son to the doctor. He may need fast treatment with a blood transfusion, IV fluids and narcotics. Rarely, surgery may be required to clear the small blood vessels in the penis.

Even if the priapism goes away, tell your doctor about it. Your son needs to tell you if he has priapism. Boys often feel shy about telling their parents about this problem. Talk to your son about this before it happens. Tell him that he must tell you if his penis gets hard and sore, even if the pain stops.

Delayed Growth

Some children with sickle cell disease are small for their age. As they become adults, most children with sickle cell disease reach full size. If your child is smaller than his friends, tell him that he will most likely catch up. It will just take him a few years longer to grow.

Some children with sickle cell disease also reach puberty later than other children. Talk about this with your child early, before he brings it up. Help him find ways to feel good about himself now. Reassure him that he will mature in time.



Some children with sickle cell disease are small for their age.

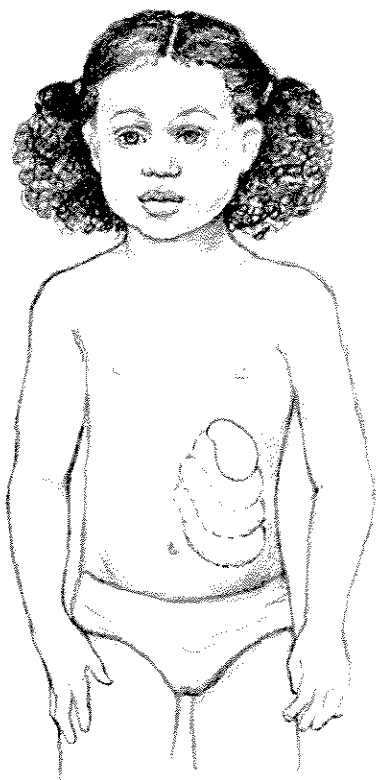
Spleen Problems

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

Enlargement and Scarring

Different types of sickle cell disease affect the spleen differently. With SS disease, the spleen often becomes very small by age 6, after being enlarged for a few years. Children with SC disease and S beta thal disease often have enlarged spleens for many years.

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



Your child's spleen may be larger than a normal spleen.

Splenic Sequestration

In some children, the spleen may become larger very quickly and begin to trap lots of blood cells. This "bleeding into the spleen" can cause the blood count to drop quickly. When this happens, it is called a "splenic sequestration crisis." It can lead to heart failure and death if not treated promptly with a blood transfusion.

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

If your child has one splenic sequestration, he will be more likely to have another one. His spleen may be taken out so that it can't happen again. He doesn't need his spleen to live. To help prevent infections, he should keep taking penicillin twice a day until his doctor tells him to stop.

Types of Sickle Cell Disease

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

Gallstones

About a third of children with sickle cell disease have gallstones by the age of seven. Many others develop them later.

Gallstones are formed from the waste products of broken-down red blood cells. This collects in the gall bladder and forms thick sludge or stones. Gallstones are not harmful. But if they get stuck in the gall bladder duct, they can cause a serious infection. Emergency surgery is then needed to take out the gall bladder.

Signs and Treatment

Often, there is a warning before gallstones get stuck in the duct. When the stones pass through the duct, they may cause pain in the right side of the abdomen. If the gall bladder is taken out after this warning, serious problems can be avoided. When gallstones are stuck in the duct, a person's skin may become very yellow.

Your doctor may suggest surgery to remove the gall bladder before an emergency happens. Taking out the gall bladder is the most common surgery in people with sickle cell disease. Most people can get along well without a gall bladder. However, they may have trouble eating a lot of fatty foods at one time.

Anemia (Low Blood)

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

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

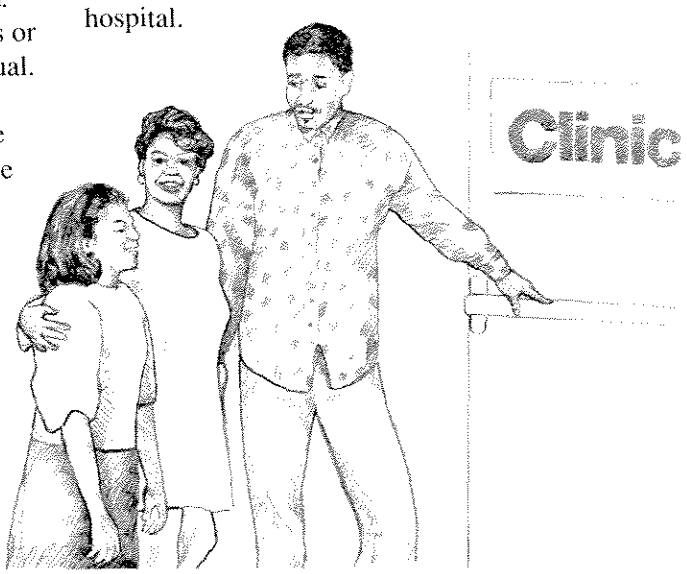
Signs

Bring your child to the doctor to have his blood count checked if you notice any of these signs:

- More tired than usual
- Pale color
- Loss of appetite
- Yellow eyes or skin
- Dark urine

Treatment

If your child's blood count falls very low, a blood transfusion may be needed. An extremely low blood count can result in heart failure and death if not treated in time. Since the blood count often falls when there are other medical problems, your doctor will usually check it daily if your child is in the hospital.



Bring your child to the doctor for a blood count.

Infections

If your child has a fever of 101°F or more, call your doctor's office.

As your child gets older, his body will get stronger. He will be able to fight infections better. He won't need to go to the hospital for infections as often. He can be treated at home and get well.

Still, some infections can cause problems. You still need to watch out for fevers. **If your child has a fever of 101°F or more, call your doctor's office.**

See Appendix I for a Temperature Conversion Chart. It shows you how to convert Celsius to Fahrenheit.

Meningitis

This is a severe infection of the membrane that surrounds the brain and spinal cord. It can be caused by a bacteria or virus.

These are the most common signs to watch for in older children:

- Fever
- Severe headache
- Throwing up
- Stiff neck (not able to touch chin to chest)

The signs are different in babies. They may be more irritable and tired than usual. Also, the soft spot on their head may bulge.

If the doctor thinks that your child might have meningitis, a spinal tap will be done to test the spinal fluid. If there is an infection, he will be put in the hospital right away and given antibiotics by IV.

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

Acute Chest Syndrome and Pneumonia

Acute Chest Syndrome is chest pain caused by sickle cells in the lungs or an infection. With an infection, it is called pneumonia. Low blood oxygen often goes with it.

Early treatment will keep it from getting worse. Watch for these warning signs:

- Fever
- Coughing
- Rapid breathing
- Shortness of breath
- Difficulty breathing or "grunting"
- Severe chest pain

If you see any of these signs, take your child to the doctor right away. Call first and describe the signs your child is showing.

Treatment is often given in the hospital. Your child will be given antibiotics and may need oxygen. A blood transfusion may also be required. Most of the time, this treatment makes people feel better.

Sometimes, the doctor will order a chest X-ray every day in the hospital as well as a few days after discharge. The doctor may also repeat lung tests after your child goes home to check on how well the lungs have healed.

Acute Chest Syndrome can be brought on when a child does not take deep enough breaths. This can happen when too much narcotic medication is given for pain or surgery. While taking drugs for pain, your child should expand his lungs by blowing into a balloon or an "incentive spirometer." This is a mouthpiece that shows how hard he is blowing.

Primary School Years:

Children between the ages of 6 and 12 can have most of the problems that affect younger children. The most common problems are:

- Infections
- Meningitis
- Acute Chest Syndrome and Pneumonia
- Anemia (Low Blood)
- Gallstones
- Spleen Problems
- Priapism
- Delayed Growth
- Problems With Kidneys and Urine
- Strokes and Other Brain Problems

In this chapter, we will describe these problems and how they affect primary school-aged children.

Different centers and doctors may use other approaches to treat these problems. Follow the doctor's advice. If you are concerned about what you are told, get a second opinion from another doctor.

Exercises for Older Children

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

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

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

Teaching Tips

- **Keep your voice low and soothing.**
Speak slowly, but not in a monotone.

- **Keep it natural.**
These are just outlines of what to say. Say what feels good to you. Soon, you will find your own words for the same things.

- **Work with a psychologist.**
It can be helpful to learn these exercises from a psychologist. He or she will pick the ones that will work best for your child and teach them to both of you.

- **Make tapes of the exercises.**
A psychologist can also make a tape of these exercises for your child to use as a guide. She can listen to it when she is in pain and can't relax on her own. You or your child can also make your own tape. Add music or a story if it helps.

- **Keep it positive.**
Your child does not have to try to relax all of the time. Make it enjoyable so that she'll be willing to try.

Self-Hypnosis

A psychologist can teach your child self-hypnosis. This is a more detailed relaxation technique that may be helpful to your child.

What To Expect

It is important for your child to practice these techniques when she doesn't have pain. This practice will help give her the best results.

Relaxation techniques can help your child manage pain. If she feels pain coming on, she should drink extra fluids, take Tylenol and relax in whatever way works best for her. Relaxation exercises and other home remedies alone will often take care of the pain. Other times, medicine may also need to be used.

Your child can listen to a relaxation tape.

Describing the Pain

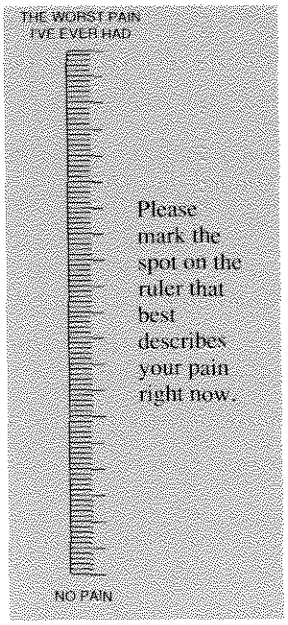


Figure 1. Pain Scale

You and your child must know how to describe her pain so she can get the right treatment. There are many ways to describe pain. Use the one that works best for your child.

- **How severe is the pain?**

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

- **Where is the pain?**

Her doctor will also want to know where she feels the pain. Sometimes it is easiest to describe where it is. Other times, it is helpful to mark the places on an outline of the body.

- **How does the pain feel?**

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

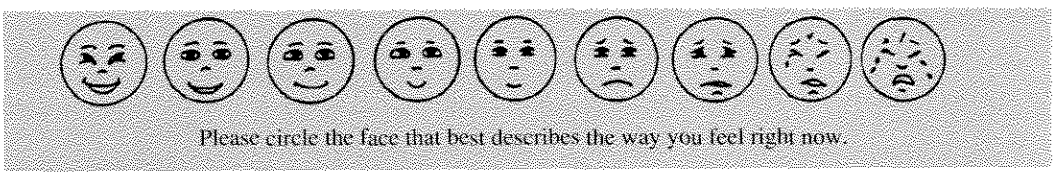


Figure 2. Pain Scale

Living with Pain

Pain Itself is not a Cause for Panic

While sickle cell pain hurts, it rarely causes serious problems. Most of the time, it can be managed at home. This pain is usually not a sign of something worse. It is part of the disease.

You and your child need to know when you should call your doctor with pain. Call your doctor if your child has any of these:

- Chest pain
- Severe headaches
- Severe pain in the belly
- No relief after trying "home remedies"

Not all pain may be caused by sickle cell disease. If you have questions about the cause of your child's pain, ask your doctor.

Get Help With Pain Problems

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

Don't let pain take over your child's life. Help your child find ways to ease her pain and to live with it. She will learn more about when she needs to manage her pain and when she can ignore it.

Teaching Your Child to Relax

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

Deep Breathing

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

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

You can help lead your child through relaxation exercises.

Exercises for Younger Children

Exercise #1: The Rag Doll

"Pretend that you are a robot (or wooden doll), all stiff and straight. Your arms and legs don't bend at all. They just stay straight."

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

Exercise #2: Noodles

"Pretend that you are noodles in a package that has not yet been opened. You are all stiff and straight." (Have your child hold this for a few moments.)

"Now you're cooked spaghetti, all over the plate. Are you covered with sauce or meatballs?"

Exercise #3: My Special Place

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

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

Narcotics

TENS (Transcutaneous Electrical Nerve Stimulation)

A TENS unit may help block the pain. It is a small device prescribed by physical therapy. Your child can use it at home when she starts to feel sickle cell pain.

A TENS unit may help to relieve pain.

If your child goes into the hospital for severe pain, she will most likely get narcotics. The most common ones are codeine, Demerol and morphine.

If your child needs one of these narcotics, you will get a prescription from the doctor for a small amount. These drugs should not be stockpiled at home. Also, these drugs (like all medicines) should not be shared with other family members.

Getting the Right Dose

The right dose of narcotics can be very helpful with severe pain. If your child's pain is not better, she may not be getting enough. On the other hand, too large of a dose can cause problems, like sleepiness or pneumonia. If you are concerned about how much pain medicine your child is getting, talk to the doctor.

Side Effects

Narcotics have some side effects. Often, they cause constipation. If this happens, give your child something to help soften her stools, such as prune juice. Feed her a diet high in fiber, with lots of fruit, whole grains and beans. If the constipation goes on, call your doctor for advice. Other side effects include itching and mood changes. Narcotics can cause serious problems, but these are rare.

Narcotic Use Does Not Cause Addiction

Narcotics for short-term sickle cell pain are not addicting. Many parents worry about whether their children will get addicted. Short-term use of narcotics to relieve pain will not lead to addiction.

Addiction Stems From Other Problems

Most children with sickle cell disease are not more likely to abuse drugs than other children. All children, including those with sickle cell disease, are at risk of drug abuse.

In most cases of addiction, other problems besides sickle cell pain are involved. Narcotics may be abused by a teen or adult who is having problems with home, school or friends. When this happens, it is likely that other drugs would be used if the narcotics weren't around.

Prevention Begins at Home

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

Chronic Pain Syndrome

Narcotics are only for acute pain. They are not meant to be used for chronic, long-term pain. People with chronic pain need to learn other ways to control their pain besides taking pills.

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

Easing Pain at Home

Home Remedies

There are many things your child can do at home to ease sickle cell pain. She can try any of these:

- Drink lots of fluids.
- Rest or play quietly.
- Take a warm bath.
- Put a heating pad or warm, moist towel on sore places.
- Massage the place that hurts.
- Take Tylenol (Acetaminophen) or Advil or Nuprin (Ibuprofen).

Keep Your Child Busy

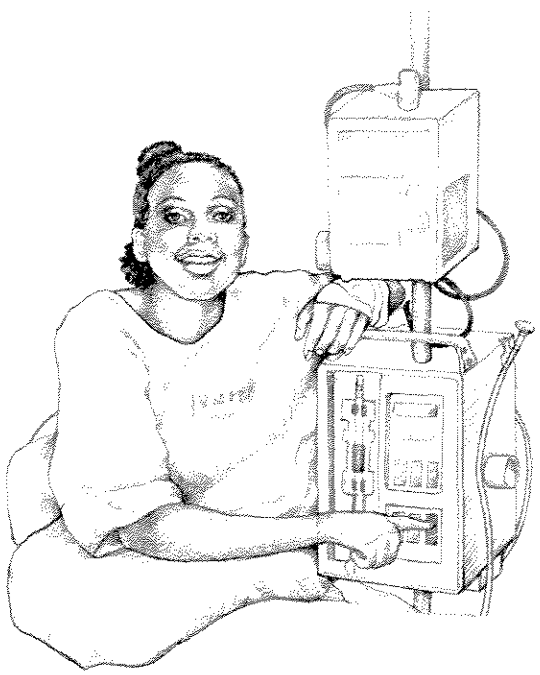
Keeping your child busy is an important way to deal with pain. She can talk to people, watch TV, play games or listen to music. Let her pick what she wants to do to keep her mind off the pain.

Try not to leave your child alone when she is hurting. Make sure that someone is with her to help keep her involved in other things.

Help Your Child Learn to Relax

Your child may hurt less if she is able to relax. You can help your child learn to relax when she is in pain. With practice, she may be able to stay calm and relaxed. Turn the page to find out how to teach this to your child.

Hospital Care for Pain



Your child can control her own pain medicine with a PCA pump.

At the hospital, your child may get fluids through an IV (in a vein). She may also get stronger pain medicine through an IV or a shot. This helps most children feel better quickly so they can go home. At home, the pain may last a few more days, but it should keep getting better. Again, use fluids, Tylenol, relaxation and other home remedies to help ease the pain.

If the pain does not get better, your child may need to stay in the hospital for a longer time. She will be given more fluids by IV and more pain medicine. Physical therapy may also be used to help with pain. This can include whirlpools, massage, exercise and heat packs. After 3 to 5 days in the hospital, your child should feel well enough to go home.

PCA (Patient Controlled Analgesia)

At some hospitals, your child will get a PCA pump so that she can control her own pain medicine. With a PCA pump, your child can decide when she needs more medicine. She presses a button to pump the medicine into her veins. The pump is set up so that she can't get too much medicine. This gives her better control of the pain in a safe way.

How to Deal with Pain

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

As soon as the pain starts, your child should drink lots of fluids and take Tylenol. She can also try other home remedies for pain.

If the pain gets worse or doesn't get better, call your doctor. Your doctor may ask you to find out if your child has a fever. The doctor may also tell you to give your child more to drink, to try heat packs or to use other medicines.

If the pain is too severe, your child may need to go to your doctor's office or the Emergency Room. Call your doctor's office before you take your child to either place so your child will be seen as soon as you get there.

This chapter will cover:

- Easing Pain at Home
- Hospital Care for Pain
- Narcotics
- Teaching Your Child to Relax
- Describing the Pain

When to Call the Doctor or Nurse

Call to Have Your Child Seen Right Away If Your Child Has One of These Danger Signs:

Fever:	101° F or higher
Head/Neck:	Severe headache or dizziness Stiff neck
Chest:	Pain or trouble breathing
Stomach:	Severe pain or swelling
Color:	Loss of normal skin color, very pale or gray
Penis:	Painful erection
Behavior:	Seizures Weakness or paralysis (can't move arm or leg) Can't wake up

If you can't reach the doctor, take your child to the Emergency Room.

Call For Advice If Your Child Has One of These Problems:

Stomach:	Vomits more than once Has diarrhea more than once
Fever:	100° F which lasts more than 24 hours
Color:	Jaundiced (eyes or skin look yellow)
Arms, Legs and Back:	Pain, with no other symptoms
Chest:	Coughs, without fever or chest pain
Nose:	Runny or stuffed nose
Behavior:	Acts strangely Refuses to take medicine Refuses to eat or drink Less active than usual

See Appendix H for a copy of this list to post in your home.

Drinking Fluids and Taking Breaks

Children with sickle cell disease need more fluids, bathroom breaks and rest than other children. When your child is away from home, it may be hard for him to take care of these needs. You or your child should tell teachers and other adults about his special needs so it will be easier for him.

Allow your child to take care of these needs himself as he gets older. Check to see how he is doing, but don't follow him around. Help him think of ways to make sure he rests when he is tired. Help him to understand that if he doesn't take good care of himself, he may have more problems.

Extra Fluids Are Needed:

- With a fever
- With pain
- When it is hot outside
- With a high level of activity
- When traveling

When your child needs extra fluids, the amount needed every 24 hours is based on your child's weight.

At 40 lbs: Drink 7 cups
At 50 lbs: Drink 8 cups
At 60 lbs: Drink 9 cups
Over 60 lbs: Drink 10 or more cups

A cup is 8 ounces of fluid. The fluids should be clear fluids, like water or bouillon.

Taking Trips

See Appendix G for a sample Travel Letter.

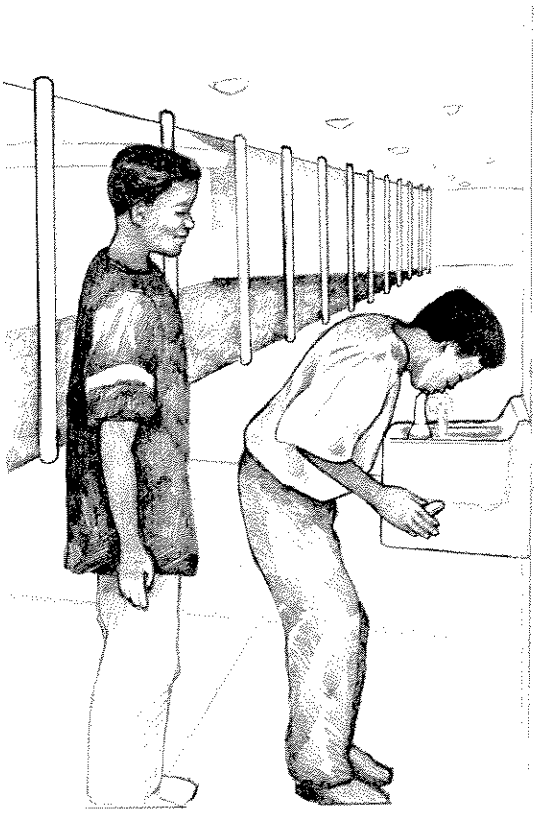
Most travel is fine for children with sickle cell disease. There are just a few rules for your child to follow which will help him stay healthy:

- Drink plenty of fluids while traveling.
- Fly only in pressurized planes.
- At elevations over 5,000 feet, drink extra fluids and rest often.
- Carry a "Travel Letter" from the doctor.

Watching for Danger Signs

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

Both you and your child can watch for early signs of problems. These danger signs are listed on the next page. Go over them with your child so he will also know what signs are important to share.



Your child needs to drink plenty of fluids.

Taking Care of Their Own Health Needs

Children must play an active role in meeting their health care needs. As they get older, their part will get larger and you will need to do less. By the time they are teens, they should be taking care of most of their own health care needs.

Primary School-Aged Children Can Do These Things:

- Learn the danger signs and tell you when they have one.
- Take their medicine (you still need to track how much they take).
- Speak up at doctor visits by asking questions and giving answers about themselves.
- Drink fluids when they are thirsty and at other times when they need more fluids.
- Take breaks and rest when they feel tired.
- Learn to manage mild pain by taking Tylenol, drinking extra fluids, using "home remedies," staying busy or relaxing.

Your teen can talk to a genetic counselor.

Teens Can Do These Things:

- All of the things that younger children can do.
- Call the doctor if they have a danger sign and tell you about their concerns.
- Take their own medicine and tell you how much they are taking.
- Take their own temperature.
- Go to doctor visits without you.
- Manage mild pain by themselves.
- Go to a teen support group.
- Talk to a genetic counselor.
- Keep you informed about what they are doing and how they are feeling.

Learning About The Disease

Talk to younger children about sickle cell disease so that they can tell others about it. Older children can write a report for school, read books and ask their health care providers for more information.

Taking Medicine

Younger children should know what medicine they take and be able to remind someone when they need it. Teens are able to take most medicine on their own. At any age, you need to keep track of the medicine your child takes.

Nutrition

Your child needs to make sure that he eats a well-balanced diet. Children with sickle cell disease need to eat good foods full of protein, vitamins and minerals every day. These foods will help them build new red blood cells. They also need to keep taking 1 mg folic acid every day.

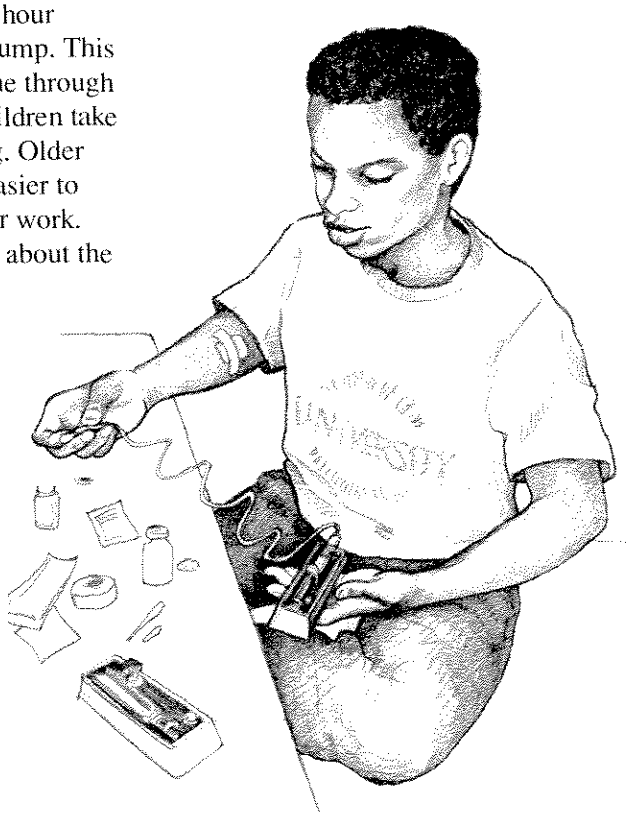
Desferal

Sometimes, children with sickle cell disease need monthly blood transfusions. Chronic transfusions prevent sickling, but they can cause iron poisoning. Desferal prevents and treats iron poisoning.

If your child has chronic transfusions, he must take Desferal. You will need to give your child the infusions until he is old enough to do it for himself. At age 10, most children are able to learn to infuse themselves. Make sure that your child is taught how to do this.

Desferal is given over an 8 to 10 hour period each day by an infusion pump. This pump slowly lets out the medicine through a needle under the skin. Most children take it at night when they are sleeping. Older children and adults may find it easier to take it during the day at school or work. You can give your child a choice about the timing.

As long as your child gets transfusions, he must take Desferal. Even after the transfusions stop, it can take awhile to get rid of all the iron. Your doctor will tell you when your child can stop using Desferal.



At age 10, most children are able to learn to infuse themselves with Desferal.

Taking Medicine at Home

Penicillin

Make sure that your child keeps taking penicillin until your doctor says to stop. Whether your child will take penicillin after age 5 depends on your doctor's evaluation. If your child has had a lot of infections or had his spleen taken out, he will most likely need it. Many doctors believe that all children should keep taking it. Research is being done now to find out how long penicillin is helpful.

If your child is on penicillin, he will take 250 mg. twice a day, morning and night. By age 6, most children should be able to take it in pill form. Since the liquid doesn't keep long, it is better for your child to take the pills. Still, a few children aren't able to swallow the pills and have to take the liquid.

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

Other Common Medicines

There are a few medicines that children with sickle cell disease often take at home:

- Tylenol (Acetaminophen) for fever and/or mild pain
- Advil or Nuprin (Ibuprofen) for mild pain
- Tylenol with codeine for severe pain (by prescription only)
- Folate (a vitamin pill taken once a day)

See Appendix F for Suggested Acetaminophen Dose by age.

Antibiotics

If your child has an infection, he will probably be given antibiotics. These must be taken until they are finished and there is no medicine left. Even if he feels fine, he needs to take the medicine until it is gone.

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

Antibiotics And Pain Pills Are Not The Same

Antibiotics must be taken for as long as they are prescribed. Even if your child feels better, he should keep taking the pills until all of them are gone.

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

NOTE: Don't forget to call your doctor if your child has a fever!

Routine Health Care and Home Care

Children with sickle cell disease need to take special care of their bodies. With special care, many problems can be prevented.

You can help your young child do what is needed to take care of himself. You can take him for well child exams and make sure he gets all the shots he needs. You can work with his doctor to make sure that he gets helpful referrals and tests. You can also make sure that he takes any medicine that he needs.

It is best to help your child do more now so that he will be able to take care of himself when he gets older. When your child is young, you will need to help him do many things. As he gets older, he will be able to do more and more things on his own. By the time he is an adult, he should be able to take care of all of his health and home care needs.

This chapter covers these health and home care topics:

- Well Child Exams
- Shots
- Common Lab Tests
- Using Other Doctors (Specialists)
- Taking Medicine at Home
- Taking Care of Their Own Health Needs
- When to Call the Doctor or Nurse

Let Your Teen Do for Himself

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

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

Starting To Let Go

Your teen may not take care of himself as well as you took care of him. Try to let him do it anyway. If you are afraid that he is hurting himself, talk to him about it. Explain what might happen and find out why he doesn't want to do something. Help him figure out a better way to take care of himself.

Now is the time to shift control from you to your child. You can start slowly, but keep moving towards giving him more control. You need to trust him to ask for help when he needs it. If he makes a mistake, help him learn from it. If he learns now, he can take charge of his own life as an adult.

Doing Other Things His Own Way

There are many ways for your teen to be independent besides caring for his disease. The way he talks, the clothes he wears, the music he likes, and the way he does his hair are all ways to express himself.

You may not like the way he looks or acts, but try to accept it. Unless he is hurting himself or others, try to leave him alone. He wants to be different from you. He wants to be himself.

Growing Up May Be Scary

Some teens with sickle cell disease are afraid to grow up. They don't want to take care of themselves and face their lives. They may try to act younger than their age. Since they often look younger, you and others may be tempted to treat them that way.

Don't let this happen. Expect your teen to act his age, not his size. Stop doing everything for him. If you don't do it all, he will find out that he needs to take care of himself.

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

Your teen wants to be herself.

Living with Your Teen

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

The teen years can be hard for families, with or without a chronic illness. There are also special problems for this age group that come with sickle cell disease. These problems can make these years a challenge for everyone.

Your teen will learn to take care of himself more and more during these years. He is getting ready for the time when he will be on his own. He is growing up and learning to live his own life. Your teen needs limits, but he also needs freedom. You and your teen will keep trying to find a balance that works.

As a parent, there is much that you can do to help your teen during these years. Each of these guidelines will be described in this chapter.

- Let Your Teen Do for Himself
- Set Limits for Your Teen
- Build Self-Esteem
- Help with School and Future Plans
- Get Help for Your Teen and Your Family

Aseptic Necrosis (Bone Changes)

This is the term for damage to the bones caused by sickle cell disease. When the hip bone is involved, it can cause chronic pain in the hip joint when a person walks.

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

Your teen will know if he has this problem. His hip will hurt when he walks or runs. Make sure he tells the doctor if he has hip pain. Early treatment helps.

The treatment depends on the extent of the problem. Sometimes a person needs to use crutches for a few months to take the weight off the joint. Other times, your doctor may suggest surgery or transfusion to stop the hip from changing shape.

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

Appearance

Late Puberty

Some teens with sickle cell disease reach puberty about two years later than others their age.

Puberty means many changes. These changes are the same for teens with and without sickle cell disease. The only difference is when they happen. For girls, puberty is when their breasts grow and their periods start. In boys, their face hair grows, their muscles get bigger and their voices deepen. Puberty also means being able to get pregnant or to make someone pregnant.

Late puberty is not a problem in itself. But it can make your teen feel bad about himself. Talk to your doctor or a counselor if you or your teen feel concerned about this issue.

Smaller Size

Some teens may be small and thin for their age. Children with sickle cell disease catch up late in their teen years. If your teen is very small or thin, talk to your health care team. Usually, nothing is needed. In a few cases, a special diet or hormones may be helpful.

Yellow Eyes

People with sickle cell disease can have yellow eyes from time to time. This is caused by a yellow colored substance called "bilirubin" that comes from broken-down red blood cells. In some people, the yellow tint lasts for a long time. It may always be there. It is not a medical problem unless the eyes are much more yellow than you've seen before. If the color is much stronger, call your doctor to see if your teen needs to be checked.

What Can Be Done

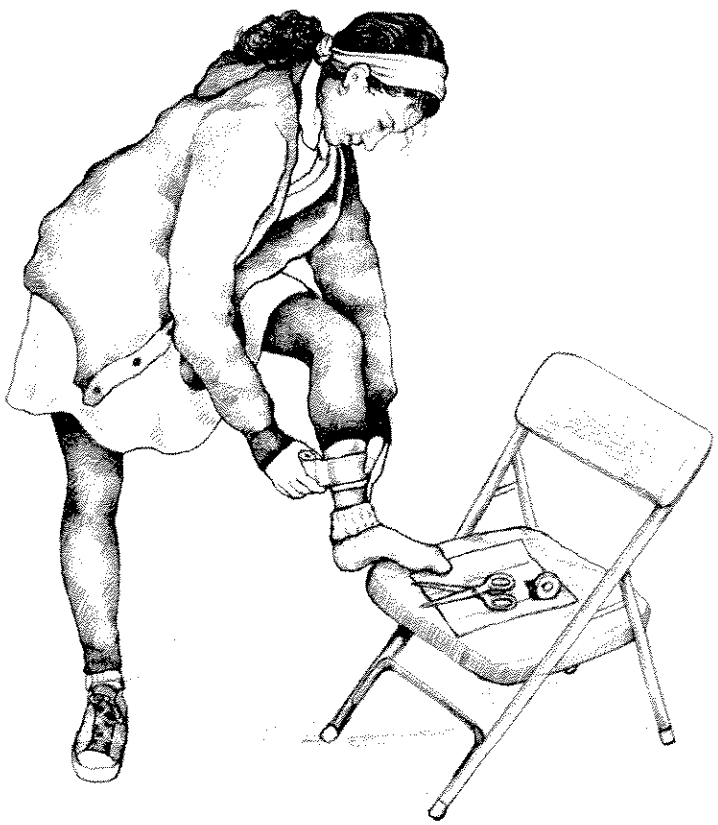
If your teen is concerned about his looks, reassure him. You and other family members can support him in feeling better about himself. It may also be helpful for him to talk with a counselor or other teens with sickle cell disease about his feelings.

Leg Ulcers

Ulcers usually start as a small sore on the ankle. They can grow large and get infected. Some ulcers heal quickly, while others can take a long time to heal.

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

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



Put on a fresh bandage every day.

Signs of Ulcers

Take your teen to see the doctor if you see either of these signs:

- **A cut or wound that doesn't heal**
- **A patch of dry, itchy skin**

It is much better to treat leg ulcers when they are small than when they are larger.

Treatment

These are the basics to good treatment for ulcers:

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

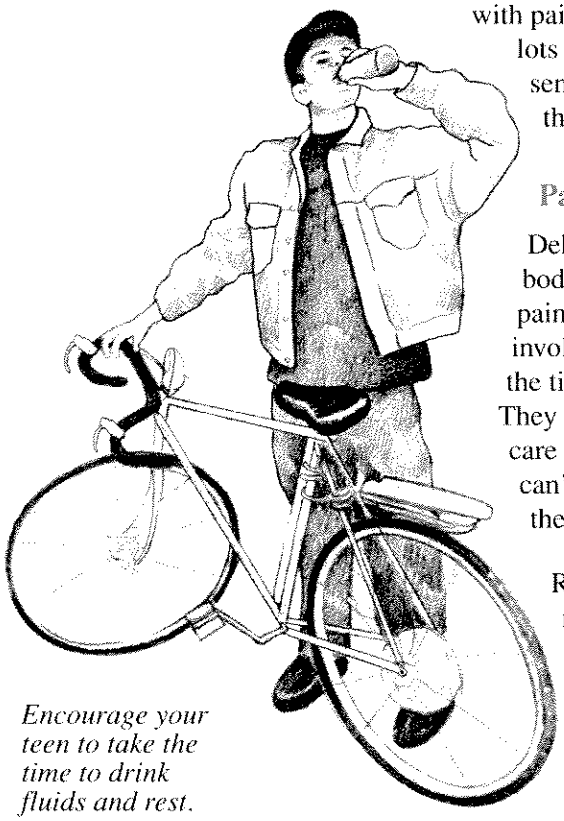
If the ulcer is large or has not started to heal in a few weeks, your child may need to go into the hospital. There, he will be on strict bed rest and get special wound care. Transfusions may also be used to try to bring more oxygen to the tissues.

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

Pain

Some teens with sickle cell disease have more pain as they get older. The pain may feel worse or just come more often. The treatment for pain is the same for teens as it is for children. (See Chapter 3 for more about pain.)

By now, if your teen has had a problem with pain, he has most likely tried lots of ways to ease it. It makes sense for him to use the ones that work best for him.



Encourage your teen to take the time to drink fluids and rest.

Pain Triggers

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

Rarely, girls get severe menstrual pain which brings on sickle cell pain. The doctor may be able to prescribe a hormone to prevent this pain.

Your teen will learn more about what triggers his pain. As he does new things, suggest that he watch to see what happens with the pain. If he finds that something often leads to pain, he can stay away from it or take special care with it.

Pain Contracts

For teens with chronic pain, a written contract may be helpful. The contract is an agreement about how pain will be handled. These contracts are often used in the hospital, but can be helpful at home.

Pain contracts have two parts:
1. The first part says what the health care staff will do to help the teen manage his pain.
2. The second part states what the teen will do for himself to manage the pain.

See Appendix L for a sample Pain Management Contract

NOTE: If a teen has a fever with pain or if the pain is in his stomach or chest, he should see a doctor. Call first so he can be seen right away.

Eye Problems

Sickle cell disease can cause eye damage and, rarely, blindness. This is why your teen needs to be checked by a special eye doctor (an ophthalmologist) once a year. By the time your teen complains of poor vision, the changes may have gone too far.

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

The Teen Years:

Teens can still have many of the same problems which younger children have from sickle cell disease. Infections, pain, low blood counts and strokes can affect people with sickle cell disease at any age.

Some of these problems are less dangerous for teens, like infections. Most infections can now be treated with pills at home, not an IV in the hospital. Still, your teen needs to see a doctor for a fever over 101°F.

Here are the most common health problems that teens with sickle cell disease face:

- Pain
- Eye Problems
- Leg Ulcers
- Aseptic Necrosis
- Appearance
- Infections
- Gallstones
- Acute Chest Syndrome
- Priapism

Many of these problems have been described in detail in earlier chapters. Pain and the last four problems will be described in this chapter.

Job Worries

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

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

- When you allow him to take care of himself, you are preparing him for the future.
- When you help him feel better about himself, you are raising his chances of success.
- When you help him do well in school as well as get involved in other interests, you are giving him the best training for future work.

Help for Teens

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

Places for Teens to Get Help With School or Work

- School counselor
- Vocational counseling programs
- Educational Consultants
- Tutoring Programs
- Guidance clinics
- Churches
- YMCA or YWCA

Help your teen do her best.

Get Help for Your Teen and Your Family

Get Help for Your Teen

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

- Your teen is angry a lot or taking dangerous risks.
- Your teen seems very depressed or anxious.
- Your teen won't take care of his health.
- Your teen is doing poorly in school.

Get Help for Your Family

Families often have tough times with teens. When any of these things happen, reach out for help for your family:

- You and your teen feel like you can't talk to each other.
- Your other children are very upset about the way your teen is acting.
- Someone close to your teen gets divorced or dies.

Get Help For Yourself

When teens have problems, parents have problems. Getting help for yourself is often the best way to help your teen. With support and guidance, you will be better able to give your teen what he needs.

Get Help Before Problems Get Worse

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

Where to Go For Help

Find a counselor who knows about sickle cell disease and chronic illness in teens. Most sickle cell centers have counselors on their staff who know a lot about the disease. Or ask your doctor or nurse who they think would be most helpful to your teen or your family. You may also be able to get help through your church.

Many cities have support groups for parents of children with sickle cell disease. Some large cities also have support groups for teens with sickle cell disease. Ask your doctor if there is a program like these that you or your teen can join. Parents and teens can gain a lot from talking with other people like themselves.

See Appendices K, M, and P for additional resources.

Find a counselor who knows about sickle cell disease.

Sex and Reproduction

In the teenage years and through adulthood, people face choices about sex and having children. It is not always easy to know what is right to do. This is true for both children and parents.

People with sickle cell disease face the same challenges as other people. They need to decide when they want to have sex. They also need to choose if they want to have children. There are some differences in how these choices may affect their health. These differences will be discussed in this chapter.

Parents can help their children face these important questions. You can talk to your children so that they know the facts. You can also share your feelings and values to help guide their choices. This chapter will help you talk to your child with sickle cell disease about these issues. It will also inform you about the impact of the disease on these areas of your child's life.

This chapter will address:

- Talking to Your Teen About Sex
- Birth Control Basics
- Sexually Transmitted Diseases and AIDS
- Pregnancy

Sex and Teens

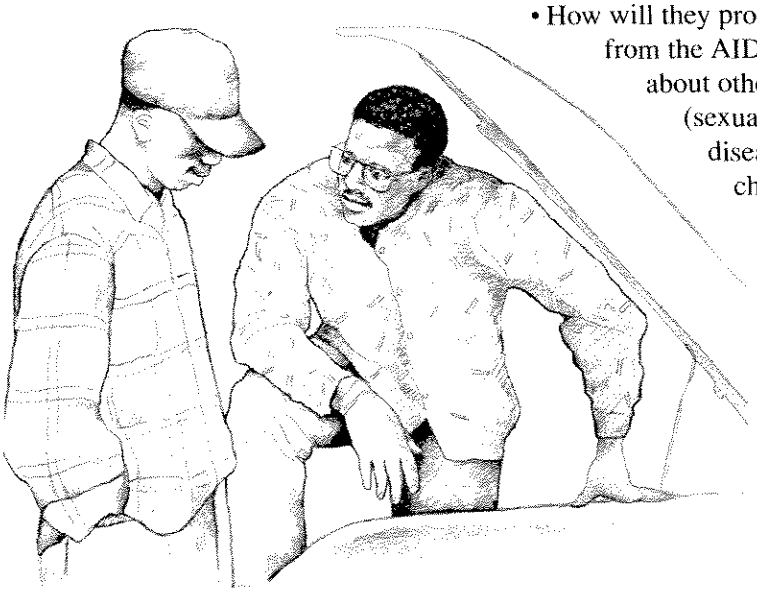
Like all teens, teens with sickle cell disease face challenges with sex. As they reach puberty, their bodies go through many changes. They begin to have strong sexual feelings. Girls are able to get pregnant, and boys are able to make someone pregnant. All teens have choices to make which will have a major impact on their lives.

Questions for Teens To Answer

Here are some of the questions which teens need to answer for themselves:

- How will they respond to their sexual feelings?
- When will they have sex? With whom? Will they wait to have sex until they are married?
- Do they want to have children?
- If they have sex but don't want children now, what kind of birth control will they use?
- How will they protect themselves from the AIDS virus? What about other STDs (sexually transmitted diseases) like chlamydia or genital warts?

These are not easy questions. Teens need to give these questions a lot of thought and make good choices for themselves and their future families.



Listen to your teen and share your values.

Concerns With Sickle Cell Disease

Teens with sickle cell disease may have concerns about maturing late. Boys may wonder whether they can have sex if they have not reached puberty or if they have a problem with priapism. Boys and girls may worry about whether they will be able to have children.

These concerns can add to normal fears about dating. Teens with sickle cell disease often worry about whether they will find a boyfriend or girlfriend. Having a good time with other teens of both sexes may ease some of these worries.

Teens need to be reassured that they will be able to get pregnant and have sex like other people. Remind them that getting pregnant or having sex are not good ways to prove that they are OK.

Talk To Your Teen About Sex

You don't have complete control over what your teen does about sex, but you can talk to her. You can give her the facts and help her make sense of all that she has heard. You can share your feelings and values so that she knows where you stand. You can listen to her feelings and thoughts and help her decide what is best for her.

Don't wait until your child becomes a teen to start talking about sex. Your child may ask you questions about these issues. Answer what she asks. You don't have to explain everything at once. If your child doesn't ask you about sex, bring up the subject. Look for times to ask about her thoughts or feelings and to share yours. It could be while watching TV or when you hear or read something about sex or love.

Don't assume your teen knows it all even if she acts like she does. Research shows that teens who know the **least** about sex are more likely to have sex sooner. More often than not, it is what teens **don't know** about sex that can hurt them.

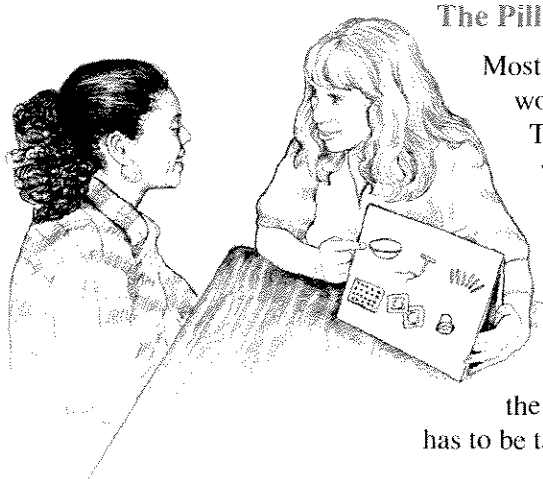
Birth Control Basics

People with sickle cell disease can have children. Girls who have reached puberty can get pregnant. Boys past puberty can get a girl pregnant. If they choose to have sex and don't want children, they need to use birth control. There are many kinds of birth control available. Only the IUD (intrauterine device) should not be used by women with sickle cell disease.

Condoms and Foam

One of the best forms of birth control for people with sickle cell disease is the condom. Condoms are good because they also help protect a person from the AIDS virus and other STDs.

Condoms don't always keep a woman from getting pregnant. For more protection, many women also use a special foam with a condom. This foam is put in the vagina before sex. It kills sperm that the condom doesn't stop.



There are many choices of birth control methods.

The Pill

Most **low-dose** pills are safe for women with sickle cell disease. These pills almost always keep a woman from getting pregnant, but they don't give protection from STDs or the AIDS virus. Some couples use both condoms and pills.

A prescription is needed to get the pill. Once the pill is started, it has to be taken every day.

Norplant

Norplant may also be a good choice for women with sickle cell disease. It is made up of 6 thin, soft, small capsules that are placed in the upper arm under the skin. These capsules slowly release a hormone into the body.

Norplant almost always keeps women from getting pregnant. It does not protect them from the AIDS virus or other STDs. Women who want to use Norplant need to talk to their doctor about whether it is right for them.

Depo-Provera (DMPA)

Depo-Provera is a shot given every 12 weeks. The hormone in it keeps women from getting pregnant. Like Norplant, it almost always keeps women from getting pregnant. It does not prevent STDs or the AIDS virus. Women should ask their doctor about whether it would be a good method for them.

No IUDs!

The IUD is not a good method for women with sickle cell disease. It can lead to pelvic infections which can cause many problems.

Choosing a Birth Control Method

There are other choices besides condoms and foam and the low-dose pill. A woman can also use a diaphragm, sponge or cervical cap.

Your teen should talk with her partner and her health care team about what method of birth control to use . Then she can choose what will work best for her.

Using Birth Control

Planned pregnancies are very important with sickle cell disease. Without planning and special medical care, pregnancy can be dangerous for the mother and the baby. If your teen is going to have sex and she isn't ready for children, she needs to use birth control.

STDs (Sexually Transmitted Diseases)

For More Information

If you or your teen need more information about AIDS or other STDs, ask your doctor. You can also get help by calling one of these **toll free** numbers.

National AIDS hotline: 1-800-342-AIDS

National STD hotline: 1-800-227-8922

STDs are a group of diseases that are spread by having sex. They include diseases like genital warts and chlamydia.

People with sickle cell disease are not able to fight infections as well as other people. So if they have sex with someone who has an STD, they are more likely to get one.

Signs of STDs

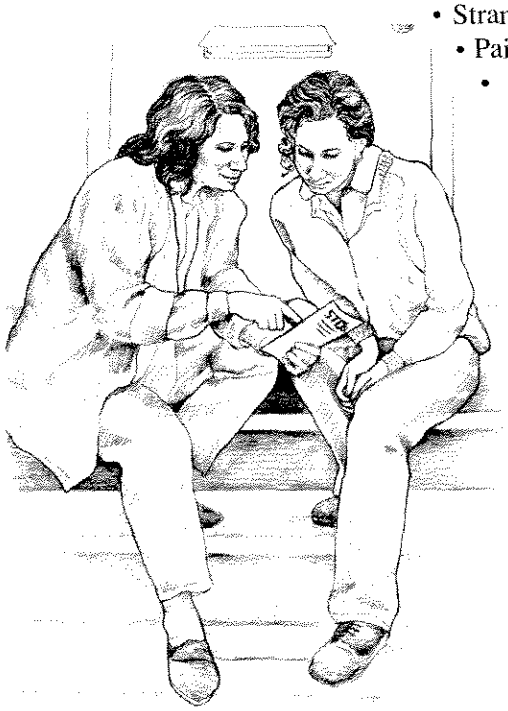
Most STDs can be cured if they are treated early. If your teen is having sex, she should watch for these signs. If she finds any of these, she should call the doctor right away.

In men:
- Drip from the penis
- Pain or burning when passing urine (peeing)
- Sores, rashes or growths on or near the genitals

In women:
- Strange discharge from the vagina
- Pain in the lower abdomen and fever
- Sores, rashes or growths on or near the genitals

Your teen should be checked by the doctor right away if she is worried about any of these signs. STDs can make her very sick or infertile if they are not treated.

Talk to your teen about STDs. Explain what they are and how they are spread. If she doesn't have sex, she won't get an STD. If she chooses to have sex, condoms will help protect her and her partner.



Talk to your teen about STDs.

The AIDS Virus (HIV)

Like other STDs, the AIDS virus can also be spread through sex. Once you have this virus, you can't get rid of it. Over time, it causes people to get sick with AIDS. Some people get sick quickly. Other people stay well for years. As of now, most people who have the AIDS virus end up with AIDS.

AIDS is a deadly disease. There is no cure. There are only treatments to strengthen the body and to deal with some of the problems that can occur.

The best way to deal with AIDS is to protect yourself from infection. The main way that people get the AIDS virus is by having sex with someone who has it. Some people also get AIDS by sharing needles through IV drug use.

Talk to your teen about how she can protect herself from this deadly disease. If she has sex or uses IV drugs, she is at risk of getting infected. Go over these guidelines with her:

- **The only sure way to avoid the virus is not to have sex or use IV drugs.**
- **If you have sex, condoms will help protect you from the virus. Also use foam or cream with Nonoxynol-9 to help protect you from the virus.**

In the past, some people have gotten the virus from blood transfusions. The risk now is very low because all blood is carefully checked.

Pregnancy

See Appendix N for a chart showing the Chances of Having a Baby with Sickle Cell Disease.

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

Getting Pregnant

While a woman with sickle cell disease can have a healthy baby, there are risks. Both she and her baby need to be watched closely. It takes planning and work to have the best chance for a healthy baby.

Women with sickle cell disease may wonder if they can get pregnant and have healthy children. They can. But they need early care to prevent or lessen problems.

Early prenatal care helps the baby. Early care can lower the risk of miscarriage. It can also decrease the risk of having a baby that is too small.

Early prenatal care also helps the mother. The mother needs to be monitored so that any problems can be found and treated early.

Prenatal care should be done by an obstetrician (OB) who is an expert in high-risk pregnancy. It is best if the OB knows a lot about sickle cell disease.

A woman with sickle cell disease needs to plan when she wants to have children. From the start of her pregnancy, she has to be careful about what she does. Alcohol, most medicines and other drugs can all harm her baby. This can be hard for women who rely on certain drugs to help them manage their disease. They may have to live with more pain while they are pregnant or go without treatment for certain problems.

A pregnant woman needs to check with her doctor before taking any medicine. Her doctor will tell her what medicines might cause problems.

It is best to start prenatal care before getting pregnant or right after. The OB and her sickle cell doctor can work together to help keep the mother and her baby healthy.

Partners Should be Tested

Both men and women with sickle cell disease can pass the disease on to their children if their partner has a hemoglobin trait. If a partner has a hemoglobin trait, there is a 50% (1 out of 2) chance that a baby will have the disease. If both partner have sickle cell disease, all their children will also have the disease.

If a partner does **not** have a hemoglobin trait, a couple **can't** have a baby with sickle cell disease. Each of their babies will have a trait.

A partner should have hemoglobin testing **before** a couple gets pregnant. Then the couple will know ahead of time if they might have a baby with a hemoglobin disease. The test can also be done when a woman finds out she is pregnant.

Testing the Baby for Sickle Cell Disease

Many couples want to find out before birth if their baby has sickle cell disease. The unborn baby can be tested in the early months of pregnancy.

If tests show that the baby will have sickle cell disease, the couple can make choices. Some people use the time to prepare for their baby's special needs. Others choose not to continue the pregnancy. It is up to the couple to decide what is right for them. The health care staff should support their decision.

NOTE: Some women with sickle cell disease choose not to have children. Others choose to have children. The choice is up to each woman and her family.

Teen Pregnancy

Teens who get pregnant have more problems than older women who get pregnant. Pregnancy is stressful. Since teens are still growing themselves, pregnancy places an added strain on their bodies.

Teens with sickle cell disease are at risk for even more problems. They have all of the risks that teens have plus the risks that come with sickle cell disease. These risks include the chance of having a child with sickle cell disease.

Some teens with sickle cell disease try to get pregnant to prove that they are normal. Talk to your daughter about the risks before she does something that may not be right for her. Reassure her that she will be able to get pregnant when she plans it. Tell her that she does not have to prove that she is fertile now.

If your teen gets pregnant, she will need your support. If she doesn't want the baby or didn't plan to have children, she may be upset or scared. Talk to her. Listen to her feelings and let her know that you will help her. Share your feelings with her, too, but try to do it in a way that doesn't turn her away. It won't help if you stop talking to each other.

Pregnant teens have some hard choices to make. Some teens choose not to continue the pregnancy. Others choose to have their babies. They may raise them or give them up for adoption. No matter what their choice is, teens with sickle cell disease need to see both an OB and their sickle cell doctor if they get pregnant.

Some teenage boys may want to prove that they can father children. Make sure your teenage son knows that he is fertile. If he has sex with a woman and doesn't want her to get pregnant, they need to use birth control.

Where to get help for pregnant teens

- **Obstetrician (OB)**

It is helpful to find an OB who is an expert in high risk pregnancies.

- **Local County Hospital**

These hospitals have to provide prenatal care. This may be the best place to get care for your teen.

- **Local Health Department**

They may have a public health nurse or social worker who helps teens get the care they need. They may also have prenatal classes for teens.

- **Family Planning Clinics**

These clinics offer low cost pregnancy tests and family planning services. Some of them also provide prenatal care.

- **Community Health Centers**

These clinics often offer low cost pregnancy tests and prenatal care.

- **Medicaid (Medi-Cal)**

This government program pays most or all prenatal care costs for low income women.

- **WIC**

This program provides free food and formula to low income women who are pregnant or mothers of young children.

Your Child's Future: The Adult Years

Work, marriage and family are the main issues of adult life. These are also the main issues for adults with sickle cell disease.

Most adults with sickle cell disease manage to lead full lives. They are able to live on their own and work, take care of a home and raise children. They live with the disease and enjoy their lives.

As a parent, you can help your adult child learn to live on his own. Even adults with serious problems can often live on their own with the help of special programs.

This chapter focuses on two aspects:

- Health Issues
- Help and Support

Health Issues

Most people with sickle cell disease can expect to live into middle or late adulthood. As more is learned about this disease, people are living longer.

Find Problems Early

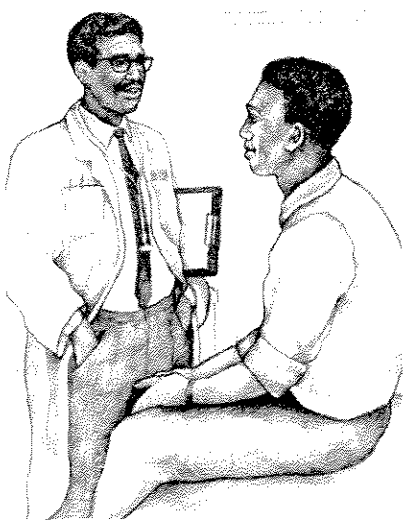
Adults with sickle cell disease still need to see a doctor at least once a year. At these routine visits, their organs are checked for signs of damage. Over time, sickle cells can affect most body organs. The kidneys, lungs, bones, eyes, heart or brain can be damaged by the disease. If signs of damage are found, proper treatment can be started early.

Hospital Care

Most of the health care needs of adults with sickle cell disease can be handled at the doctor's office and at home. Hospital care is sometimes needed.

Major Health Problems

Some adults with sickle cell disease have major health problems. Two of the most common are acute chest disease (lung disease) and kidney failure. Other problems, such as aseptic necrosis, strokes, blindness and congestive heart failure can happen to adults.



Adults with sickle cell disease need to see a doctor once a year.

- **Lungs**

Adults who keep having acute chest syndrome may need frequent blood transfusions. They may also need to have their lung function checked closely.

- **Kidneys**

Some people develop kidney failure and need dialysis or transplants. Transplants can cure kidney disease in many patients.

- **Bones**

The chance of getting aseptic necrosis in the hip increases with age. Hip replacements help many people who have this problem.

- **Brain**

Strokes can occur at any age in sickle cell disease. Transfusion and rehabilitation help most patients regain their abilities.

- **Eyes**

Blindness rarely happens. It can be prevented by yearly check-ups with an ophthalmologist (eye doctor) who knows about sickle cell disease.

- **Heart**

Older adults with sickle cell disease may get congestive heart failure. Medicine and transfusions may help.

NOTE: To help prevent and treat these problems, regular visits to the doctor are a must.

Fears of Dying

People with sickle cell disease often have fears of dying. A long or serious illness can make these fears worse. These fears can come up over and over again because each new set of problems brings new worries.

The same is true for you. Parents are often afraid that their children with sickle cell disease will die.

Talk about your feelings with each other. It will bring you closer together. Talking about these fears helps most people live with them. Honest, open talk seems to work best. You may also feel better if you share your feelings with others who are close to you.

It is important that the fear of death does not become the main focus of your life or your child's life. If either of you can't stop thinking about it, get help. Talk about it with a counselor or clergy.

Help and Support

There are some programs which provide help and support for adults with sickle cell disease. There are also other sources of support. Both kinds of support are important for adults with sickle cell disease.

Friends, family and others who care can often be a major source of help. Their support is a gift of love that tells a person that he matters.

Formal programs provide a different kind of support. They offer money or other types of help that most people can't afford to give. If your adult child needs these programs, he should make sure he gets them. Some of these programs are described here.

Health Care Costs

Many cities and states have programs to help people who have certain illnesses pay for health care. These programs often cover adults with sickle cell disease.

Job Training and Placement

Adults with sickle cell disease sometimes need help finding a job. Many states have programs to help people with chronic illnesses deal with job and career issues. Ask about job training, career counseling, placement and job support programs.

Basic Living Costs

Some people with sickle cell disease can't support themselves through work. They are fully disabled. The government has a program to help disabled people pay for their basic living expenses. It is called SSI (Supplemental Security Income). This program can provide enough money to allow adults to live on their own.

Sickle cell support groups can be a great help.

Legal Help

Your adult child may have problems getting housing or finding work because of his illness. It is against the law for someone to refuse to hire or rent to people because they have a disease. If your adult child thinks he has not been treated fairly, he should get legal help.

There are legal groups which help people when they have been wronged. Look for these groups:

- Legal Aid
- The Urban League
- The ACLU (American Civil Liberties Union)

These groups have funding to cover the cost of legal help.

Find Out About Help

There are a few places you or your child can go to find out about help. Ask the social worker at your nearest sickle cell center about these programs. Your local Department of Social Services may also know about some support programs.

Many cities also have a local branch of the National Association of Sickle Cell Disease. People in these groups often know the most about the programs in their city. If you have a group nearby, join it.

Your adult child may need to stand up for himself to get the help he needs. Support him in speaking up and asking for help. Encourage him to find out about programs and resources and to find ways to get what he needs.

Places to Ask About Help

- Nearest Sickle Cell Center
- Local branch of the National Association of Sickle Cell Disease
- Other sickle cell organizations
- Sickle cell support groups
- Local Department of Social Services
- Local Health Department
- Local churches
- The Urban League
- Local Department of Employment Services
- Community colleges
- Career Planning offices of colleges and universities

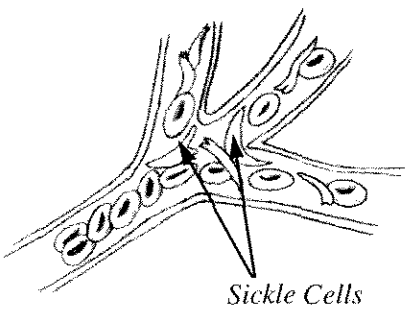
Appendices

What is Sickle Cell Disease?

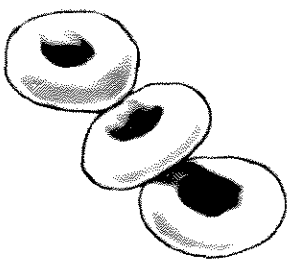
Sickle cell disease is a disease that affects a special protein inside our red blood cells called hemoglobin. Red blood cells have an important job. They pick up oxygen from the lungs and take it to every part of the body. It is the hemoglobin in these cells that carries the oxygen to different parts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

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

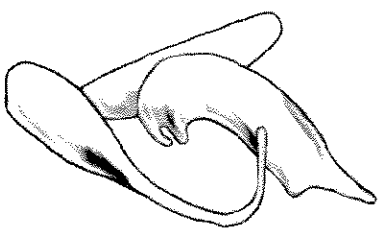
The hard, sticky sickle red blood cells have trouble moving through small blood vessels. Sometimes they clog up these blood vessels so that blood can't bring oxygen to the tissues. This can cause pain or damage to these areas.



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



Normal red blood cells.



Sickle red blood cells.

What Causes Sickle Cell Disease?

Sickle cell disease is inherited

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

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

A baby gets one hemoglobin gene from each parent

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

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

Both parents have a hemoglobin trait

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

A trait is not a disease

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

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

How to find out about your hemoglobin genes

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

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

Comprehensive Sickle Cell Disease Care Plan:

6 Years to Adult

Evaluation	Interval
General Physical Exam	
6 - 8	Every 3-4 months
9 - 18	Every 4-6 months
Over 18	Every 6-12 months
Immunizations	As advised by your doctor
TB Skin Test	Once a year
Comprehensive Social Worker Evaluation	
Interview	Once a year
Home visit	Once a year
School Assessment	Once a year
Genetic Counseling Services	
Family Studies	First visit
Counseling and Education	1-3 times a year
Hematology (red blood cell) Evaluation	Every 4 months to once a year
Liver-Gall bladder Evaluation	Every 2 years or when needed
Renal (Kidney tests)	Once a year or when needed
Cardiac (Heart tests)	Once a year
Ophthalmology (Eye tests)	Once a year
Pulmonary (Lung tests)	Once a year
Dental Evaluation	Once a year
Neurological Evaluation	Once a year
Psychological/Family Therapy Consultation	Once a year
Physical Therapy Assessment	When needed
Developmental Screen	Once a year or when needed
Formal Nutrition Assessment	Every 2 years or as needed
Teen Center Evaluation	Variable

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

Common Lab Tests

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

Hemoglobin electrophoresis

This is the test which is used to find out a person's hemoglobin type. It is this test which tells you what type of sickle cell disease your child has.

This test is also done when it is important to know how much sickle hemoglobin is in your child's blood. Before a blood transfusion, doctors use the test to help decide how much blood should be given. After a transfusion, the test is used to see if enough blood was given to lower the amount of sickle hemoglobin and prevent complications from sickling.

Complete blood count (CBC)

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

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

Reticulocyte (Retic) count

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

Kidney and liver function tests

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

Urine test (urinalysis)

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

Blood Chemistry Tests

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

X-Rays

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

Health Care Providers

Audiologist

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

Cardiologist (heart specialist)

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

Child Life Specialist

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

Dentist

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

Family Guidance Services

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

Family Practice Physician

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

Genetic Counselor

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

Hematologist (blood specialist)

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

Nephrologist (kidney specialist)

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

Neurologist

This is a doctor who is an expert in the nervous system. The nervous system includes the brain. This specialist checks for seizures, strokes or related problems.

Nutritionist

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

Occupational Therapist

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

Ophthalmologist (eye specialist)

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

Orthopedist (bone specialist)

The doctor who treats damaged bones or joints.

Pediatric Nurse Practitioner

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

Pediatrician

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

Physical Therapist

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

Psychiatrist, Psychologist (see Family Guidance Services)

Social Worker

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

Suggested Acetaminophen Dose Chart*

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

	Chewables	Junior Strength	Regular Strength	Extra Strength
Age	80 mg tablets	160 mg caplets	325 mg tablets	500 mg tablets
6-8 years	4 tablets	2 caplets	1 tablet	—
9-11 years	—	2 1/2 - 3 caplets	1 1/2 tablets	1 tablet
12-14 years	—	4 caplets	2 tablets	1 1/2 tablets
15+ & over 100 lbs	—	—	3 tablets	2 tablets

Give your child the right dose 4-5 times a day or as ordered by your doctor.

Don't give your child more than 5 doses in 24 hours.

TRAVEL LETTER

Re: _____
MR# _____ SAMPLE
DOB: _____

To Whom It May Concern:_____

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

_____under the care of_____.

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

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

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

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

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

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

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

Telephone number _____.

Dr._____is available at telephone number_____during regular business hours to provide further information about individual patients, to answer any questions, and to screen calls for appropriate physicians. During other times, or if you need to speak to the on-call hematologist, the_____ hospital switchboard can page one of our physicians 24 hours a day at _____.

When to Call the Doctor or Nurse

Call to have your child seen right away if your child has one of these danger signs:	**FEVER**	101°F or higher
	HEAD/NECK	Severe headache or dizziness Stiff neck
	CHEST	Pain or trouble breathing
	STOMACH	Severe pain or swelling
	COLOR	Loss of normal skin color, very pale or gray
	PENIS	Painful erection
	BEHAVIOR	Seizures Weakness or paralysis (can't move arm or leg) Can't wake up

If you can't reach the doctor, take your child to the Emergency Room.

Call for advice if your child has one of these problems:	**STOMACH**	Vomits more than once Has diarrhea more than once
	FEVER	100°F which lasts more than 24 hours
	COLOR	Jaundiced (eyes or skin look yellow)
	ARMS, LEGS AND BACK	Pain, with no other symptoms
	CHEST	Coughs, without fever or chest pain
	NOSE	Runny or stuffed nose
	BEHAVIOR	Acts strangely Refuses to take medicine Refuses to eat or drink Less active than usual

Temperature Conversion Chart

Celsius (°C) to Farenheit (°F)

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

Letter to Schools About Physical Education

Date:

RE: Patient _____

MR# _____

DOB: _____

To Whom It May Concern:

The above-named patient is a _____-year-old followed at _____
(Clinic or Hospital)
_____ for sickle cell disease. She/he is capable of normal participation in a general physical education program, and we would encourage this as much as possible.

However, because of his or her sickle cell disease, there are times when the patient may experience pains in the limbs, shortness of breath or other symptoms with strenuous physical activity. She/he should be excused from such activity at these times, but alternatives should be sought which will allow as much participation as possible in group activities, without forcing the child beyond these temporary limitations.

If you have any questions, please feel to contact me at _____
(Clinic or Hospital)
_____.

Sincerely,

_____
(Doctor or Nurse)

_____
(Telephone Number)

National Institute of Health

Comprehensive Sickle Cell Centers (1993-1998)

Boston Comprehensive Sickle Cell Center
Boston City Hospital
818 Harrison Avenue, FGH-2
Boston, MA 02118
(617) 424-5727
Lillian E. C. McMahon, MD, Director

Bronx Comprehensive Sickle Cell Center
Albert Einstein College of Medicine
Montefiore Hospital Medical Center
Rosenthal Main
111 East 210th Street
Bronx, NY 10467
(212) 920-6310
Ronald L. Nagel, MD, Director

Columbia University Comprehensive Sickle Cell Center
College of Physicians & Surgeons
630 West 168th Street
New York, NY 10032
(212) 305-5808
Sergio Piomelli, MD, Director

Duke Comprehensive Sickle Cell Center
Duke University Medical Center
Box 3934 Morris Building
Durham, NC 27710
(919) 684-3724
Wendell F. Rosse, MD, Director

Georgia Comprehensive Sickle Cell Center
Emory University School of Medicine
Department of Medicine
69 Butler Street N.E.
Atlanta, GA 30303
(404) 616-3572
J.R.Eckman, MD, Director

Meharry Comprehensive Sickle Cell Center
Meharry Medical College
Department of Pediatrics
1005 D.B. Todd Jr. Blvd.
Nashville, TN 37208
(615) 327-6763
Ernest A. Turner, MD, Director

Northern California Comprehensive Sickle Cell Center
University of California
San Francisco General Hospital
1001 Potrero Avenue, Bldg.100, Room 331
San Francisco, CA 94110
(415) 206-5169
William C. Mentzer, MD, Director

Philadelphia Comprehensive Sickle Cell Center
The Children's Hospital of Philadelphia
34th Street & Civic Center Blvd.
Philadelphia, PA 19104
(215) 596-9500
K. Ohene-Frempong, MD, Director

South Alabama Comprehensive Sickle Cell Center
CSAB 138
University of South Alabama
College of Medicine
Mobile, AL 36688-0002
(205) 460-7334
S. Goodman, PhD, Director

USC Comprehensive Sickle Cell Center
University of Southern California School of Medicine
2025 Zonal Avenue, RMR 304
Los Angeles, CA 90033
(213) 342-1259
Cage Johnson, MD, Director

Sample Pain Management Contract:

Contract between the Teen and the Sickle Cell Care Team

GOALS:

1. The teen will perform self-care activities as specified.

2. The teen will remain as pain-free as possible.

The Teen's Responsibilities	Staff Responsibilities
1. Will discuss how well the pain medication dosage is working with the health team.	1a. Will administer the agreed upon pain medication and discuss the treatment plan with the teen. 1b. Will adjust pain medication dosage as needed within safe guidelines.
2. Will drink the needed amounts of fluids.	2. Will provide fluids of choice and encourage intake when necessary.
3. Will do deep breathing exercises every 4 hours or use incentive spirometer as agreed upon.	3. Will encourage and assist as needed.
4. Will get out of bed for 10 minutes during both day and evening shifts.	4. Will assist with ambulating when necessary and provide appropriate diversional activities.
5. Will do relaxation exercises for 30 minutes on each shift.	5. Will encourage and assist as needed.

Regional Genetic Network Coordinators

Genetics Network of New York, Puerto Rico, and the Virgin Islands, GENES
Karen Greendale, M.A., Co-Coordinator
Genetic Services Program Administrator Wadsworth Center for Labs and Research
New York State Department of Health
Empire State Plaza, Rm E275
P.O. Box 509
Albany, NY 12201-0509
518/473-8036
(New York, Puerto Rico, Virgin Islands)

Great Lakes Regional Genetics Group, GLaRGG
Louise Elbaum, Coordinator
328 Waisman Center
1500 Highland Avenue
Madison, WI 53705-2280
608/265-2907
(Minnesota, Wisconsin, Illinois, Indiana, Michigan, Ohio)

Great Plains Genetics Service Network , GPGSN
Dolores Nesbitt, Ph.D. , Coordinator
Division of Medical Genetics
Department of Pediatrics
University of Iowa
Iowa City, IA 52242
319/356-4860
(N. Dakota, S. Dakota, Iowa, Missouri, Oklahoma, Arkansas, Kansas, Nebraska)

Mid-Atlantic Regional Human Genetics Network, MARHGN
Gail Chiarrello, M.C.P. , Coordinator
Korman Building, Room B-29
5501 Old York Road
Philadelphia, PA 19141-3098
215/985-6760
(New Jersey, Maryland, W. Virginia, Delaware, Pennyslvania, Virginia, D.C.)

Mountain State Regional Genetic Services Network, MSRGSN
Joyce Hooker, Coordinator
Colorado Department of Health
FCHS-MA-A4
4300 Cherry Creek Drive South
Denver, CO 80222-1530
303/692-2423
(Montana, Wyoming, Colorado, Arizona, Utah, New Mexico)

New England Regional Genetics Group, NERGG
Joseph Robinson, M.P.H., Coordinator
New England Regional-Genetics Group
PO Box 670
Mt. Desert, ME 04660
207/288-2704
(Maine, Rhode Island, Massachusetts, Connecticut, New Hampshire, Vermont)

Pacific Northwest Regional Genetics Group, PacNoRGG
Kerry Silvey, MA, Coordinator
CDRC Clinical Services Bldg.
901 East 18th Avenue
Eugene, OR 97403
503/346-2610
(Oregon, Washington, Idaho, Alaska)

Pacific Southwest Regional Genetics Network , PSRGN
Harriet Kuliopulos, MA, Coordinator
California Department of Health Services
2151 Berkeley Way, Annex 4
Berkeley, CA 94704
510/540-2852 (w)
(California, Nevada, Hawaii)

Southeastern Regional Genetics Group, SERGG
Mary Rose Lane, Coordinator
Emory University School of Medicine
Pediatrics/Medical Genetics
2040 Ridgewood Drive
Atlanta, GA 30322
404/727-5844
(Kentucky, Tennessee, N. Carolina, S. Carolina, Louisiana, Mississippi, Alabama, Georgia, Florida)

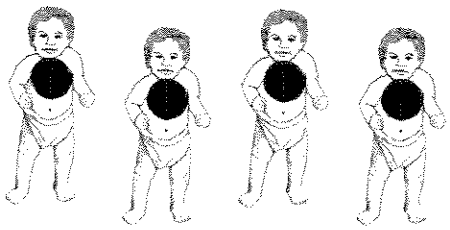
Texas Genetics Network , TEXGENE
William Moore, MHA, Coordinator
Bureau of Maternal and Child Health
Texas Department of Health
1100 West 49th Street
Austin, TX 78756-3199
512/458-7700 (w)
(Texas)

Chances of Having a Baby with Sickle Cell Disease

Both Parents Have Sickle Cell **Disease**

Sickle Cell Disease
(SS)

Sickle Cell Disease
(SS)

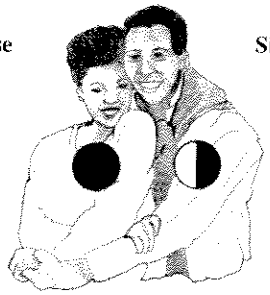


Sickle Cell Disease (SS) | Sickle Cell Disease (SS) | Sickle Cell Disease (SS) | Sickle Cell Disease (SS)

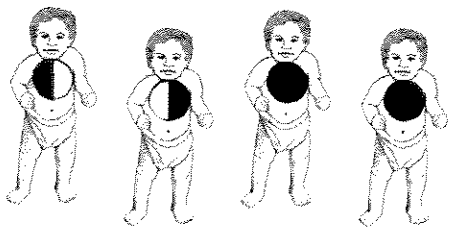
When both parents have sickle cell disease, **all** their children will also have the disease.

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

Sickle Cell Disease
(SS)



Sickle Cell Trait
(AS)



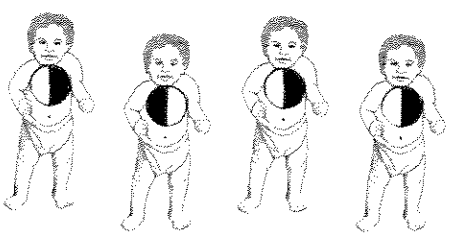
Sickle Cell Trait (AS) | Sickle Cell Trait (AS) | Sickle Cell Disease (SS) | Sickle Cell Disease (SS)

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

One Parent Has Sickle Cell **Disease** and **One** Parent Has **No Hemoglobin Trait**

No Trait
(AA)

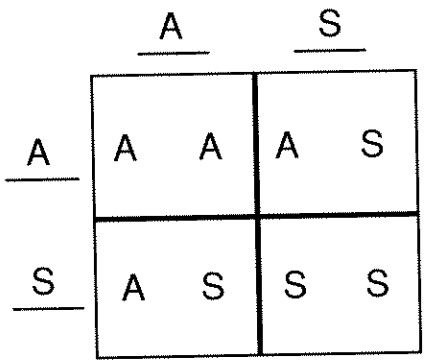
Sickle Cell Disease
(SS)

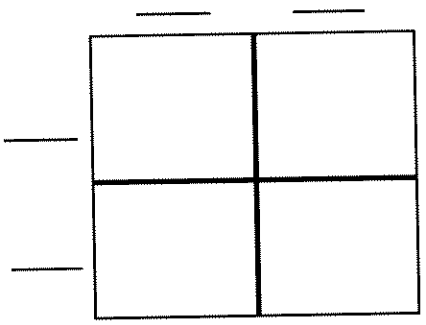


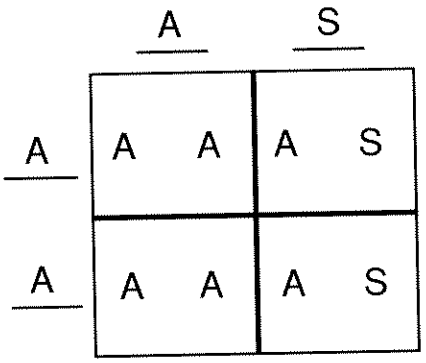
Sickle Cell Trait (AS) | Sickle Cell Trait (AS) | Sickle Cell Trait (AS) | Sickle Cell Trait (AS)

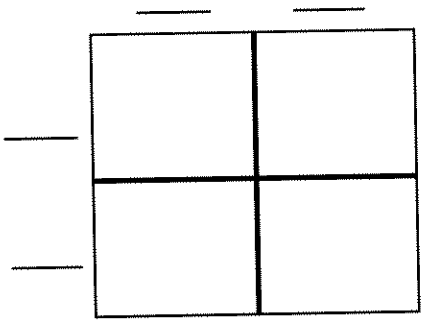
When one parent has sickle cell disease and the other has **no** trait, the couple **cannot** have a baby with sickle cell disease. Each of their babies will have a trait.

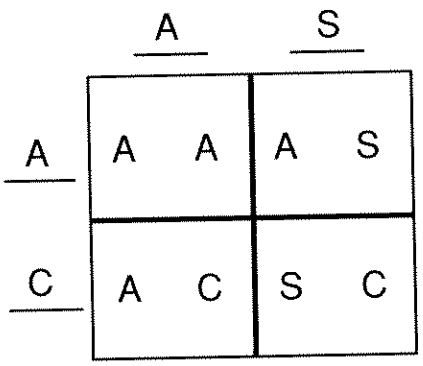
Diagrams of Inheritance *(Punnett Squares)*

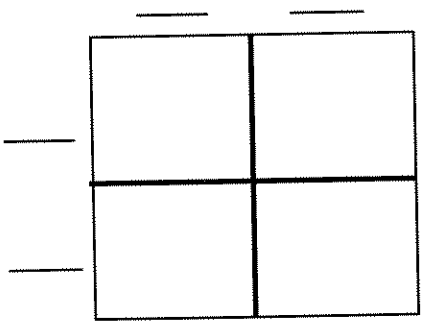












Resources for Parents

Alliance of Genetic Support Groups
35 Wisconsin Circle
Suite 440
Chevy Chase, MD 20815-7015
(800) 336-4363

Genetic Services Branch
Maternal & Child Health Bureau
Parklawn Building, Room 18A-20
5600 Fishers Lane
Rockville, MD 20857
(301) 443-1080

National Center for Education in Maternal and Child Health
2000 N. 15th Street
Suite 701
Arlington, VA 22201
(703) 524-7802

Parents may obtain publications on sickle cell disease and referrals for further information.

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

For more information on parenting:

Dinkmeyer, Don and McKay, Gary, *Raising a Responsible Child,* Simon & Schuster, New York, 1982.

Edelman, Marion Wright, *The Measure of Our Success: A Letter to My Children and Yours*, Beacon Press, Boston, 1989.

Fleming, Don, *How to Stop the Battle with your Teenager: A Practical Guide to Solving Everyday Problems*, Prentice-Hall Press, New York, 1989.

McCoy, Kathleen and Wibbelsman, Charles, MD, *Crisis-Proof Your Teenager*, Bantam Books, New York, 1991.

Poussaint, Alvin S. and Comer, James, *Raising Black Children,* Plume, New York, 1992.

Glossary

Anemia (low blood)

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

Aplastic Episode

An episode when the bone marrow stops making red blood cells. The blood count may fall much lower than usual. If it happens, it is usually with a fever or infection.

Carrier

A person who has one gene for hemoglobin A and one gene for another type of hemoglobin. This person is also referred to as having a hemoglobin trait. A carrier doesn't have the disease, but two carriers can have a baby with sickle cell disease.

Chromosome

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

Complete Blood Count (CBC)

A blood test which measures the size of the red blood cells and the amount of hemoglobin. It tells the number of red blood cells, white blood cells, and platelets.

Dehydration

A condition caused by not having enough water in the body. Dehydration can happen with diarrhea, fever or exercise. It may cause a sickling episode in someone with sickle cell disease.

Electrophoresis

One of the best blood tests to find out a person's hemoglobin type. It shows most hemoglobin traits and can determine different types of sickle cell disease.

Gene

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

Hemoglobin

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

Infarct

A blockage of blood flow that causes tissue to die because it doesn't have enough oxygen.

Inherited

A characteristic passed on from parents to their children. Sickle cell disease is an inherited disease.

Jaundice

Yellowish color of the skin or eyes. It is caused by coloring material from red blood cell breakdown.

Leg Ulcer

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

Priapism

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

Prophylactic Penicillin

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

Sickle Cell Anemia

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

Sickle Cell Disease

A term which refers to all types of sickle hemoglobin disorders, such as SS disease, SC disease and S beta thal disease.

Spleen

An organ on the left side of the body that may be felt below the rib cage. It is a filter to remove bacteria from the blood. This organ does not work well in sickle cell disease. It can trap blood and become enlarged.

Splenic Sequestration

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

Trait (see Carrier)

Transfusion

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

Vaso-Occlusive Episode

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