Sickle Cell Disease Newborn Screening Education Project

Janet Ohene-Frempong, MS

April 21, 2005
Advisory Committee
on Heritable Disorders and Genetic Diseases
in Newborns and Children

SCDAA – NCEC

Background

- Funding
- Purpose
- Mission
- Intended Outcomes

SCDAA – NCEC
Funding and Purpose

- **Funding from HRSA's Genetic Services Branch Maternal and Child Health Bureau (MCHB)**

- **To create and implement a National Coordinating and Evaluation Center (NCEC) to accomplish the goals of the Sickle Cell Disease and Newborn Screening Program**

The SCDNSP supports the comprehensive care of newborns diagnosed with Sickle Cell Disease or as carriers of SCD and their families.

SCDAA – NCEC
Mission

To increase the capacity of the HRSA funded SCD newborn screening community-based programs to provide services to families with babies identified with SCD, or as carriers of SCD or other hemoglobinopathies:

- **model education**
- **counseling**
- **follow-up**

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Intended Outcomes

<u>One of three outcomes</u>:

Within the HRSA funded SCD community-based programs, to increase knowledge about SCD:

- **for families with babies identified as carriers of SCD or other hemoglobinopathies**
- **for families with babies identified with SCD**
- **for their providers who are**

SCDAA – NCEC
Intended Outcomes

<u>One of five program priorities: Materials Development</u>

Create materials and methods of information delivery that will increase health literacy, particularly about sickle cell disease and genetics.

Through information created for families and for providers, the NCEC will establish a foundation to disseminate standardized information about sickle cell disease.

Literacy Levels of Adults in America

Reading Ability	NALS Level	Approx Grade Level	% of Pop.
Very low	1	0-5	21
Low	2	6-8	26
Marginal	3	9-12	32
Skilled	4	College	17
Highly skilled	5	Grad	3

Literacy Levels of Adults in America

Reading Ability	NALS Level	Approx Grade Level	% of Pop.
Very low	1	0-5	21
Low	2	6-8	26
			47

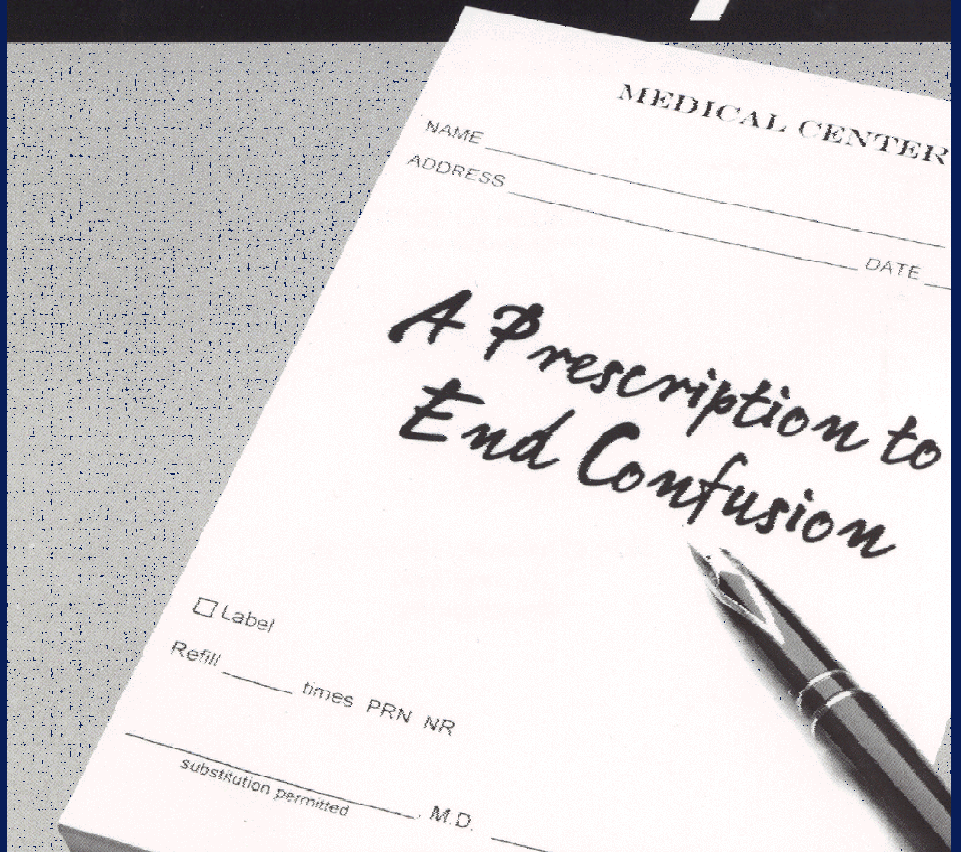
Literacy, Race and Ethnicity

NALS Level	% of Pop.	% of White	% of Hisp-O	% of A m Ind	% of Asian	% of Black	% of PR	% of Cuba	% of Mex	% of C Am
1	21	14	25	25	36	38	47	53	54	56
2	26	25	27	39	25	37	32	24	25	22
Totals	47	39	52	64	61	75	79	77	79	78

INSTITUTE OF MEDICINE
OF THE NATIONAL ACADEMIES

Executive Summary

Health Literacy



IOM Committee on Health Literacy

- Charge
  - Identify obstacles to creating a health literate public
- Recent related studies
  - Confronting Racial and Ethnic Disparities (2003)
  - Assessing Health Communications Strategies of Diverse Populations (2002)
- Report released
  - April 2004

AHRQ Systematic Review

- Charge
  - Perform a systematic review of the relationship between literacy and health and of interventions designed to mitigate literacy-related health disparities
- Support from
  - AHRQ
  - Research Triangle Institute
  - American Medical Association
- Report released
  - Spring 2004

Finally…A Moving Train

SCDAA – NCEC
Plain Language Materials Development

- Technical Assistance and Training Session 2/03
- Evaluation of NBS trait notification letters 3/03-5/03
- Field testing of most commonly used materials for trait education and disease management education 6/04
- Development of tool kit for information providers

SCDAA – NCEC
Plain Language Materials Development

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Words, Space, Pictures and Appeal

How to Evaluate and Develop Easy to Read Materials
Level 1 - The Basics

Janet Ohene-Frempong, MS
J O Frempong & Associates

Layout and Typography

1. Print is easy to see
2. Layout is short and spacious
3. Information is visually well organized
4. Font is plain
5. Avoids all caps – uses unjustified right

Illustrations

1. Are used and serve a purpose.
2. Are clear – cause very little confusion.
3. Are simple – with very few distractions.
4. Are literal - not abstract.
5. Provide context and order.

Text
Prose Literacy

1. Language is simple and friendly
2. Message is clear
3. Information seems manageable
4. Information is engaging
5. Information is repeated

Evaluating and Developing Materials

Text – Document and Quantitative

1. **Few document literacy skills are needed.**
2. **Few quantitative literacy skills are needed.**

Appeal

Intended audience is likely to find it to be:

1. Attractive

2. Easy to use

3. Personally relevant

Highly subjective. Is preferably developed and field tested With intended audience

Facts About Stroke

American Heart Association

Stroke is the third largest cause of death in America, after diseases of the heart and cancer. Although elderly people account for the vast majority of stroke deaths, stroke ranks third as a cause of death among middle-aged people.

Despite these statistics, there's good news. The age-adjusted death rate for stroke has been steadily declining in the U.S., dropping from 88 per 100,000 population in 1950 to 34 in 1984. The rate declined about one percent a year until 1972; after that it started droppingabout five percent a year. Improvements in medical care for stroke patients and the control of high blood pressure have contributed to the decline.

What is stroke?

A stroke is a form of cardiovascular disease. It affects the arteries or veins of the central nervous system and stops the flow of blood bringing oxygen and nutrients to the brain. A stroke occurs when one of these blood vessels either bursts or becomes clogged with a blood clot. Because of this rupture or blockage, part of the brain doesn't receive the flow of blood it needs. As a result, it starts to die.

Types of stroke

One of the most common types of stroke is *cerebral thrombosis*. This occurs when a blood clot forms inside an artery bringing blood to the brain and blocks the blood flow. The clot could also form in a vessel in the brain. Blood clots form most often in arteries damaged by atherosclerosis, a disease condition in which the inner walls of arteries become lined with thick, rough, fatty deposits.

Another type of stroke is a *cerebral embolism*. It occurs when the bloodstream carries

a clot to an artery leading to the brain or in the brain itself. Then the clot becomes stuck. Such clots most commonly come from diseased areas in the heart. Both these types of stroke are caused by blood clots and are called ischemic strokes.

Not all strokes are caused by blood clots, though. Some are caused when a blood vessel on the surface of the brain ruptures and bleeds into the space between the brain and the skull. This is a *subarachnoid hemorrhage*. Another type of stroke is an *intracerebral hemorrhage*. These occur when a defective artery in the brain bursts, flooding the surrounding tissue with blood. Intracerebral hemorrhage is most often associated with high blood pressure, and subarachnoid hemorrhage is usually caused by rupture of a cerebral aneurysm or aterio venous malformation.

Know the warning signals of stroke

The warning signals of stroke are:

- Sudden weakness or numbness of the face, arm and leg on one side of the body.
- Loss of speech, or trouble talking or understanding speech.
- Dimness or loss of vision, particularly in only one eye.
- Unexplained dizziness, unsteadiness or sudden falls.*

About 10 percent of strokes are preceded by "temporary strokes" (transient ischemic attacks or TIAs). These can occur days, weeks or even months before a major stroke. TIAs result when a blood clot temporarily

*If you notice one or more of these signs, talk to your doctor. Your body may be trying to tell you something.

Signs of a Stroke

American Heart Association

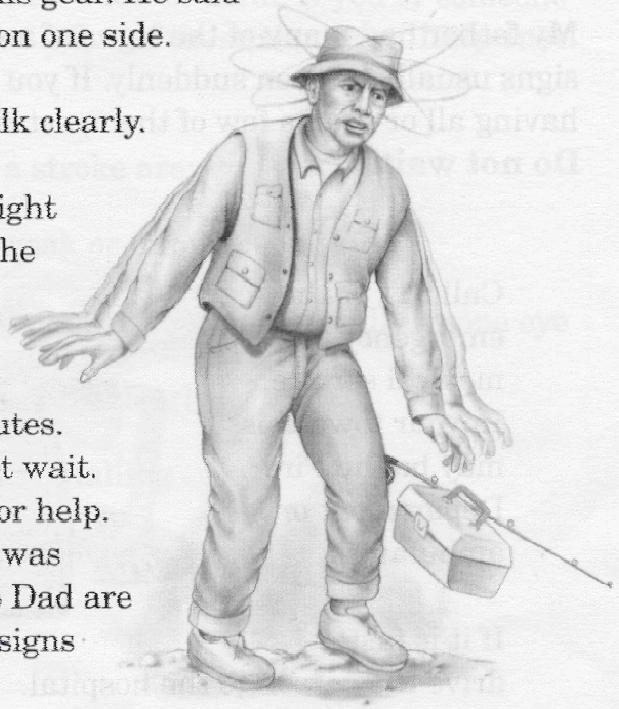
My father is alive today because I know the signs of a stroke. You can save lives, too, if you learn these signs.

Let me tell you what happened. My father has high blood pressure. Last week we went fishing.

He dropped his gear. He said he felt weak on one side.

He did not talk clearly.

He said his sight blurred, and he felt dizzy.



He felt okay in a few minutes. Still, I did not wait. I called 911 for help. I knew what was happening to Dad are the warning signs of a **stroke.**

The Infant and Young Child With Sickle Cell Anemia

Sickle cell anemia is an inherited blood disease that is particularly serious for infants and young children. About one in every 400 black babies is born with sickle cell anemia. A person with this lifelong disease has abnormally formed red blood cells. All complications of disease can be traced to changes in the makeup of the red blood cell causing it to assume a "sickle" shape, making it unusually fragile, and sometimes very rigid. These red blood cells can become trapped within the blood vessels and thus interfere with normal blood flow. This obstruction can lead to sudden pain anywhere in the body, as well as damage to body tissues and organs over time. Supportive treatment is available for sickle cell anemia, but as yet there is no cure except for a bone marrow transplant.

The young child with sickle cell anemia has special needs. If those caring for the child understand these special needs, then complications can be prevented or treated early. These complications are discussed here in detail to help parents and others provide the best possible care to the child with sickle cell anemia.

Many young children with sickle cell anemia are rarely sick, but certain complications can occur which are very serious and sometimes fatal (cause death). It is hard to believe that a healthy looking baby has a life-threatening disease, but this is true when a child has sickle cell anemia. At first parents may not want to know what can happen with this disease. They may want to pretend that nothing is wrong until something happens to indicate otherwise. This is a normal reaction to the unpleasant news that a child has a serious disease. Besides feeling sad, parents may feel angry and/ or guilty. These emotions are very normal and usually temporary. Once the disappointment is accepted, parents are able to see the importance of understanding this disease and recognizing early symptoms of serious complications.

Complications that can be fatal are: (1) septicemia (infection of the blood), and (2) acute splenic sequestration (sudden enlargement of the spleen and rapid drop in the blood count). Both of these medical emergencies can occur suddenly. Treatment is available for both

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Date

Ms. «Mothers_First_Name»«Mothers_Last_Name»
«Address»
«City», «State» «Zip»

Re: Baby «Babys_Last_Name»
DOB: «Date_of_Birth»
Unit #: «Med_Rec_No»

Dear Ms. «Mothers_Last_Name»:

All babies born in State of _____ are tested before leaving the hospital for certain genetic or inherited disorders. Sickle Cell Disease/Trait, and other hemoglobin variations are among them. Your child was found to have a **Sickle Cell Trait or Hemoglobin** "AS". **This is not a disease.** However, it is important for you to be aware of this condition and what it means for your baby.

In order for your baby to have a **Sickle Cell Trait,** it means that baby inherited one normal gene-(Hemoglobin "A"), from one parent and one sickle gene-(Hemoglobin "S") from the other parent. It is important that both you and baby's dad are tested for sickle cell trait, so that you know what your risk is for having a child with Sickle Cell Disease. **Sickle Cell Trait** usually causes no health problems. It is important for you, the parents, to know your hemoglobin status and how it could affect future pregnancies, as well as recognize that when your infant becomes of childbearing age that he/she understands the risk of having children with **Sickle Cell Disease.**

It is vital that you contact our office in the week or two to make an appointment so that we may discuss sickle cell trait and what it means for your baby. **Although rare, Sickle Cell Trait can result in some medical complications.** You will find that this brief session will provide you with the information your need to give your child the appropriate information when the time is right. Please contact us at xxx-xxx-xxxx for an appointment and more information. Inform our receptionist that you have received a letter from us indicating that your baby has **Sickle Cell Trait.** In addition, please inform your child's primary care physician that your child has Sickle Cell Trait.

We look forward to hearing from you.

Sincerely,

Coordinator, Community-Based Sickle Cell Project
Director of Pediatric Hematology/Oncology

Date

Ms. Mothers_First Name Mothers Last Name
Address
City, State Zip

Re: Baby Last Name, First Name
DOB: Date_of_Birth
Unit #: Med_Rec_No

Dear Ms. Mothers Last Name

This letter is to let you know about your baby's blood screening results.

Blood screening results

- All babies born in the state of _____ get screened for sickle cell hemoglobin (pronounced he-mo-glo-bin) right after they are born.
- Your baby's blood screening results shows that your baby has **sickle cell trait.**

Here are 6 things you should do:

1. **Do not be alarmed. Do not worry.**
   - This is NOT a disease.
   - Some people think that sickle cell trait is a disease. But this is not so.
   - A few people have problems with the trait. But this is rare.
2. **Think about getting each parent tested if you may have another baby.**
   - If your child has sickle cell trait, it means that **at least one** parent carries the sickle cell gene.
   - If **both** parents carry the sickle cell gene, it means that you could have a baby with sickle cell disease in the future. This is something to think about.
3. **Get more information, if you need it.**
   - Read the pamphlet that comes with this letter.
   - Call _____ if you have questions.
   - Come to one of our counseling sessions:
     Place:_____ Time: _____
4. **Share this letter with your baby's doctor.**
5. **Keep this letter for your records.**
6. **Keep this information for your child. When he or she is old enough make them aware of their own chance of having children with sickle cell disease.**

We look forward to hearing from you.

Sincerely

Coordinator, Community-Based Sickle Cell Project Director of Pediatric Hematology/Oncology

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NCEC Work Plan

Goal: Priority Area 1: Materials Development Increased
Objective: 1.1: Develop *Consumer* Educational Materials

Activity 1.1:1

Assess and test for readability and user- friendliness:

- **5 most frequently used sickle cell newborn screening materials**
- **5 sickle cell treatment materials most frequently used for children under 4 years of age**

NCEC Work Plan 2004-2005

Goal: Priority Area 1: Materials Development Increased

Objective: 1.1: Develop *Consumer* Educational Materials

Activity 1.1:2

Develop and test draft prototype for :

- **2 newborn screening materials**
- **2 treatment materials for children with SCD under 4 years of age**

NCEC Work Plan 2004-2005

Goal: Priority Area 1: Materials Development Increased
Objective: 1.1: Develop *Consumer* Educational Materials

Activity 1.1:3

Create, disseminate and test a draft tool kit for use by community based-providers and counselor educators in the development of user-friendly materials for families

Focus Group and Discussion Groups With SCT and SCD Parents 2001 and 2004

Sickle Cell Trait and Sickle Cell Disease Information What Parents Want to Know

Janet Ohene-Frempong, MS – Moderator
Health Literacy Consultant for NCEC

and

Christine Corbin, RN – Assistant Moderator
Health Education Coordinator for NCEC

Background

- **In June 2004, the NCEC conducted 3 discussion groups with parents of young children (age 4 and under) with sickle cell trait and with sickle cell disease.**
- **Results of these discussion groups were combined with the results of 2 parent focus groups conducted in November 2001.**

Purpose of the Study

To find out:

1. **What parents in each of the following 3 categories want to know about sickle cell trait and sickle cell disease:**
   - **Parents of newborns diagnosed with sickle cell trait**
   - **Parents of newborns diagnosed with sickle cell disease**
   - **Parents of young children who have sickle cell disease (infants to 4 years of age)**

2. **How they like materials to be designed**

3. **How the current material, most frequently used by the grantees, could be improved to better meet their needs (if at all necessary)**

Methodology

Brookdale University Hospital & Medical Center

Brooklyn, New York

1 group of 19 parents interviewed in 2 consecutive sessions

- **6 were parents of children with sickle cell trait only**
- **13 were parents of children with sickle cell disease**

Brookdale - 2004 Discussion Groups

Part 1 - Trait Notification Discussion Group

Parents were asked to:

- **first, discuss their reactions to the trait diagnosis, then**
- **give their opinions on selected materials designed to give parents information about sickle cell trait.**

Part 2 - Disease Management Discussion Group

- **Parents were asked to give their opinions on selected materials designed to give parents information about the management of sickle cell disease**

Methodology

Children's Hospital of Philadelphia (CHOP) – Philadelphia, Pennsylvania

<u>1 group of 20 parents of children with sickle cell disease</u>

- **some were also parents of children with sickle cell trait**

CHOP - 2004 Discussion Groups

Disease Management and Trait Notification Discussion Group

Parents were asked to:

1. **First, give their opinions on 1 specific piece of material, designed to give a comprehensive overview of how to manage sickle cell disease, identify what they felt may be missing and what they would like to know more about - including psychosocial issues.**

2. **Next, view a 6-minute video on health literacy problems and then let us know what kinds of things they think parents are most likely to find confusing in the management of sickle cell disease.**

3. **Finally, for those who had children with sickle cell trait as well, look at trait notification materials and tell us what they think parents of newborns diagnosed with sickle cell trait should be told and how.**

CHOP - 2001 Focus Groups

Disease Notification Focus Groups

Parents who had children with sickle cell disease were asked to reflect on how they were informed that their newborn 1) MAY have sickle cell disease and then 2) DID have sickle cell disease, and to let us know their opinions on:

- **How a parent may want to receive this information**
- **What a parent may need to know at these two points in time**
- **What a parent may NOT want to know at these tow points in time**

Findings and Recommendations

Selected Issues – Trait Notification

- What the main message should be
- Where to place the emphasis
- How parents want information presented
- Other issues

In their words…

Selected Issues – Trait Notification

“You can make choices

if you know what you’re dealing with…”

Materials Tested – Trait (Carrier)

Materials Tested

The 5 most frequently used by grantees for the management of sickle cell trait

- **Sickle Cell Trait (AS) & Your**
- **Sickle Cell Testing for Newborns**
  What every expecting parent should know
- **About Sickle Cell Disease and Sickle Cell Trait**
- **Trait Notification Letter - Standard Letter Format**
  with Longer Paragraphs
- **Trait Notification Letter - Bulleted List Format**
  with Shorter Paragraphs
- **The Family Connection**

Sickle Cell Trait (AS) & Your Baby

Prepared by
Janet Fithian and Kwaku Obene-Frempong, M.D.
Comprehensive Sickle Cell Center, The Children's Hospital of Philadelphia, for the Sickle Cell Disease Association of America, Inc.®

Reviewed and approved in 1996 by the SCDAA, Inc. Medical and Research Advisory Committee.

Will sickle cell trait make my baby sick?

No. Sickle cell trait is not an illness. Your baby will not have to get special medical care because of sickle cell trait.

You say sickle cell trait is not a problem. Then why was my baby tested?

Babies are tested to see if they have sickle cell disease. Your baby was tested to make sure that *she did not have sickle cell disease.

What is sickle cell disease?

Sickle cell disease is a disease of red blood cells. People with sickle cell disease have red blood cells that become hard and pointed instead of being soft and round. Sickle cells cause anemia, pain and many other problems.

Why is it called sickle cell?

Many red blood cells in people with sickle cell disease look like a sickle, a tool used by farmers. So, a red blood cell that looks like a sickle is called a **sickle cell**.

Normal red cell *A sickle* *Sickle cell*

Can sickle cell trait ever turn into sickle cell disease?

Never. Your baby's hemoglobin types are hers for life. They do not change.

What should I do for my baby?

Your baby does not need special medical care. Give her the best care possible - not because she has sickle cell trait, but because she is your baby. She needs love and good medical care to grow up happy and healthy.

I want to learn more...

Sickle cell trait - what does that really mean?

It means that your baby makes the usual hemoglobin called "A" and some of the sickle cell hemoglobin called "S" in her red blood cells. Put the two together in a red blood cell and you have sickle cell trait: **A and S → AS.**

Lots of African Americans (about two and a half million) have sickle cell trait . It is common in people of African, Mediterranean, Middle Eastern, and Indian origins. It is not only black people who have sickle cell conditions.

How did my baby get sickle cell trait?

She got or inherited sickle cell trait the same way she got the color of her eyes, the shape of her nose and the texture of her hair. She got it through the genes that her mother and father passed on to her. Genes also tell the body what kind of blood to make.

She got an abnormal gene for hemoglobin S from one of her parents and the normal genes for hemoglobin A from the other.

Why is it called trait?

Trait is a common word for a condition where a person gets an abnormal gene from one parent and the normal type of that gene from the other parent. If she gets the abnormal genes from both parents, she is said to have the **disease.**

---

*Sickle cell trait is found equally in both boys and girls. To make this easy to read, we have used **she** and **her** in this pamphlet. In other pamphlets we have used **he, his** and **him.**

What exactly is hemoglobin?

Hemoglobin is inside the red blood cells. It helps them carry oxygen from the air in our lungs to all parts of the body. Hemoglobin also gives blood its deep red color.

What's so special about hemoglobin S?

Hemoglobin S acts differently than hemoglobin A. Normal red blood cells, with mostly hemoglobin A inside, are round and soft and squeeze through small blood vessels very easily.

But red blood cells with mostly hemoglobin S inside can become hard and pointed. They break up easily and cause **anemia,** or low blood count. Sometimes they can't get through tiny blood vessels. This can cause **tissue damage** and lead to **pain** and many other medical problems.

A child with sickle cell trait, **AS,** does not have enough hemoglobin S in her red blood cells for them to become sickle cells easily in the body.

How do babies get sickle cell disease?

Remember, the gene that causes sickle cell disease is called the **S gene.** If a baby gets one **S gene** from one parent and one **S gene** from the other parent, she will have **two S genes,** or **SS.** This means she has the SS type of sickle cell disease.

Or, if she got the **S gene** from one parent and a gene for a different abnormal hemoglobin from the other parent, she would have another form of sickle cell disease.

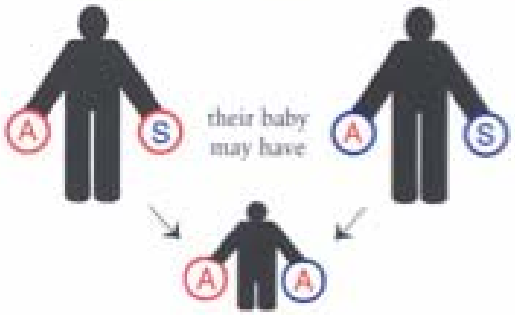
Should we, the parents, take a blood test?

Before you have your next baby, we suggest that you and your partner get a special blood test. We all have two sets of genes for hemoglobin. One set is passed on to the baby from each parent.

(Continued on back panel)

Inheritance Pattern for Sickle Cell Disease-SS

When each parent has sickle cell trait (AS)



AA (normal hemoglobin)

or

AS (sickle cell trait)

or

also, AS (sickle cell trait)

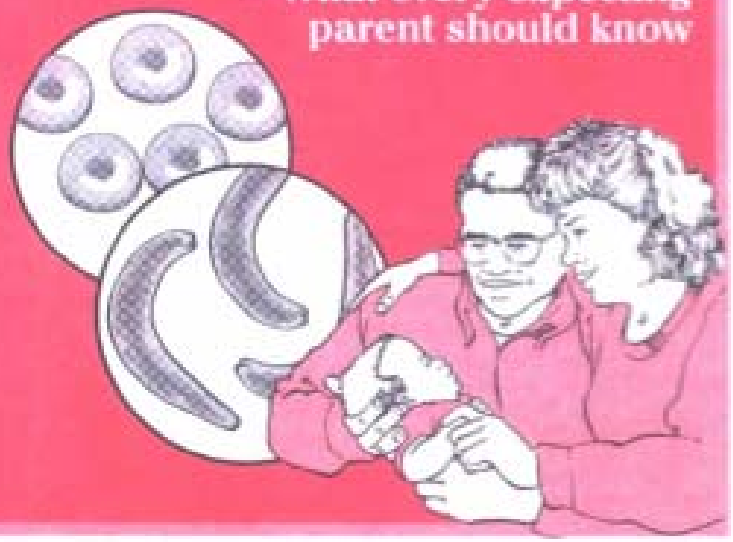
or

SS (the most common type of sickle cell disease)

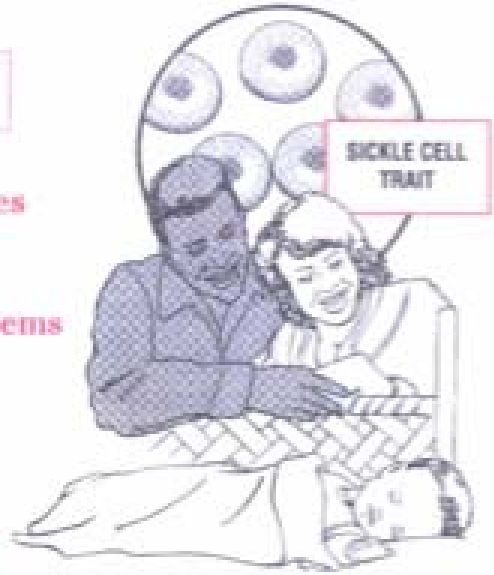
It does not matter what their other babies have, their next one has the same three possibilities: AA, AS, or SS.

Sickle Cell Testing for Newborns

-- What every expecting parent should know



Sickle cell trait:



Also runs in families

Does not usually cause health problems

Sickle cell trait does not change the shape of red blood cells.

Is related to sickle cell disease

People with sickle cell trait will not get sickle cell disease. But their children may be born with the disease.

Sickle cell testing is simple.

- A little blood is taken from your baby's heel or finger.
- Experts study the blood.
- The blood will show if your baby has sickle cell disease or trait.

Ask your doctor if you need to call for the result.

If you think you may want more children:

Ask about sickle cell testing for you and your partner.

It is most important to get tested if your baby has the trait or the disease.

Know the risk of sickle cell disease in your children.

- There is no risk if only one parent has the trait or the disease.
- There is a 1 in 4 chance of disease if both parents have the trait.
- There is a 1 in 2 chance of disease if 1 parent has the trait and the other has the disease.

These chances are for each pregnancy.

Talk to a genetic counselor.

This person can give you more facts. Knowing the facts can help you decide about having more children. Ask your doctor about finding a genetic counselor.

If your baby has sickle cell disease,

he or she will need special treatment right away. That is why the sickle cell test is so important! Your baby will need:

Special medical care

This may include:

- medicines
- immunizations (shots)
- transfusions
- extra doctor visits

Call the doctor right away any time your baby seems sick.

Special care at home

This includes making sure your baby:

- takes penicillin (a medicine) every day
- eats healthy foods
- drinks lots of fluids
- does not get too hot or cold
- takes vitamins (if the doctor says to).

Ask your doctor for details.

If your baby has sickle cell trait, he or she needs the same care as any other baby.

ABOUT SICKLE CELL DISEASE

and sickle cell trait

Sickle Cell Foundation
of Palm Beach County, Inc.
1600 North Australian Avenue, West Palm Beach, Florida 33407-5621
(561) 833-3113 • Fax (561) 659-4505

SICKLE CELL DISEASE CAN CAUSE SEVERAL HEALTH PROBLEMS

For example:

INFECTIONS

Curved, sticky blood cells can clog the spleen and prevent it from doing its job -- helping the body kill germs.

ANEMIA

Sickle cells have a shorter life than regular red blood cells. The body cannot make new red blood cells fast enough to replace old ones. With fewer red blood cells, organs and tissues do not get the oxygen they need.

ORGAN DAMAGE

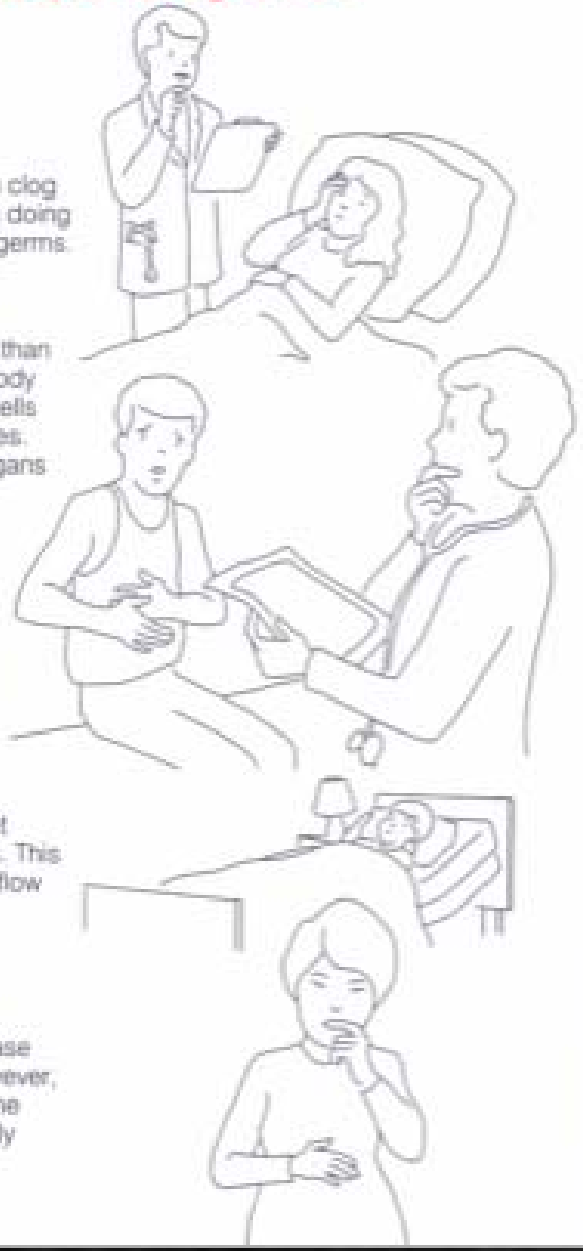
Lack of oxygen over many years can lead to tissue and organ damage in any part of the body.

PAIN

Crescent-shaped cells can get stuck in smaller blood vessels. This may block blood and oxygen flow to tissues, resulting in pain.

COMPLICATIONS DURING PREGNANCY

A woman with sickle cell disease can have a healthy baby. However, risks are involved, and both she and the baby should be closely monitored by a health-care provider. Prenatal care is very important!



Date

Ms. «Mothers_First_Name»«Mothers_Last_Name»
«Address»
«City», «State» «Zip»

Re: Baby «Babys_Last_Name»
DOB: «Date_of_Birth»
Unit #: «Med_Rec_No»

Dear Ms. «Mothers_Last_Name»:

All babies born in State of _____ are tested before leaving the hospital for certain genetic or inherited disorders. Sickle Cell Disease/Trait, and other hemoglobin variations are among them. Your child was found to have a **Sickle Cell Trait or Hemoglobin** "AS". **This is not a disease.** However, it is important for you to be aware of this condition and what it means for your baby.

In order for your baby to have a **Sickle Cell Trait,** it means that baby inherited one normal gene-(Hemoglobin "A"), from one parent and one sickle gene-(Hemoglobin "S") from the other parent. It is important that both you and baby's dad are tested for sickle cell trait, so that you know what your risk is for having a child with Sickle Cell Disease. **Sickle Cell Trait** usually causes no health problems. It is important for you, the parents, to know your hemoglobin status and how it could affect future pregnancies, as well as recognize that when your infant becomes of childbearing age that he/she understands the risk of having children with **Sickle Cell Disease.**

It is vital that you contact our office in the week or two to make an appointment so that we may discuss sickle cell trait and what it means for your baby. **Although rare, Sickle Cell Trait can result in some medical complications.** You will find that this brief session will provide you with the information your need to give your child the appropriate information when the time is right. Please contact us at xxx-xxx-xxxx for an appointment and more information. Inform our receptionist that you have received a letter from us indicating that your baby has **Sickle Cell Trait.** In addition, please inform your child's primary care physician that your child has Sickle Cell Trait.

We look forward to hearing from you.

Sincerely,

Coordinator, Community-Based Sickle Cell Project
Director of Pediatric Hematology/Oncology

Date

Ms. Mothers_First Name Mothers Last Name
Address
City, State Zip

Re: Baby Last Name, First Name
DOB: Date_of_Birth
Unit #: Med_Rec_No

Dear Ms. Mothers Last Name

This letter is to let you know about your baby's blood screening results.

Blood screening results

- All babies born in the state of _____ get screened for sickle cell hemoglobin (pronounced he-mo-glo-bin) right after they are born.
- Your baby's blood screening results shows that your baby has **sickle cell trait.**

Here are 6 things you should do:

1. **Do not be alarmed. Do not worry.**
   - This is NOT a disease.
   - Some people think that sickle cell trait is a disease. But this is not so.
   - A few people have problems with the trait. But this is rare.
2. **Think about getting each parent tested if you may have another baby.**
   - If your child has sickle cell trait, it means that **at least one** parent carries the sickle cell gene.
   - If **both** parents carry the sickle cell gene, it means that you could have a baby with sickle cell disease in the future. This is something to think about.
3. **Get more information, if you need it.**
   - Read the pamphlet that comes with this letter.
   - Call _____ if you have questions.
   - Come to one of our counseling sessions:
     Place:_____ Time: _____
4. **Share this letter with your baby's doctor.**
5. **Keep this letter for your records.**
6. **Keep this information for your child. When he or she is old enough make them aware of their own chance of having children with sickle cell disease.**

We look forward to hearing from you.

Sincerely

Coordinator, Community-Based Sickle Cell Project Director of Pediatric Hematology/Oncology

SCDAA – NCEC
Plain Language Materials Development

- Technical Assistance and Training Session 2/03
- Evaluation of NBS trait notification letters 3/03-5/03
- Field testing of most commonly used materials for trait education and disease management education 6/04
- Development of tool kit for information providers

Development of Trait Information Toolkit

- Guide to Reader-Friendly Materials Development
- Checklist for Evaluating Reader-Friendliness
- Template for a trait notification letter
- Template for a trait notification brochure
- A “What If – Future Babies Card”
- Fact sheet on sickle cell trait
- 5 things parents want to know

Next Steps For Families

Modify or develop, then test and refine materials to provide:

- **Welcome kit for parents of newly diagnosed babies with SCD**
- **Easy-to-read web content on disease management and system navigation skills for parents of infants and children with a longstanding diagnosis of SCD**

Next Steps
Materials for Providers

Modify or develop, then test and refine materials to provide easy-to-access web content for:

- **community based providers, including Primary Care Providers and Emergency Room physicians for**
- **Hemoglobinopathy Counselor/Educators**

Findings and Recommendations

Selected Issues – Disease Notification

When notified that there MAY be a problem

- How they wanted to receive this information
- Things they briefly wanted to be told

When notified that there IS a problem

- What they wanted to know
- What they did NOT want to know
- The one thing thy MOST wanted to hear

Selected Issues – Disease Management

- What parents wanted more information about
- Where to place the emphasis
- What they are most likely to find confusing
- How they wanted information presented

Materials Tested - SCD

Materials Tested

The 5 most frequently used by grantees for the management of sickle cell disease

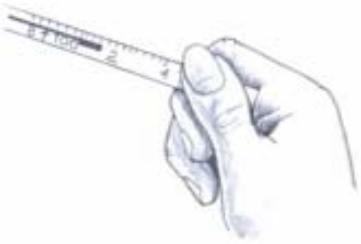
- **A Parents' Handbook for Sickle Cell Disease**
  Part I - Birth to Six Years of Age
- **The Infant and Young Child with Sickle Cell Anemia**
- **You Can Help Your Baby with Sickle Cell Disease Stay Well**
- **Hemoglobin Sickle C Disease**
- **Chest Syndrome**

A PARENTS' HANDBOOK FOR SICKLE CELL DISEASE

PART I

Birth to Six Years of Age

Table of Contents



Time to call the doctor.

Fevers

It is important to know when your child has a fever and what to do about it. When your child has a fever, it is a sign that her body is fighting an infection. Infections can be very serious in children with sickle cell disease. Catching an infection early can let you take actions to prevent it from getting worse.

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

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

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

When to call the doctor

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

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

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

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

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

The Infant and Young Child With Sickle Cell Anemia

A Guide Especially for Parents

The Infant and Young Child With Sickle Cell Anemia

Sickle cell anemia is an inherited blood disease that is particularly serious for infants and young children. About one in every 400 black babies is born with sickle cell anemia. A person with this lifelong disease has abnormally formed red blood cells. All complications of disease can be traced to changes in the makeup of the red blood cell causing it to assume a "sickle" shape, making it unusually fragile, and sometimes very rigid. These red blood cells can become trapped within the blood vessels and thus interfere with normal blood flow. This obstruction can lead to sudden pain anywhere in the body, as well as damage to body tissues and organs over time. Supportive treatment is available for sickle cell anemia, but as yet there is no cure except for a bone marrow transplant.

The young child with sickle cell anemia has special needs. If those caring for the child understand these special needs, then complications can be prevented or treated early. These complications are discussed here in detail to help parents and others provide the best possible care to the child with sickle cell anemia.

Many young children with sickle cell anemia are rarely sick, but certain complications can occur which are very serious and sometimes fatal (cause death). It is hard to believe that a healthy looking baby has a life-threatening disease, but this is true when a child has sickle cell anemia. At first parents may not want to know what can happen with this disease. They may want to pretend that nothing is wrong until something happens to indicate otherwise. This is a normal reaction to the unpleasant news that a child has a serious disease. Besides feeling sad, parents may feel angry and/ or guilty. These emotions are very normal and usually temporary. Once the disappointment is accepted, parents are able to see the importance of understanding this disease and recognizing early symptoms of serious complications.

Complications that can be fatal are: (1) septicemia (infection of the blood), and (2) acute splenic sequestration (sudden enlargement of the spleen and rapid drop in the blood count). Both of these medical emergencies can occur suddenly. Treatment is available for both

The Infant and Young Child With Sickle Cell Anemia

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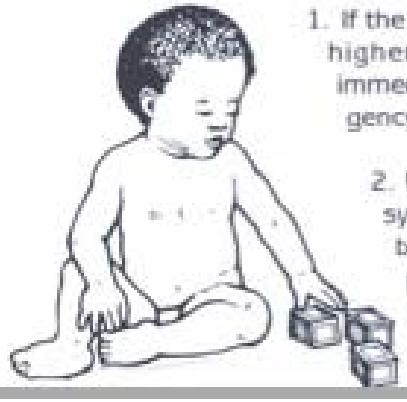
Normal


Sickle Cell Anemia

Fever

Fever is normal response of the body to infection. Fever may be due to a minor illness, but it could be the first sign of a very serious infection. When fever first begins, it is impossible to tell how serious the infection is. The child with sickle cell anemia is more susceptible to serious infections such as septicemia (infection of the blood or blood poisoning). If the child does have septicemia, treatment must be started early to save his or her life. Fever can be the first symptom of septicemia, so it is important for parents to know what to do when their child seems sick.

Whenever a child seems sick the temperature should be checked with a thermometer. Parents should know how to read a thermometer and always have one at home or with them if they are away from home.



1. If the temperature is 101 degrees F. or higher the child should be taken immediately to a facility that gives emergency care.

2. When a child has fever and other symptoms such as pale color, trouble breathing, unusual sleepiness, chest pain, severe cough, abdominal pain, diarrhea, or vomiting, your child should be taken to the doctor immediately.

You Can Help Your Baby With Sickle Cell Disease Stay Well

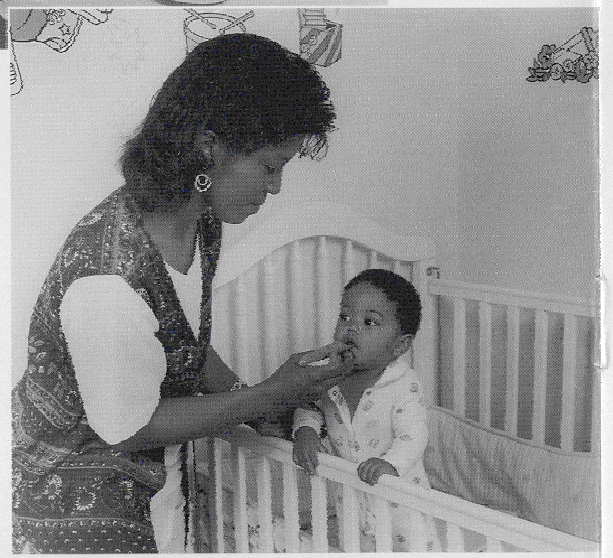
Follow these tips each day:

- ✔ Give your baby penicillin two times each day.
- ✔ Learn to take your baby's temperature.
- ✔ Know when to take your baby to the doctor.

You can help your baby with sickle cell disease stay healthy!

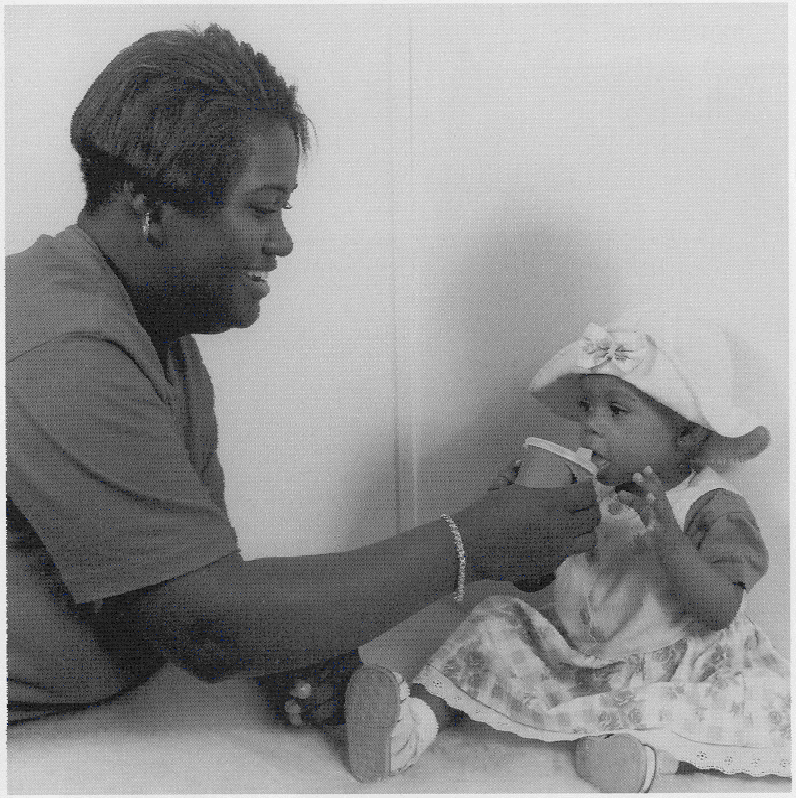
Give your baby penicillin two times each day. Use a special spoon each time!

Give baby a dose before breakfast!



Give baby a dose before bed time!

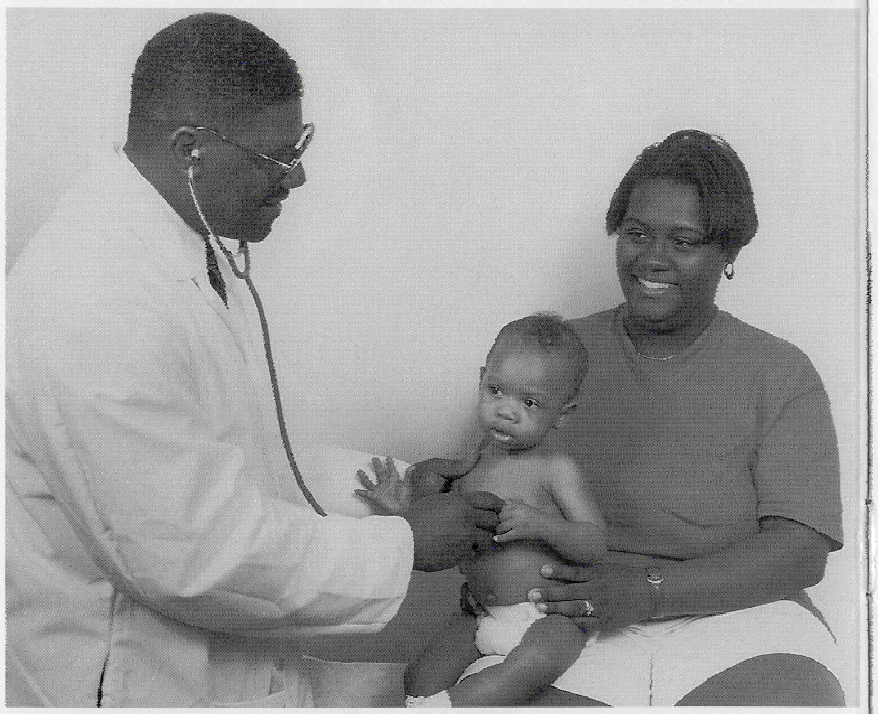
Your baby must have more to drink than other children.



Offer your baby lots of water and juice after she drinks her milk.

Take your baby to the doctor before he gets sick to:

- Get baby shots.
- Get a special shot called pneumovax (new-mo-vax).
- Get penicillin orders.
- See a special doctor, called a pediatric hemotologist, for your child's blood disease. Go by age two months, then at least one time a year.



Have check-ups on time.

Call the doctor right away if:

- Your baby has a fever over 101 degrees.
- Your baby has swollen hands or feet.
- Your baby is crying a lot or is very fussy.
- Your baby has very white or yellow skin.
- The left side of your baby's tummy is swollen.

You can get help if your baby has sickle cell disease

Our sickle cell program can:

- Test your baby for sickle cell disease.
- Tell you about your baby's sickle cell test and about sickle cell disease.
- Teach you and other people you know about the special care your baby needs.
- Help you meet others parents whose babies have sickle cell disease.
- Help you find ways to pay for your baby's special care.

HEMOGLOBIN SICKLE C DISEASE

PROBLEMS SEEN IN CHILDREN WITH SICKLE C DISEASE

Anemia

Your child will always have a slight decrease in his blood count - this is called anemia. Except for occasionally causing tiredness and or weakness in some children, the mild anemia usually does not cause any problems.

Pain

The red blood cells of Sickle C disease are rigid and stiff and may sometimes "clog up" the small blood vessels in the bones and other parts of the body. Because enough oxygen cannot get into the bones, this can cause pain. Painful episodes occur most commonly in the arms, legs, stomach and back. These episodes can last for hours, days, or up to a week. The pain can vary from mild to moderate to severe. The location, length, and degree of pain can vary from episode to episode. How often these crises occur is variable. Some children with SC Disease have no crises at all, but most will have a few each year.

Pneumonia and Other Infections

A child with SC disease has an increased risk of getting certain infections, particularly pneumonia. The abnormal RBC's can "clog up" in the lungs and increase the risk of infection there. This is called chest syndrome. Symptoms to watch for include: fever, fast breathing, trouble breathing, retractions (ribs "suck in" when breathing), very congested cough, and chest pain. If these occur, your child needs to see a doctor immediately.

Spleen

The spleen is normally a small organ located on the upper left side of the abdomen up under the rib cage. It acts as part of the body's defense system that fights infection by removing bacteria (germs) from the blood. Children with SC disease may have an enlarged (big) spleen but this does not happen until they are about age 5 years or older. This big spleen usually does not cause any problems. Occasionally, teen-agers and/or adults may have pain over the spleen and a drop in the blood count - this is called a "spleen crisis."

Eyes

Older children (over age 10 years) and adults with sickle C disease may develop damage to the retina in the back of the eye. This may cause blindness if it is not treated in time. Regular eye check-ups by an ophthalmologist (medical eye doctor) are necessary to diagnose and treat this problem in its early stages.

THE CHILD WITH HEMOGLOBIN SICKLE C DISEASE

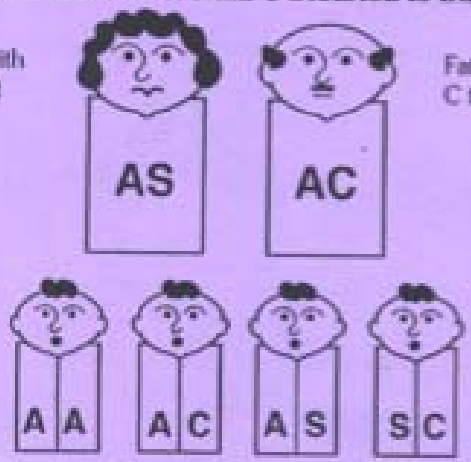
Hemoglobin sickle C disese is a "mild" form of sickle cell anemia. Your child's red blood cells (RBC's) contain two abnormal hemoglobins, called hemoglobin S and hemoglobin C.

Hemoglobin is the substance in the RBC's which carries oxygen to all parts of the body. Instead of appearing round (donut-shaped), your child's RBC's are somewhat misshapen, and some even appear like the sickle-shaped cells found in sickle cell anemia. Others are folded or football-shaped.

RED BLOOD CELLS

Normal RBC's | SC RBC's | SS RBC's (sickle cell anemia)

HOW HEMOGLOBIN SICKLE C DISEASE IS INHERITED



When one parent has sickle trait and the other parent has C trait, each baby has a 1 in 4 chance of inheriting Sickle C disease.

Chest Syndrome

WHAT IS CHEST SYNDROME?

Chest syndrome is a common cause of hospitalization in children with sickle cell anemia. Chest syndrome is a term to describe the chest pain, fever and "pneumonia-like" cough in a person with sickle cell disease. Many times it is mistaken for pneumonia, but your child can have pneumonia and chest syndrome at the same time. In either case, this problem can be fatal in the child with sickle cell disease.

WHAT CAUSES CHEST SYNDROME?

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

WHAT ARE THE SYMPTOMS OF CHEST SYNDROME?

- Sometimes the chest hurts so bad that the pain spreads to the stomach.
- Fever of 102 F (38.8 C) or higher.
- Very congested cough.
- Trouble breathing.
- Fast breathing.
- You may see your child's ribs "suck in" when he/she breathes in.

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

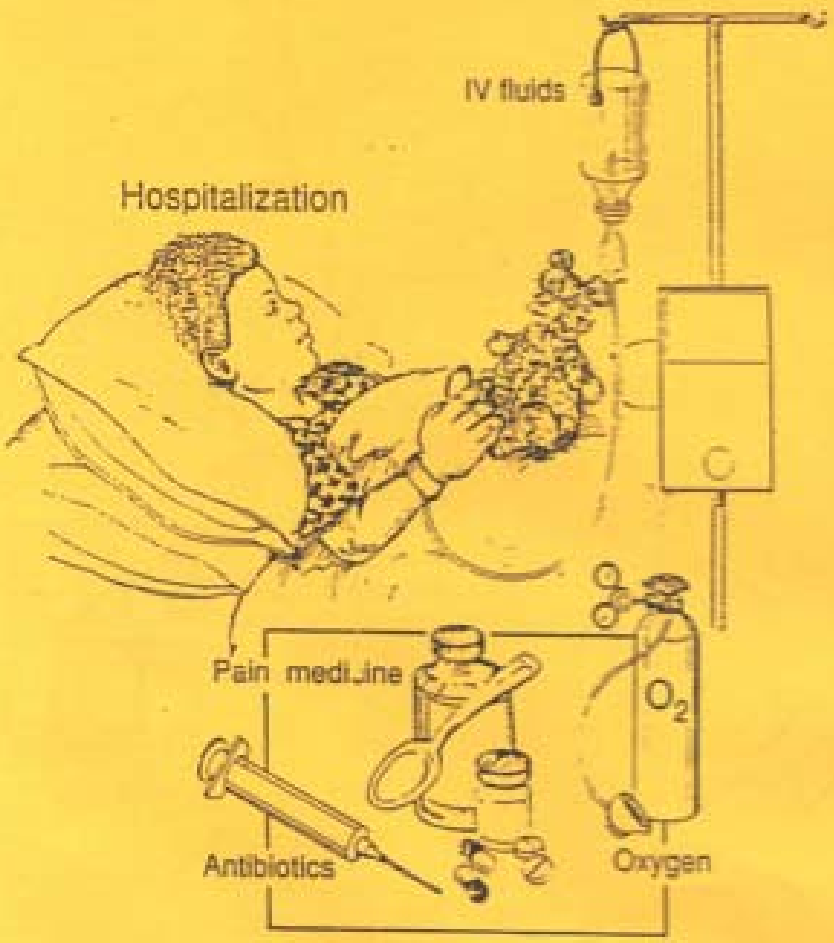
TREATMENT

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

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

OUTCOME

With proper therapy, children with chest syndrome usually do very well. However, some have many episodes. The long-term effects of chest syndrome on lung function during adulthood is unknown. More research is needed.



NCEC Work Plan 2004-2005

Goal: Priority Area 1: Materials Development Increased

Objective: 1.1: Develop *Consumer* Educational Materials

Activity 1.1:2

Develop and test draft prototype for :

- **2 newborn screening materials**
- **2 treatment materials for children with SCD under 4 years of age**

NCEC Work Plan 2004-2005

Goal: Priority Area 1: Materials Development Increased

Objective: 1.1: Develop *Consumer* Educational Materials

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Develop and test draft prototype for :

- **2 newborn screening materials**
- **2 treatment materials for children with SCD under 4 years of age**

NCEC Work Plan 2004-2005

Goal: Priority Area 1: Materials Development Increased

Objective: 1.1: Develop *Consumer* Educational Materials

Activity 1.1:3

Create, disseminate and test a draft tool kit for use by community based-providers and counselor educators in the development of user-friendly materials for families

NCEC Work Plan 2005-2008

Goal: Priority Area 1: Materials Development

Objective: 1.1: Develop *Consumer* Educational Materials

Activity 1.1:1:a

a. SCD *Notification* Materials

Develop prototype materials for a Welcome Kit for parents of newborns newly diagnosed with SCD

NCEC Work Plan 2005-2008

Goal: Priority Area 1: Materials Development
Objective: 1.1: Develop *Consumer* Educational Materials

Activity 1.1:1

b. SCD - *Management* Materials

Develop easy-to-read web content on disease management and system navigation skills for parents of infants and children with a longstanding diagnosis of SCD

NCEC Work Plan 2005-2008

Goal: Priority Area 1: Materials Development

Objective 1.2: Develop *Provider* Materials

Activity 1.2:1 – Materials Review

Engage national provider groups in the review of existing materials (use prototypes developed by the American College of Medical Genetics and the American Pain Society) for ease of use by providers of care.

NCEC Work Plan 2005-2008

Goal: Priority Area 1: Materials Development
Objective 1.2: Develop *Provider* Materials

Activity 1.2:2 - Materials for Physicians

Modify or develop and test and refine easy-to-access materials (i.e. Physician's Guide for community based providers (treatment centers) including Primary Care Providers and Emergency Room physicians for posting on SCDAA website

NCEC Work Plan 2005-2008

Goal: Priority Area 1: Materials Development

Objective 1.2: Develop *Provider* Materials

Activity 1.2:3 - Materials for Hemoglobinopathy Counselors

Modify or develop and test and refine easy-to-access materials for Hemoglobinopathy Counselor/Educators for posting on SCDAA website with SCD