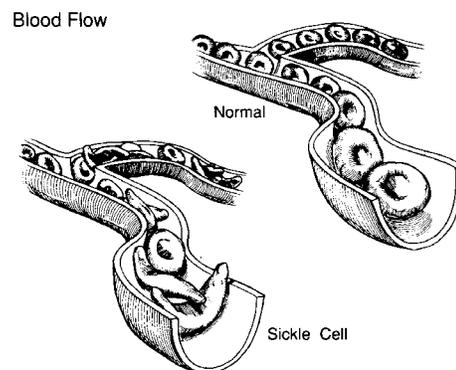
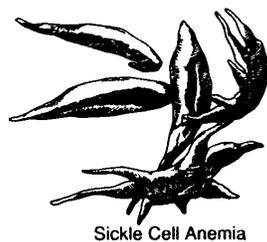
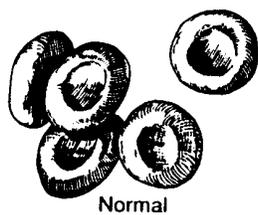


## **Sickle Cell Anemia: A Parent's Guide for the School Aged Child**

### **Important Facts About Sickle Cell Anemia (Hb SS)**

Sickle cell anemia is the most common serious genetic disease in Black Americans. About one in every 400 black infants is born with the condition. A person with this life-long disease has an abnormality of the hemoglobin, which leads to abnormally shaped red blood cells. All complications of this disease can be traced to these abnormal shaped red blood cells. The cell sometimes assumes a "sickle" shape that is very rigid. Rigid red blood cells can become trapped and cause "log jams" within the blood vessels interfering with normal blood flow. The resulting obstruction can lead to sudden pain anywhere in the body, as well as damage the body tissues and organs over time. The altered structure of the red blood cell causes it to break down more rapidly leading to a chronically low blood count, or anemia. The manifestations and severity of the disease are unpredictable. Some children have almost no symptoms while others have multiple serious problems. Most children have occasional mild symptoms. Supportive treatment is available for complications associated with sickle cell anemia. The only known cure is a Bone Marrow Transplant.

Understanding the special needs of the child with sickle cell anemia is essential for the child's health and well being. The intent of this guide is to educate parents and others about how to provide the best care to the child with sickle cell anemia.



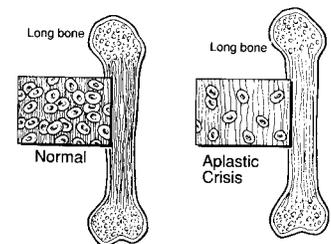
### **DIAGNOSIS OF A SERIOUS DISEASE IN A HEALTHY CHILD**

Children with sickle cell anemia are well most of the time, but certain complications can occur which are very serious and sometimes fatal (cause death). It may be hard to believe that a healthy looking child 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. Eventually parents work through their

feelings and want to learn everything they can about this disease to provide the best care for their child.

## ANEMIA

New red blood cells are produced in the bone marrow every day to replace old blood cells. In a child with sickle cell anemia a red blood cell will last about 14 days. A red blood cell will last about four months in the child with normal hemoglobin. Because the red blood cells do not last long and because the bone marrow cannot make new blood cells fast enough, anemia results. Anemia exists when the number of red blood cells in the circulation is below normal. Sometimes anemia is referred to as a low blood count. Anemia is common in all individuals with sickle cell disease. Even though a child's usual blood count is quite low, this causes few if any real problems. The blood count in a child with sickle cell anemia can drop suddenly. When a child who is already anemic has a drop in the blood count, this is potentially serious. The symptoms of a very low blood count are headache, irritability, unusual sleepiness, lethargy, rapid heartbeat, and pale color. If the lips and fingernails have no pink color even when the hands are warm, the child is pale. A child can have a seriously low blood count without many symptoms. Sometimes the only sign may be that the child is less active or sleeping all the time. If a child has symptoms of a low blood count a doctor should be consulted immediately. In most cases no treatment is necessary and the blood count comes up on its own. Sometimes a blood transfusion is necessary.

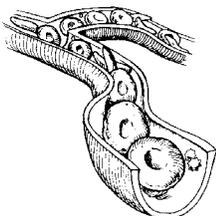


### Aplastic Crisis: Sign and Symptoms

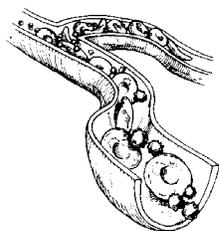
- Paleness
- Lethargy
- "Not feeling good"
- Headache
- Fever
- Low blood count (anemia)
- Recent upper respiratory infection
- Passing out (fainting)

## FEVER

Fever is a normal response of the body to infection. Fever is usually due to a minor illness, but it may 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,



Infection



Septemia (blood poisoning)

treatment must be started immediately to save his life. Fever may be the first symptom of septicemia, so it is important for parents to know what to do when their child seems sick and has a fever.

Whenever a child seems sick, their temperature should be checked with a thermometer. Parents should always have one at home or with them if they are away from home.

1. IF THE TEMPERATURE IS 101.5 DEGREES OR HIGHER take the child immediately to a facility that gives emergency care. Be sure to tell the doctor that the child has sickle cell anemia.

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 for emergency care.

3. SUDDEN WORSENING OF ANY ILLNESS IS ENOUGH REASON TO CALL OR SEE THE DOCTOR SOON.

4. IT IS IMPOSSIBLE TO KNOW HOW HIGH THE FEVER IS BY FEELING THE SKIN. The child's temperature must be taken with a thermometer. Do not give medication for fever before checking the temperature with a thermometer.

Fever medication makes a child feel better and reduces fluid losses from the body, but it does not treat the cause of fever. If a child has a fever of 101.5 degrees or more, you may give the child medication for fever and take the child to a facility that provides emergency care.

There are other things to do when a child has a fever. Dressing the child in light clothing, keeping the room about 70 degrees, and giving a lukewarm bath may help lower the fever. If the child is sleeping, use only light covers. Anything that causes shivering should be avoided because it will make the fever rise. A child with fever needs to drink lots of fluid. If a child has a temperature of 101.5 degrees or higher, it is not safe to wait and see if the fever comes down. The child should be taken to the doctor for immediate treatment.

## INFECTION

The child with sickle cell anemia will get colds, sore throats, and ear infections just like other children. These minor infections are not serious. The more serious infections that are more likely to occur in the child with sickle cell anemia are septicemia (infection of the blood), meningitis (infection around the brain), pneumonia (infection in the lungs), and osteomyelitis (infection of the bone).

A child with septicemia may not seem very sick. A fever of 101.5 degrees or higher may be the only symptom at first. Other symptoms are unusual sleepiness, rapid breathing, pale color, vomiting, stiff neck, and diarrhea.

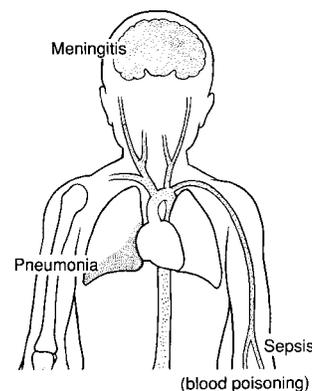
Septicemia is more common in young children, but does occur in older children with sickle cell anemia. Early and aggressive treatment is the best hope for recovery from this some times fatal complication.

Meningitis is very similar to septicemia in its symptoms and can also be fatal. The child with this problem is usually very irritable and may have a stiff neck or seizures.

Pneumonia can be mild with very few symptoms or it can be quite serious. The symptoms are high fever, rapid breathing, and shortness of breath, chest pain, abdominal pain, and cough.

Osteomyelitis causes fever, pain, swelling, and or redness over a bone. A child will often have a fever of 102 degrees or more. If this complication occurs and treatment is delayed, serious and permanent damage to the bone or joint can occur.

All the infections named here are treatable and complete recovery is possible. It is also true that even with treatment, permanent disabilities and sometimes death can result. A doctor should see the child with symptoms of a serious infection as soon as possible. Early recognition and treatment of infection offers the best chance for complete recovery. Pneumococcal vaccines are given to decrease the risks of blood infection (septicemia), meningitis, and pneumonia.



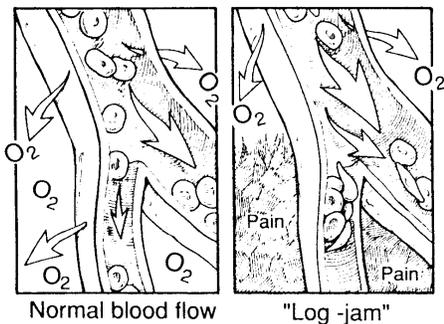
## **PAINFUL EPISODES**

Painful episodes occur in children with sickle cell anemia as a complication of their disease. These episodes are more common in older children, but sometimes happen in babies. Most often the pain seems to be in the bone, but occasionally it occurs in other areas. These episodes usually are not dangerous and may last for several hours to several days, to sometimes a week or ten days.

### **Where Is the Pain?**

Most often painful episodes seem to be in the bone, but occasionally the pain will occur in the chest and abdomen. A complication that may occur in boys with Sickle Cell Anemia is a prolonged, painful erection called Priapism. This may take several hours or days to resolve. If the erection is not better after 2 hours, notify the doctor or take the child to a facility for immediate care. Sometimes admission into the hospital is necessary for pain control.

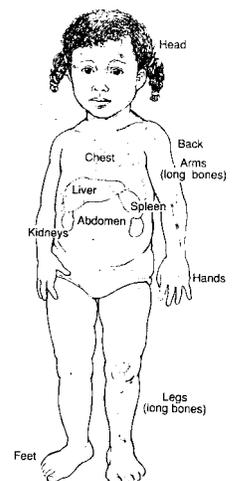
### **WHAT IS THE CAUSE OF THESE PAINFUL EPISODES?**



The exact cause of these episodes of pain is unknown, but it is thought that red blood cells become trapped causing "log jams" inside a blood vessel and interfere with normal blood flow. If blood flow is reduced in even a small area of the body it can cause pain. Sometimes swelling is seen in the area of pain. In babies swelling often occurs in the hands and feet. Older children can have swelling in the arms and legs. Swelling usually does not mean that something is seriously wrong, but in rare cases swelling and pain is caused by infection in the bone. A child with swelling other than hands and feet should be seen by a doctor as soon as it is observed.

### **What Can Be Done to Ease the Pain?**

Taking medication such as Acetaminophen, Children's Advil, or Acetaminophen with codeine at home usually eases painful episodes. Also it is very important that the child drinks lots of fluid to prevent dehydration. A child may refuse to use the part of the body that has the pain. If a child will not stand or walk it is best not to force him. As soon as the pain is better he will be active again. Other measures that may help the pain are rest, and an application of warmth such as a heating pad. If a child cannot be made comfortable at home with medication by mouth, then it may be necessary for the pain to be treated with stronger medication in the hospital.



### **How Long Does the Pain Usually Last?**

Pain usually lasts only a few hours, although it may last several days up to a week or 10 days. If the pain continues for more than four days the parent should call the doctor for possible intravenous therapy.

## **How Do You Know Whether the Pain is From Sickle Cell Anemia or Something Else?**

If the child does not have other symptoms of illness, the pain is probably due to the sickle cell anemia. If the child has fever of 101.5 degrees or above, an infection may be present, and should be seen by a doctor immediately. If the child has chest or abdomen pain with cough, rapid breathing, shortness of breath, pale color, or high fever, you should take the child to a facility that provides an emergency care.

## **Can Too Much Activity Cause a Painful Episode?**

Some parents worry that their child is too active and will cause himself/herself to develop a painful episode. A child that is active is a healthy child. Active behavior should not be discouraged. A child with sickle cell anemia needs to be treated as a normal. Encourage your child to know their limit of how much they are able to tolerate. However, certain conditions may precipitate a painful episode. Studies have shown that extreme cold or hot weather, swimming in an unheated pool, injury, and emotional distress, may cause development of painful episodes as well as unknown factors.

## **Can Anything Prevent Painful Episodes?**

There is not enough known about the cause of the painful episodes to prevent them. Getting plenty of rest, drinking plenty of fluids, and avoiding extremes of cold and heat may minimize the chances of developing pain. There is no known nutritional factor that affects the development of painful episodes. Children with sickle cell anemia who eat a balanced diet usually do not need extra vitamins.

## **HYDROXYUREA**

In the last decade, small doses of a chemotherapy drug called Hydroxyurea has been used to make the clinical course of sickle cell disease milder. This medication is taken by mouth twice every day. Hydroxyurea works by raising the fetal (baby) hemoglobin level in a person with sickle cell disease. The Red blood cells with fetal hemoglobin do not sickle, helping to decrease the pain episode and chest syndrome.



## **ENURESIS & NOCTURIA**

Enuresis (incontinence of urine during the day) and nocturia (bedwetting at night) can occur in the child with sickle cell anemia from about age three years into adulthood. It is more commonly seen in boys than in girls. Enuresis and nocturia can occur daily, intermittently, or occasionally. Current therapies used for bedwetting such as restricting fluids before bedtime does not appear to make a difference in the bedwetting. It is important to remember the child is not wetting the bed intentionally. When a child wets the bed he feels badly. The best approach to bedwetting is a casual one - don't make a big deal out of it, don't spank, punish or yell at your child - this only serves to make him feel ashamed of a behavior which he has no control over.

## **WHAT IS A STROKE?**

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

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

A repeat stroke causes greater brain damage and increases the risk of death. Repeat strokes occur in at least 60% of the children who have already suffered one stroke unless treatment is given.

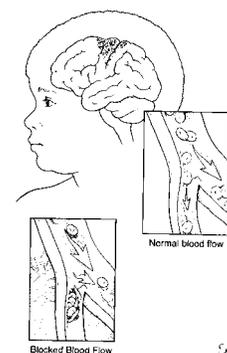
There is a test called Transcranial Doppler Ultrasound (TCD), which can predict strokes in some children with sickle cell.

## **What Causes a Stroke?**

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

## **Symptoms of a Stroke**

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



## **YELLOW EYES**

Occasionally the eyes of a child with sickle cell disease may appear yellow. Usually the yellow eyes result from the build up of by-products from the increased destruction of red blood cells. Sometimes the eyes may appear yellow at the time of a painful episode or other illness; other times there may be no associated problem. Yellow eyes do not usually mean that there is something seriously wrong with your child. They do not need to stay home or be sent home from school.

## **GROWTH AND ADOLESCENCE**

It is important for the parent and child to know that there can be a delay as long as three years in reaching full adult development. The adolescent with sickle cell anemia may still look and feel like a child while his or her friends are developing adult characteristics. Reassurance that they too will grow up and mature will be necessary to relieve anxious feelings.

Girls with sickle cell anemia can and do become pregnant. Serious complications can happen, therefore pregnancy should be planned and under the supervision of a doctor especially

knowledgeable in the management of women with sickle cell anemia. Women with sickle cell anemia can use several different birth control methods successfully.

## SCHOOL

The education of the child with sickle cell anemia is just as important as it is for any child. Sickle cell anemia alone does not affect learning ability. However, excessive absence due to the complication of the disease sometimes affects a child's ability to meet up with class work. Motivating and helping a child to achieve his or her potential in school is important because that child has a future. Extra encouragement and home school when they are unable to be in the classroom would be helpful when illness interferes with school. Keeping up with schoolwork is important. These efforts can make a great difference in educational achievement. The child with aptitude for it should be prepared to attend college in order to qualify for employment that relies upon mental rather than physical skills. The child who does not want to attend college should be encouraged to train for a career that will not require strenuous physical activity.

## LIVING A NORMAL LIFE

Just because a child has a chronic illness does not mean he or she cannot live a normal life. A child with sickle cell anemia has very few limitations. No special diet is needed, just well balanced meals. There is no reason to isolate a child from others to keep him or her from catching colds or other minor infections. Exercise and play should not be discouraged.

Physical endurance may be less because of the anemia, so a child may tire more easily.

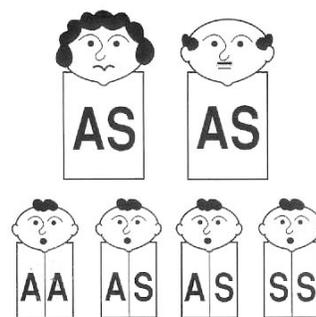
Children will learn their own bounds for physical endurance and will usually stop and rest when they need it. Discipline should be the same for the child with sickle cell anemia as it is for other children in the family. Treating a child with sickle cell anemia in a normal manner is an important part of helping a child to develop a healthy identity as a person.

## THE INHERITANCE OF DISEASE

Sickle cell anemia is only one of many diseases that are inherited. Inherited diseases are passed from parents to children through genes. Genes are what make the physical characteristics of a person such as height, eye color, features of the face, and blood type. Every kind of gene comes in twos with one gene inherited from the mother and one gene inherited from the father. When a person has sickle cell trait there is one gene for normal hemoglobin and one gene for sickle hemoglobin. Because there is one gene for normal hemoglobin the person does not have the disease. The child with sickle cell anemia has two genes for sickle hemoglobin and this causes the disease.

There is a one in four chance, or 25% chance, that a child will be born with sickle cell anemia when both parents have sickle trait. When predicting the chances of having another child with disease, it is important to remember that it is possible to have another child with sickle cell anemia with each pregnancy.

Parents usually want to know if their other children could have sickle cell anemia. This can be learned by blood testing and is a good idea when one child is already known to have disease. Parents need to know that future children can also have disease.



## General Guidelines

- Complications are treatable, the available cure is Bone Marrow Transplant
- Discourage "sick person" identity
- No special diet
- Discipline the same
- Stress importance of school work
- No restrictions
- Regular follow up by medical team
- Encourage normal lifestyle (realistic goals)



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