

Sickle Cell Anemia: A Parent's Guide for the Infant and Young 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 abnormally 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. These "log jams" interfere with normal blood flow. The resulting obstruction can lead to sudden pain anywhere in the body, as well as damage to 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.



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 most common complications of sickle cell anemia are discussed below 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 of Sickle Cell Anemia

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 septicemia and splenic

Family Education Sheet • Sickle Cell Anemia: A Parent's Guide for the Infant and Young Child

sequestration, but it must be started in time. In the sections that follow, more detailed information is given about these and other problems that can occur in the infant and young child with sickle cell anemia.

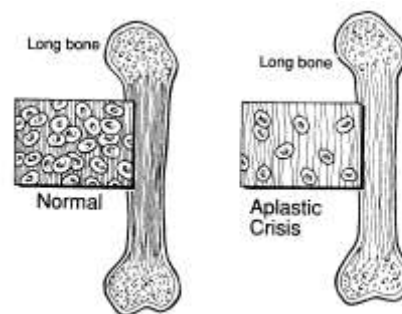
Anemia

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.

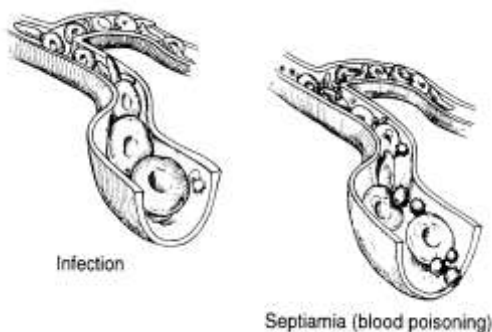
Aplastic Crisis

Aplastic crisis occurs when the bone marrow stops making red blood cells. This may cause significant anemia. The signs and symptoms of aplastic crisis include:

- Paleness
- Lethargy (tiredness)
- "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, treatment must be started immediately to save his/her 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.

Family Education Sheet • Sickle Cell Anemia: A Parent's Guide for the Infant and Young Child

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 A 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 usually serious in the child with sickle cell anemia. The serious infections that are more likely to occur in the child with sickle cell anemia include septicemia (infection of the blood), meningitis (infection around the brain), pneumonia (infection of the lungs), and osteomyelitis (infection of the bone).

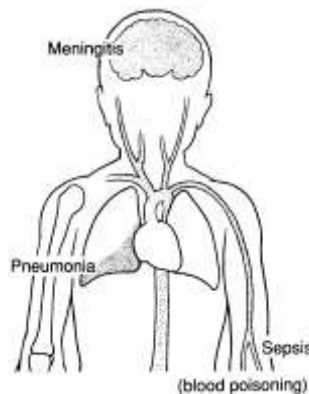
A child with septicemia may not seem very sick initially. A fever of 101.5 degrees or higher may be the only sign at first. Other symptoms are unusual sleepiness, rapid breathing, pale color, stiffness, vomiting, and diarrhea. Septicemia is the major cause of death in young children with sickle cell anemia. Early treatment is the best hope for recovery.

Meningitis is very similar to septicemia in its symptoms. A 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, shortness of breath, chest pain, and cough.

Osteomyelitis causes fever, pain, and swelling over a bone. At first it is quite similar to a painful episode, but a child usually seems sicker with an infection of the bone.

All the infections named here are treatable and complete recovery is possible. It is also true that even with treatment permanent disabilities and even death can result. A doctor should see the child with serious infection symptoms as soon as possible. Early recognition and aggressive



Family Education Sheet • *Sickle Cell Anemia: A Parent's Guide for the Infant and Young Child*

treatment of infection offers the best chance for complete recovery. Pneumococcal vaccines are given to decrease the risks of blood infections (septicemia), meningitis and pneumonias.

Why is Penicillin so Important?

The infant and young child with sickle cell anemia is more susceptible to septicemia (infection of the blood). Scientific studies indicate that penicillin prevents fatal cases of septicemia. When given every 12 hours (two times each day), penicillin can kill bacteria before they grow in the blood and cause life-threatening septicemia.

What if a Child Doesn't Get Penicillin for a Few Days?

A child who is not taking penicillin may be going without important protection. About 12 hours after a dose, the penicillin is almost gone from the body. You should refill the penicillin before it runs out.

Can Septicemia Develop Even if the Penicillin is Given Regularly?

Yes, it is possible since a few bacteria are resistant to penicillin. If a child is taking penicillin and develops a fever of 101.5 degrees or higher the child should see a doctor immediately. (see Fever section).

Will the Penicillin Lose its Effectiveness?

No, penicillin remains very effective over many years. However, some bacteria are resistant to the penicillin. If an infection develops, other antibiotics can be used for bacteria that are resistant to penicillin.

Will the Penicillin Prevent Colds Too?

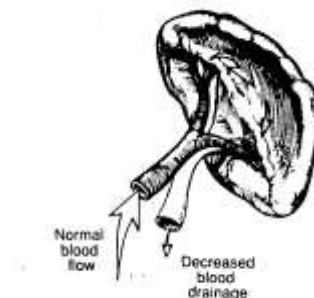
No. Because viruses and not bacteria cause colds they are not affected by penicillin. Children with sickle cell anemia recover from minor illnesses without special problems.

The Spleen

The spleen is a small organ located under the rib cage on the left side of the body. It functions as part of the body's defense against infection by removing bacteria (germs) from the bloodstream. In the child with sickle cell anemia the spleen is not able to remove bacteria, so bacteria can grow in the bloodstream and cause septicemia (infection of the blood).

What Causes the Spleen to become Larger?

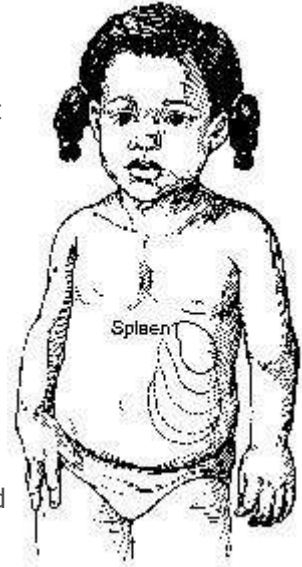
When sickled cells are trapped in blood vessels leading out of the spleen, the normal flow of the blood is blocked. Blood stays inside the spleen instead of flowing through it. This is called sequestration. When this happens, the spleen is very large and it is easy to feel. Sometimes sequestration is painful. Medication can be given to ease the discomfort.



Family Education Sheet • Sickle Cell Anemia: A Parent's Guide for the Infant and Young Child

Is Splenic Sequestration a Serious Problem?

If the spleen enlarges suddenly, the red blood cell count may drop causing severe anemia. This is a serious and potentially life-threatening problem. When the spleen gradually gets larger over several weeks, the blood count does not change as much, so it is not as serious. Any enlargement of the spleen is of concern and must be watched for changes. Parents should check their child's spleen every day, particularly when the child is sick. They should know how their child's spleen usually feels, so that whenever the child seems sick they can check the spleen to see if it is bigger. If the spleen suddenly becomes larger, a doctor should check the child as soon as possible. If the child also has symptoms of a low blood count, then the situation becomes an emergency.



What are the Symptoms of a Low Blood Count?

A child with a low blood count is likely to be irritable, unusually sleepy, and has a rapid heart beat. If the lips and fingernails do not have any pink color even when the fingers are warm, the child is pale. A child can have a seriously low blood count without many symptoms. Sometimes the only symptom is that the child is less active.

At What Age Do Problems with the Spleen Usually Occur?

Babies and young children are at greatest risk of splenic sequestration and septicemia. Complications can develop as young as 2 months of age, but usually occur between the ages of 6 months and 5 years. As children get older the spleen becomes smaller. Eventually it may not enlarge anymore.

Can Splenic Sequestration Happen More than Once?

Yes. A child who has one episode of splenic sequestration is likely to have other episodes.

What is the Treatment for Splenic Sequestration?

If the blood count is dangerously low, then red blood cell transfusion is the treatment. If a child has several episodes, surgery to remove the spleen (splenectomy) may be recommended.

What Can Be Done to Help Children with Sickle Cell Anemia Fight Infection Since the Spleen Doesn't Work?

Prevention and early treatment of infection is the best defense against serious complications. The child who is sick should be carefully watched for symptoms of serious infection. A fever of 101.5 degrees or higher should always be considered a symptom of possible septicemia or bacteria in the blood (see fever section). Penicillin is given twice daily to prevent infection. Pneumococcal vaccines (a shot) are given to boost immunity to harmful infection.

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

Family Education Sheet • Sickle Cell Anemia: A Parent's Guide for the Infant and Young Child

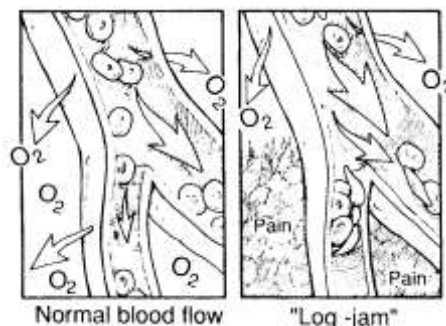
seems to be in the bone, but occasionally it occurs in other areas. These episodes usually are not dangerous. Episodes may last for several hours to several days and sometimes up to a week or more.

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 and cause "log jams" inside a blood vessel. These "log jams" 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 (Tylenol®), 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/her. As soon as the pain is better he/she 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 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 emergency care.

Family Education Sheet • Sickle Cell Anemia: A Parent's Guide for the Infant and Young Child

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 activity 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.

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 once daily. 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 chance of painful episodes and chest syndrome.



Enuresis and 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/she 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/her feel ashamed of a behavior which he/she has no control over.

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.

Family Education Sheet • Sickle Cell Anemia: A Parent's Guide for the Infant and Young Child

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 catch 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.

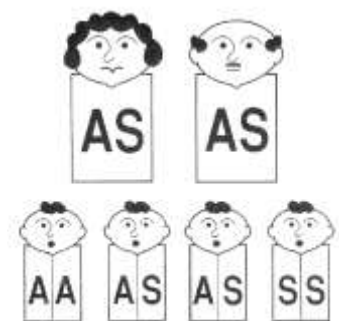
1. With the exception of karate, participation in sports activities is not limited.
2. Physical endurance may be less because of the anemia, so a child may tire more easily.
3. Children will learn their own bounds for physical endurance and will usually stop and rest when they need it. Discipline should 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 the 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 the disease.



Family Education Sheet • Sickle Cell Anemia: A Parent's Guide for the Infant and Young Child

General Guidelines

- Complications are treatable. The only available cure is a 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)

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