TEXAS Health and Human Services | Texas Department of State Health Services

THE INFANT AND YOUNG CHILD WITH SICKLE CELL ANEMIA

A Guide Especially for Parents

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.

Certain complications of sickle cell anemia 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 septicemia and splenic 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.

FEVER

Fever is normal response of the body to infection. Fever is 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.

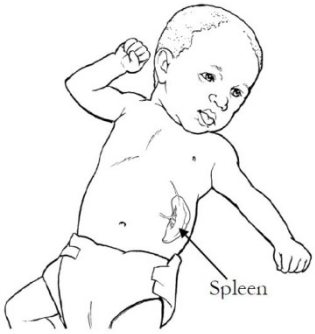
1. If the temperature is 101 degrees F. or higher the child should be taken immediately to a facility that gives emergency care.
2. When a child has fever and other symptoms such as pale color, trouble breathing, unusual sleepiness, chest pain, severe cough, abdominal pain, diarrhea, or vomiting, your child should be taken to the doctor immediately.
3. Sudden worsening of any illness is reason enough to call or see the doctor.
4. It is impossible to know a child has a high fever by feeling the skin. The 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 degrees or more you may give the child fever medication and take the child to a facility for 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 a few covers. Anything that causes shivering should be avoided because it will make the fever rise. A child with fever needs to drink more and should have such things as water, juice, or a soft drink frequently. If a child has a temperature of 101 degrees or higher, it is not safe to wait and see if the fever will come down. The child needs to be taken to the doctor for immediate treatment.

THE SPLEEN

The spleen is normally 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 and your doctor can help treat the pain.

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 heartbeat. 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 DOES NOT WORK?

Prevention and early treatment of infection is the best defense against serious complications. The child who is sick should be carefully watched for symptoms of serious infection. A fever of 101 degrees or higher should always be considered a symptom of possible septicemia or bacteria in the blood (see fever section). Penicillin is given twice daily to prevent infection. Pneumococcal vaccines (a shot) are given to boost immunity to harmful infection.

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 more 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 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 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 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 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 degrees or higher the child should see a doctor immediately (see Fever).

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.

PAINFUL EPISODES

Painful episodes occur in children with sickle cell anemia as a complication of the 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 the abdomen. These episodes are not dangerous and will disappear usually in several hours or sometimes after several days.

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 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.

WHAT CAN BE DONE TO EASE THE PAIN?

Taking medication for pain such as acetaminophen or ibuprofen usually relieves discomfort. Even with relief from most of the pain, 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 plenty of fluids, rest and application of warmth such as a heating pad. If a child is not comfortable at home with medication by mouth, then it may be necessary to treat the pain with stronger medication in the hospital.

HOW DO YOU KNOW IF THE PAIN IS FROM THE SICKLE CELL DISEASE OR FROM SOMETHING ELSE?

If the child does not have other symptoms of illness, the pain is probably due to the sickle cell disease. If the child has fever of 101 degrees or above, an infection could be present. A doctor should see the child who has pain in the chest or abdomen, cough, rapid breathing, shortness of breath, pale color, or high fever in addition to pain in the bones as soon as possible.

CAN TOO MUCH ACTIVITY CAUSE A PAINFUL EPISODE?

Some parents worry that their child is too active and will cause themselves to develop a painful episode. Usually this is not the case. The child who is active is a healthy child. Active behavior should not be discouraged. A child with sickle cell disease needs to be treated as normally as possible, especially when they are acting normally!

CAN ANYTHING PREVENT PAINFUL EPISODES?

Not enough is known about the cause of painful episodes to prevent them. Getting plenty of rest, drinking lots of fluid, and avoiding extremes of heat and cold may minimize the chances of developing pain. No nutritional factor is known to affect the development of painful episodes. Children with sickle cell anemia usually do not need extra vitamins.

HYDROXYUREA

In the last decade, small doses of a chemotherapy medication called Hydroxyurea has been used to make the clinical course of the sickle cell disease milder. This medicine, taken by mouth, raises the fetal (baby) hemoglobin. Red blood cells with fetal hemoglobin do not sickle, helping to decrease pain episodes and chest syndrome. Research is currently active in identifying other treatments for sickle cell disease.

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 the 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 (A) and one gene for sickle hemoglobin (S). Because there is one gene for

normal hemoglobin the person does not have a disease. The child with sickle cell anemia has two genes for sickle hemoglobin and this causes disease.

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. There is 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 four children who all have sickle cell anemia.

When both parents have sickle cell trait, each baby has a one in four chance of having sickle cell disease.

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