SICKLE CELL DISEASE:

A FAMILY GUIDE

(Third Edition)
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Dedication

The members of the Sickle Cell Advisory Committee of New Jersey wish to dedicate this manual to the:

- approximately 80-90 infants born each year in New Jersey with sickle cell disease or other hemoglobinopathies;
- parents and family members who provide these children with the home care, love and encouragement for an active and healthy childhood;
- dedicated staff of the statewide network of treatment centers which specialize in treating children with sickle cell disease;
- hospital emergency room staff who provide prompt emergency treatment to these children;
- pediatricians and family practice physicians who provide routine health care to these children and their families; and
- local community organizations that advocate for individuals with sickle cell disease.
HOW TO USE THIS GUIDE

WHO SHOULD USE IT?

If you, your baby, or a child you care for has one of the sickle cell diseases, this guide is for you. SICKLE CELL DISEASE: A FAMILY GUIDE is mainly for use at home by parents. It also contains important information for other people who care for children with sickle cell disease. Baby sitters, day care and school staff, and other family members may also need to know how to give your child emergency care as well as daily care.

WHAT’S IN IT?

The table of contents will help you quickly find the information you need about day-to-day living with sickle cell disease.

WHERE SHOULD IT BE USED?

Keep this guide at home in a place where it can be easily seen and used. Keep important phone numbers by your telephone.

You can obtain additional copies of this guide by contacting the Comprehensive Sickle Cell Treatment Center where your child receives specialty care. This document is not protected by copyright laws and may be used and reprinted without special permission. The New Jersey Sickle Cell Advisory Committee appreciates citation as to source.
Sickle cell disease is a common inherited disorder of red blood cells. In the United States, it is more common among African Americans. Sickle cell disease is also common in other racial/ethnic groups in the United States and around the world. The map on the next page shows those areas of the world where sickle cell disease is most common.

Most children with sickle cell disease feel and look fine most of the time. When they do have medical problems, they can become sick very quickly and with little warning. For this reason, the State Department of Health and Senior Services now screens all newborns in New Jersey for sickle cell disease. Identifying infants with sickle cell disease in the first weeks of life can help prevent some of the serious medical problems of the disease.

Most of the medical care of children with sickle cell disease involves preventing problems or treating problems as they occur. To make specialty care available to more children, the Department of Health and Senior Services has established sickle cell treatment centers throughout New Jersey. Each center has a team of specialists who work with the child's regular doctor. Together, they help the family manage the child's health care needs.

One of the most important aspects of caring for these children is educating families about the disease. Reading about all of the problems may, however, be scary. Just remember that most of these problems will probably never affect your child! If you are not aware of what might happen, you may not recognize the problems soon enough. **The sooner you notice a problem, the sooner your child can get the treatment he/she needs.**

Over the years, we have found that most families ask the same types of questions about the disease. This section includes some of these commonly asked questions. The answers and entire guide have been written at a level for older children and adults to understand. If you want a more detailed answer or you have other questions, ask the health care team at the treatment center. Information about sickle cell disease is also available on the Internet.
EMERGENCY TELEPHONE NUMBERS

Call 911 or Emergency Ambulance (_____)________________________________________

Dr. ______________________________________ (_____) ____________________________
Sickle Cell Specialist

Dr. ______________________________________ (_____) ____________________________
Child's Regular Doctor

Local Hospital ________________________________________________________________

Address _______________________________________________________________________

Directions _____________________________________________________________________

____________________________________________________________________________

____________________________________________________________________________

Other Numbers: (____)____________________________________________

(____)____________________________________________

SICKLE CELL TREATMENT CENTER TELEPHONE NUMBERS:

From 9 a.m. to 5 p.m. (_____)___________________________________________________

Evenings, Holidays, and Weekends (_____)________________________________________

Your Child's Doctor: __________________________________________________________

Hematologist (Sickle Cell Specialist)

Names of Staff: ________________________________________________________________

____________________________________________________________________________

____________________________________________________________________________
### WARNING SIGNS

Watch for these signs and symptoms of serious problems. Call your child's doctor right away or seek emergency care at a hospital emergency room. Any change from what you feel is normal in your child should be reported to your child's doctor.

<table>
<thead>
<tr>
<th>SIGNS</th>
<th>SYMPTOMS</th>
</tr>
</thead>
<tbody>
<tr>
<td>FEVER</td>
<td>101 degrees or above thermometer reading. * * * Do Not Wait to See if Fever Comes Down.</td>
</tr>
<tr>
<td>IRRITABILITY</td>
<td>crying, fussing, sleepiness, lack of interest in playing.</td>
</tr>
<tr>
<td>PALLOR</td>
<td>looks pale in face, lips, or tongue.</td>
</tr>
<tr>
<td>BREATHING</td>
<td>shortness of breath nostry breathing or grunting sounds.</td>
</tr>
<tr>
<td></td>
<td>fast breathing when resting.</td>
</tr>
<tr>
<td>HEADACHE</td>
<td>sudden or constant</td>
</tr>
<tr>
<td>WEAKNESS</td>
<td>one side of body</td>
</tr>
<tr>
<td>HEART BEAT</td>
<td>pounding or faster than normal.</td>
</tr>
<tr>
<td>PAIN IN THESE AREAS</td>
<td>abdomen   joints   chest   stomach   penis (with prolonged unexpected erection) head</td>
</tr>
<tr>
<td>SWELLING</td>
<td>hands   feet   joints</td>
</tr>
</tbody>
</table>
COMMON QUESTIONS ABOUT SICKLE CELL DISEASE

What is the Difference Between Sickle Cell Disease and Sickle Cell Anemia?

Sickle cell anemia refers to the specific and most common type of sickle cell disease where a child has inherited two genes that produce an abnormal hemoglobin called "S" hemoglobin ("SS" disease). There are several other, less common types of sickle cell disease where one gene produces "S" hemoglobin and the other gene produces "C" hemoglobin (SC disease) or a "beta-thalassemia" type of hemoglobin ("S-beta thalassemia").

Taken all together, all of these conditions are referred to as sickle cell disease. Your child will have inherited only one of these types. It is important for you to know which one. Your doctor can explain this to you.

Is Sickle Cell Disease a Common Disease?

Sickle cell disease is a common, life-threatening disease. Each year in the United States, about one in 400 African-American infants are born with sickle cell disease. Sickle cell disease also occurs in children from other racial and ethnic backgrounds. The most common type of sickle cell disease is sickle cell anemia (SS). Other forms of sickle cell disease include hemoglobin SC disease and sickle beta-thalassemia.

Hemoglobin S-C disease tends to be milder than sickle cell anemia. Sickle beta-thalassemia can be mild or severe.

All of these types of sickle cell disease can be identified by the newborn state-screening test.

The focus of this manual is on sickle cell disease. Your child's doctor can explain more about the particular type that your child has when you visit the hospital.

How Does a Child Get Sickle Cell Disease?

Sickle cell disease is inherited through genes. Genes contain messages that are passed on to the child through the mother's egg and the father's sperm. These messages control such qualities as eye color, blood type, and the kind of hemoglobin a person will have, etc.

Germs do not cause sickle cell disease and you cannot catch it from another person like you catch a cold.

For a child to have any form of sickling disease, each parent will have an abnormal hemoglobin. One possibility is that each parent has sickle cell trait (AS). Another possibility is when one parent has the disease (SS) and the other parent has sickle cell trait (AS).

In hemoglobin SC disease, one parent has sickle cell trait and the other parent has a different trait (hemoglobin C). In sickle beta-thalassemia, one parent has sickle cell trait (or sickle cell anemia) and the other parent carries the trait for beta thalassemia (or has thalassemia major).
Inheritance Pattern When Each Parent Has Sickle Cell Trait

When each parent has sickle cell trait, there is a one in four chance (25% chance) with every pregnancy that the baby could be born with sickle cell disease.
**What Does Sickle Cell Disease Do To Blood?**

It affects the hemoglobin part of red blood cells. Hemoglobin gives blood its red color and carries oxygen from the lungs to other parts of the body.

Normal red blood cells are round, soft, and flexible. Since they can squeeze through small blood vessels, blood flows easily.
In sickle cell disease, red blood cells sometimes change from round to half moon or sickle shaped [C] when the hemoglobin gives off its oxygen.

When red blood cells are shaped like a sickle, they are hard and rigid. These sickled cells can get stuck and plug up small blood vessels. The flow of blood and oxygen can be slowed down or stopped.

Pain, damage to parts of the body, and anemia can result. When the blood flow stops or slows suddenly, the problem is called a sickle cell crisis. The following chart shows how some body organs and tissues may be affected by the plugging of blood vessels.
## Plugging of Blood Vessels

<table>
<thead>
<tr>
<th>Organ/Tissue Involved</th>
<th>Problem Caused</th>
</tr>
</thead>
<tbody>
<tr>
<td>Kidney</td>
<td>bed-wetting, blood in the urine, kidney failure</td>
</tr>
<tr>
<td>Hands &amp; Feet</td>
<td>Swelling and pain</td>
</tr>
<tr>
<td>Spleen</td>
<td>increased risk for serious infections, Splenic sequestration (large amount of body’s blood pooled in the spleen)</td>
</tr>
<tr>
<td>Lungs</td>
<td>Pneumonia, Acute chest syndrome</td>
</tr>
<tr>
<td>Bones</td>
<td>Infection, Bone damage</td>
</tr>
<tr>
<td>Brain</td>
<td>Stroke, Headache</td>
</tr>
<tr>
<td>Skin</td>
<td>Slow - healing sores on legs and ankles</td>
</tr>
<tr>
<td>Penis</td>
<td>Painful unexpected erection</td>
</tr>
<tr>
<td>Eyes</td>
<td>Vision problems</td>
</tr>
<tr>
<td>Liver</td>
<td>Increased size, Gallstones, Gives yellow color to eyes and skin</td>
</tr>
</tbody>
</table>

* Not all of these problems happen to everyone with sickle cell disease. You need to know, however, that they can happen. Tell your child’s doctor right away if you think your child has any of these problems.
Is There A Cure For Sickle Cell Disease?

At this time, there is no cure for most people with sickle cell disease. Medical advances are constantly happening. For up to date information, ask your doctor at the Pediatric Sickle Cell Treatment Center.

Should Other Members Of My Family Be Tested?

Yes. Because the sickle hemoglobin abnormality is inherited, your other children may also carry the sickle hemoglobin gene. If both parents are carriers, there is a 25% chance for a child to be born with sickle cell disease. You should know the sickle cell status of yourself, your spouse, and your children (see Diagram 29). Your health care provider, Sickle Cell Treatment Center or Genetic Testing and Counseling Center (listed on page 42-44) can help to determine who should be tested.

Can Sickle Cell Disease Be Diagnosed Before The Baby Is Born?

Yes. Prenatal testing is available. The testing is quite accurate and can tell whether the child has sickle cell trait, sickle cell disease, or neither. If you are pregnant and would like additional information, you may contact the Genetic Testing and Counseling Center nearest you. Centers are listed on page 44 of this guide.

What Is Sickle Cell Trait?

Like other children, the child born with sickle cell trait has two genes for hemoglobin. One gene is normal while the other gene makes sickle hemoglobin.

The normal gene is often referred to using letter (A). The letter (S) is used to refer to the gene that makes sickle hemoglobin. When combined, the letters (AS) refer to the pair of genes that result in sickle cell trait.

Sickle cell trait is a condition. It is not a disease. If a child has only one gene that makes sickle hemoglobin, the child does not have sickle cell disease (SS). The child will not develop the disease later in life; however, the child should know he or she has trait.

This information will be very important to a person when he or she is planning to start having children. If both partners have sickle cell trait, for each pregnancy there is a 25 percent chance the child could be born with sickle cell disease. (See picture of inheritance pattern on page 8).
Checking for Fever
MEDICAL CARE

FEVER AND INFECTION

Infection is a life-threatening problem of sickle cell disease. Unlike other children, children with sickle cell disease have less resistance to fight infections.

For this reason, children with sickle cell disease may have more infections than other children do. They also tend to easily get more serious infections.

The child with a serious infection may not seem very sick at first. It is important to learn the early warning signs of an infection and know when to take your child right away to the doctor or hospital.

- **Signs and Symptoms**
  - Fever (high temperature of 101 degrees or higher) is the most common symptom of infection.

- **Special Care for Fever**
  - Do not give any medicine such as Tylenol before checking the temperature.
  - Use a thermometer to check for fever.
  - Do not wait for a fever to go down.

**Call the child's doctor right away or take the child to a hospital emergency department.**

Special Care to Help Prevent Infections

- **Penicillin**
  - It is important for the child to take penicillin twice a day. Antibiotics such as penicillin help prevent dangerous infections.
  - It is now recommended that some children above the age of 5 or 6 years stop taking penicillin under certain conditions. Do not stop taking antibiotics unless you have been advised by your doctor. If your child has had his/her spleen removed or if a bacterial infection in the bloodstream has occurred in the past, he/she should not stop taking penicillin. Also, we recommend that your child receive a second vaccination with Pneumovax before stopping penicillin.

- **Pneumovax**
  - Pneumococcal vaccine (Pneumovax) should be given to the child by 2 years of age. We recommend that this vaccination be repeated in 3 to 5 years. It may need to be repeated later as well. This vaccine is given to children with sickle cell disease to help prevent infections caused by the bacteria called pneumococcus.

- **Other Immunizations**
  - Your child must also have the usual childhood immunizations (baby shots).
  - Hepatitis B vaccine is recommended for all children as part of their routine childhood vaccinations whether they are affected by sickle cell disease or not. It helps to prevent a serious viral illness that can be transmitted by blood transfusions.
  - Prevnar is also recommended for all children, but it is especially important for patients with sickle cell disease.
ANEMIA

Normal red blood cells live about four months before they break down. Your body makes new red blood cells all the time to replace the older ones that are slowly breaking down. Sickle red blood cells live less than one month. Even though the body tries, it cannot make new red blood cells fast enough to replace the old ones. This causes the child's red blood cell count to be low (anemia).

When red blood cells break down, the hemoglobin changes into a yellow colored substance called bilirubin. People with sickle cell anemia make a lot of bilirubin. As a result, their skin, eyes and urine may look yellow (jaundice).

Children with sickle cell disease usually have anemia. They also usually adjust and do not have problems from it. There are times, however, when their blood count drops lower than usual. This can happen as a result of an illness or infection.

At certain times, the child cannot make blood cells quickly enough. Their blood count drops suddenly, making them tired, pale and having a poor appetite. The term aplastic crisis is used to describe the problems resulting from this sudden drop in blood count.

- **Signs and Symptoms**
  - Tire easily.
  - Poor appetite.
  - Paleness.
- **Special Care**
  - If you notice any of these signs and symptoms, call the child's doctor. Hospitalization and blood transfusion may be required.

**Splenic Sequestration Crisis**

The spleen is an organ on the left side of the belly under the rib cage. It helps the body fight infection. In children with sickle cell disease, the spleen may get plugged with sickle cells. When the plugging in the spleen makes it suddenly fill with blood and get bigger, the problem is called splenic sequestration crisis. Splenic sequestration crisis can happen with little notice in a few hours and is life-threatening.

- **Signs and Symptoms**
  - sudden weakness.
  - paleness (especially of lips, gums, nails).
  - belly pain.
  - enlargement (swelling) of the spleen.
- **Special Care**
  - Take your child to the emergency room immediately.
  - Immediate hospitalization and blood transfusion may be needed.
  - You can feel the spleen normally in some children with sickle cell disease. It is important for you to know what is normal for your child.
  - Ask your doctor or nurse to teach you where the spleen is and how to feel for changes. See pictures on next page.
Normal spleen and swelling spleen
**LUNG PROBLEMS**

Pneumonia is a common lung infection in children with sickle cell anemia. If a child with sickle cell disease gets pneumonia, his/her lungs do not work as well. Trouble breathing and chest pain can result. The combined symptoms are called acute chest syndrome. Acute chest syndrome can be life-threatening.

- **SIGNS AND SYMPTOMS**
  - chest pain.
  - cough
  - shortness of breath.
  - fast, noisy, or difficult breathing.
  - fever.
- **SPECIAL CARE**
  - It is extremely important to report any of the above signs and symptoms to the child's doctor or nurse. The child with pneumonia/acute chest syndrome needs to be in the hospital. Intravenous (in the vein) antibiotics will be given to fight any infection that may be present. Transfusions may also be necessary in treating this problem.

**KIDNEY PROBLEMS**

All children with sickle cell disease pass urine frequently because their kidneys do not hold water well. Bed-wetting often happens and is not the fault of the child. Children with sickle cell disease also get kidney infections more often than other children.

- **SIGNS AND SYMPTOMS**
  - Pain when urinating.
  - Blood in the urine (hematuria).
  - Bed-wetting (when it has not happened before)
  - More frequent urination than usual.
- **SPECIAL CARE**
  - Make sure your child is getting enough fluid.
  - Bed-wetting may occur until your child realizes that he needs to get up at night to use the bathroom. Try to be supportive and understanding towards your child. It is sometimes worthwhile to wake your child up in the middle of the night to use the bathroom to prevent bed-wetting. **IT IS IMPORTANT THAT YOU DON'T GIVE THE CHILD LESS TO DRINK DURING THE DAY TO TRY TO CORRECT THIS PROBLEM. THE CHILD WITH SICKLE CELL ANEMIA NEEDS EXTRA FLUIDS.**
  - During the day, your child will also need to use the bathroom more often. This may become a problem when the child starts school. Have your doctor or nurse contact the school to give information about Sickle Cell Disease. Also, a letter giving permission for the child to use the bathroom as needed, and drink extra fluids during the school day may be helpful.
  - Infections should be treated with antibiotics.
STROKE

When there is a blockage of blood vessels supplying the brain, a stroke results. Stroke in a sickle cell disease is uncommon but a stroke may be life-threatening.

For the past few years, doctors have been using an ultrasound instrument, called the Transcranial Doppler, to measure blood flow in the vessels that supply the brain. The test is similar to the sonogram used to detect a fetal heartbeat or to look for gallstones.

These doctors have found that children with sickle cell anemia, who have a high rate of blood flow in the brain, are at an increased risk for having a stroke. This is the first time that we have had a way of measuring the risk of a serious problem before it happens. These children have the option of receiving prophylactic blood transfusions.

- **SIGNS AND SYMPTOMS**
  - Problems talking.
  - Weakness of one side of the body.
  - Loss of vision, or blurred vision.
  - Fainting and dizziness.
  - Severe headache.
  - Coma, child won’t wake up.

- **SPECIAL CARE**
  - Immediate hospitalization for observation, tests, and treatment. Treatment includes blood transfusion.
  - Sometimes, the child who has a stroke may return to normal within several days. However, there may be physical weakness or disabilities requiring special therapy.

PAIN CRISIS

Almost all children with sickle cell disease will have a pain crisis at some point in their lives. Some children have them very often. A fever or infection, exposure to extreme heat or cold, physical exhaustion and unusual stress or anxiety may also trigger the pain. However, most often there is no obvious reason why a child gets a painful crisis.

- **SIGNS AND SYMPTOMS**
  - Pain differs from one person to another and varies from mild to very severe. The pain can happen in any part of the body.
  - Infants and young children who can’t say that they hurt may cry a lot and be irritable.
  - Swollen and painful hands and feet (The most common in first type of crisis in an infant or young child).
  - Abdominal (belly) pain
    - Pain, swelling of the belly.
    - Problem breathing.
  - Fever of 101 or higher thermometer reading.

- **SPECIAL CARE**
  - If your child has abdominal pain, call your child’s doctor to make sure the problem is not serious.
  - Speaking to your child’s doctor is especially important if your child has not had a pain crisis before and you are not sure what to do.
  - Check for fever before giving Tylenol. If the child has a fever of 101 degrees or more, call the child’s doctor right away.
  - During a painful crisis, it is important for the child to drink a lot of fluids. Children with sickle cell disease lose a lot of fluids because they have to urinate (pee) a lot. When too much water is lost, the child becomes dehydrated (loss of too much water).
Dehydration can make sickle cell disease worse by causing the red blood cells to sickle faster.

- Encourage the child to drink liquids like juice, fruit drinks and water.
- Discourage the child from drinking fluids that contain caffeine, like soda, tea, and coffee.
- Bed rest.
- If your child does not get better with medication or develops a fever or seems to be getting worse, call your doctor.

**OTHER PROBLEMS**

- **Aseptic Necrosis**
  When the blood vessels supplying bone get blocked by sickle cells, the bone can become damaged. This causes pain which will not tend to go away like a crisis. There is no infection (aseptic), but there is a gradual breakdown or destruction of bone (necrosis). This is called aseptic necrosis. This problem happens most commonly in the hipbone but may affect the knee and shoulder joints as well. Most children with sickle cell disease do not develop aseptic necrosis. There are no ways to prevent it from starting but rest and using crutches may delay the process from worsening over time.

- **Eye Problems**
  Children with sickle cell disease may develop eye problems which can occasionally lead to blindness. The most common eye problems happen very slowly and rarely occur before the age of ten years. Eye problems are more common in children with hemoglobin S-C disease.

  After ten years of age, children should be seen every one or two years by an eye doctor, (an ophthalmologist), who is familiar with sickle cell disease. If your child has sudden problems with vision, call your child’s doctor immediately!

- **Priapism**
  A prolonged unexpected erection of the penis is called priapism. This problem happens when sickle cells block the circulation of the penis. Young boys as well as teenagers can have this problem.

  If priapism lasts more than a couple of hours, notify the child’s doctor. Adolescents may find it embarrassing, so they may not tell you. Talk to them and tell them this problem might happen. Tell them to let you know right away if it does. The child may have to be hospitalized.

- **Skin Problems**
  Some patients develop open skin sores (ulcers), especially around their ankles. If your child has an open cut which is not healing normally, notify your child’s doctor. This problem does not happen often and it usually affects teenagers and young adults with sickle cell disease.
NEW APPROACHES TO THE TREATMENT OF SICKLE CELL ANEMIA

BONE MARROW TRANSPLANT

Bone marrow transplant (BMT) is a complex medical procedure that has been used for many years to treat patients with various kinds of cancers, such as leukemia. The bone marrow is the place where all of the blood cells are produced: red blood cells (that contain hemoglobin), white blood cells (that fight infection) and platelets (that help blood clotting).

In the past few years, this treatment has been used for some children and young adults with sickle cell anemia. Not everyone who has sickle cell anemia can have a transplant nor should everyone with sickle cell anemia have a transplant.

The procedure involves taking a small amount of bone marrow from either a brother or sister (donor sibling) of the patient with sickle cell anemia. First, the donor must be a "match" for the child with sickle cell anemia. This "match" involves some very complex tests that are done on each child's blood. Only about 1 in 4 siblings will be a match for the brother or sister. Also, the donor must NOT have sickle cell anemia. It would not help to transplant bone marrow from someone who makes sickle cells into someone who already has them. Taking bone marrow from the donor involves an operation under general anesthesia, but it is not very dangerous.

Before the child with sickle cell anemia (recipient) can receive the bone marrow transplant, he or she must be given very strong medicines that will destroy the child's own bone marrow cells. Then the donor's marrow is given to the recipient in a way that looks much like a blood transfusion only it is the donor's bone marrow cells that are going into the patient's body. During this time, and for some months afterwards, there are very serious risks of infection for the child.

After a few weeks, if all goes well, the donor's bone marrow cells will begin to make normal red blood cells (and white cells and platelets) in the patient. If that happens, the patient will no longer have sickle cells in their blood and should not have any symptoms of the disease.

Part of the problem is trying to decide who should have a bone marrow transplant. Not every child with sickle cell anemia is seriously ill. Most children have not had a major complication from the disease such as stroke. And not every child will have a "matched" sibling so that even if someone needed a transplant, there might not be a suitable donor for them. There are risks to having a bone marrow transplant. The child who receives the transplant can become very ill and possibly die from the procedure. It is a major medical decision to undergo such a procedure.

Doctors are working on ways to try to identify patients who are likely to have the more serious complications of sickle cell anemia, i.e. stroke, before these events happen. Perhaps in the future, we will be able to recommend bone marrow transplant to those children who are at increased risk for serious complications from sickle cell disease before any major life-threatening event happens. Ask your doctor for more information about bone marrow transplant.
**HYDROXYUREA**

Hydroxyurea is actually a chemotherapy medicine that has the side effect of increasing the amount of fetal hemoglobin in the blood. Fetal hemoglobin is produced before the baby is born and protects the red cells from taking on the sickle shape.

In a large study of adult patients with sickle cell anemia who were given Hydroxyurea, patients had fewer hospitalizations and fewer problems with acute chest syndrome. The medicine did not increase fetal hemoglobin levels in all patients. To be effective, this medication must be taken every day.

Some doctors have started to use Hydroxyurea in children who have had serious problems with the disease. It is too early to know if there is any long-term benefit to taking this medication.

There are a variety of side effects associated with Hydroxyurea. It can cause skin and nail changes, anemia, sores in the mouth, lowering of the white blood cell counts and platelets. There may also be some long-term effects on a child’s growth and fertility. Hydroxyurea may also affect a developing fetus and it should not be taken by someone who is planning a pregnancy.

**BLOOD WORK**

Your child will need to have periodic blood tests. One test, called a complete blood count or CBC, is done to determine the amount of white blood cells, hemoglobin and platelets in the child’s blood (see below). The child with sickle cell disease usually has a low red blood count (low hemoglobin or anemia).

It is important for you to know the hemoglobin level and retic count (see below) that is normal for your child.

The most common blood work drawn on children with sickle cell disease will be discussed in this section. There will be times that other blood may be drawn from your child. Do not ever hesitate to find out what tests are being done and why.

- **CBC (COMPLETE BLOOD COUNT)**
  
  When blood is drawn for this test your doctor is looking at three different values.
  o **WBC (white blood cells)** – These cells help the body to fight infection.
  o **Hemoglobin** – hemoglobin is inside the red blood cells and gives the blood its red color and carries oxygen to the muscles, organs, and body tissues.
  o **Platelets** – The part of the blood that helps make blood clots.

- **RETICULOCYTE OR RETIC COUNT**
  
  Reticulocytes are young red blood cells. Children with sickle cell disease make more young red because their blood cells don’t live very long. The retic count will then be high.

- **TYPE AND CROSS MATCH**
  
  This test is done before giving blood transfusions to make sure that the blood you child will be given matches your child’s blood type. It is done before every transfusion is given.

- **HEMOGLOBIN ELECTROPHORESIS**
  
  This test will show the type of hemoglobin you and your child have. Normal hemoglobin is shown by “AA”, sickle cell trait hemoglobin by “AS”, and sickle cell hemoglobin by “SS”.
BLOOD TRANSFUSIONS

Blood transfusions can be very important in treating some of the problems of sickle cell disease. Your sickle cell doctor will not give a transfusion unless it is absolutely necessary. Many parents are very anxious about transfusions. These concerns are real. If you have religious beliefs or concerns about receiving blood transfusions, please discuss them with your child's doctor or nurse.

Since people with sickle cell disease cannot be transfused with blood that has the sickle trait, family members often cannot donate blood for the child's transfusions. People are always concerned about the risk of getting infections like AIDS and hepatitis from a transfusion. All blood donors are routinely screened for hepatitis and AIDS. We will only transfuse your child if we feel that the risk of the transfusion is less than the risk of the sickle cell problem we are treating.

ANESTHESIA

Before your child receives anesthesia for any reason, let your child's sickle cell doctor know. It is important for your child's sickle cell specialist to know even if general anesthesia is planned for dental work. Special precautions are required for a child with sickle cell disease before he/she receives any type of anesthesia. This must be discussed with your doctor before any procedure is done.

Always let all your child's doctors, including dentists, know that your child has sickle cell disease.

SURGERY

Before your child has minor or major surgery, contact your child's sickle cell doctor and the child's regular doctor. These doctors can help the surgeon and hospital staff prepare the child for surgery and help manage his care afterward.
ROUTINE MEDICAL CARE

- Give your child the medicines that doctors prescribe. Penicillin must be taken twice every day. **IMPORTANT.** The penicillin must be given each morning and each night. If you forget a dose, don't double or triple the dose. Restart it as soon as possible. If your child goes away on a trip, be sure to take enough penicillin with you. Carry your child's Medication and Treatment Record with you to regular doctor's appointments, visits to the sickle cell treatment center, and emergency hospital visits. **DON'T LEAVE HOME WITHOUT IT.**
- An emergency card can also be given to baby sitters, parents of friends, school personnel, and family members.
- Take your child to your regular doctor (pediatrician or family doctor) for childhood shots and regular check-ups.
- Carry an emergency card with you and your child to let health care workers and others know that your child has sickle cell disease.
- Take your child to a Sickle Cell Treatment Center regularly.
- Take your child to other medical and dental specialists, as recommended by your child's regular doctor and sickle cell specialist.

HOSPITAL STAY

- Hospital stays can be frightening for a child.
- Prepare your child by playing hospital.
- Take your child to visit the hospital when the child is not sick.
- When your child is in the hospital, visit him/her as often as possible.

DENTAL CARE

- Take your child for a dental check-up at least once a year.
- Tell the dentist and dental hygienist that your child has sickle cell disease.
- If your child needs to have a tooth pulled, major dental work or general anesthesia is needed, ask your dentist to speak with your sickle cell doctor first.
- Learn how fluorides, sealants, and diet may help protect your child's teeth.
- Learn what you can do at home and what dental programs are offered at your child's school.
- Antibiotics may be necessary before dental work.

GROWTH AND DEVELOPMENT

- Children with sickle cell anemia may be shorter than their friends of the same age. They also may have delay in puberty (sexual development). Usually, their height and sexual development will eventually be normal.
- A peer support group of other children with sickle cell disease may help your child deal with feelings and questions. Ask your child's doctor about a group in your area.
NUTRITION AND DIET

Your child should:

- Eat a well balanced diet for the child’s age group. There is no special diet for a child with sickle cell disease.
- Drink plenty of fluids such as water and juice to avoid dehydration (the loss of too much water).
- Drink extra fluids during increased physical activity, when temperatures are warm, and during fevers.
- Avoid caffeinated drinks like regular coffee and soda.
- During a crisis, it is more important to drink fluids than it is to eat.
- Take the vitamin folic acid daily, if advised by your doctor.

PHYSICAL ACTIVITY

- Children should take an active part in any physical activity that interest them. Let them set their own pace.
- Aerobic exercise like running, swimming, walking, and biking are okay. The body loses more fluid during exercise so it is important to drink more fluids.
- Swimming in very cold water may trigger a painful crisis in some children. Before the child goes swimming, make sure the water is not too cold. If the water feels cold to the touch, it is probably too cold for swimming. Children should dry off with a towel as soon as they come out of the water.

WEATHER

Extremes of heat and cold should be avoided. Extreme temperatures may trigger a crisis. Too many clothes in hot weather should be avoided. Warm clothes are needed in cold weather but the child should not be overdressed.

TRAVEL

- Pack a thermometer, emergency (information) card, antibiotics, Tylenol, and make sure your child can get plenty to drink.
- Persons with sickle cell disease can have trouble during flights at high altitudes in a non-pressurized airplane. Since commercial airlines are pressurized, flights on these planes are not a problem.
- It is a good idea to discuss travel plans with the Sickle Cell Team. They can often suggest a hospital or doctor who may be helpful if your child has a problem far from home.

DISCIPLINE

Treating a child with sickle cell disease in a normal manner is an important part of helping the child develop a healthy identity. For parents to agree on how to discipline a child is just as important for the child with sickle cell disease as it is for other children.
PREGNANCY AND SEXUAL ACTIVITY

Teenage and adult women with sickle cell disease have many more problems with pregnancy than other women. They can have children, but they need early and regular medical care during pregnancy.

Unplanned pregnancy should be avoided. As soon as you are aware that you are pregnant, please let the doctors at the hematology center know. You should also arrange to have regular visits with an obstetrician as soon as possible once you know you are pregnant.

Women with sickle cell disease can usually have normal sexual activity and can usually have children without serious problems. When your child is ready, ask your child’s doctor about:

- risk of and protection against, sexually transmitted diseases such as AIDS.
- pregnancy and birth control options.
- genetic counseling (risk of having children with sickle cell disease and the options that are available).

YOUR CHILD IN SCHOOL

Your child can grow and become almost anything he or she wants to be in life. There are people with sickle cell disease who have become doctors, lawyers, teachers, nurses, etc.

Your child should be urged to fully participate in school at an early age. Register your child in pre-school and/or kindergarten at the right age. It is important for you and school staff to understand, however, that your child may miss a lot of school because of sickle cell disease.

It is important to talk to your child's teachers and school nurse about sickle cell related problems your child may have at school. The Sickle Cell Treatment Center staff is available to help the teachers, nurse, principal, and your child's classmates understand sickle cell.

THE SCHOOL SHOULD KNOW:

- Your telephone number at home and/or work so that you can be called if your child becomes ill at school.
- The telephone number of the Sickle Cell Treatment Center.
- Your child may need to use the bathroom more often than other children since children with sickle cell pass more urine (pee) than other children.
- Your child may need to be allowed to drink lots of fluids.
- Your child may tire more easily in gym class.
- Your child should not be exposed to hot or cold temperatures without proper clothing. For example during a fire drill in cold weather, your child must be allowed to put on a coat, gloves, and a hat to go outside.
- Your child should be sent to the school nurse if he/she does not feel well or looks tired.

YOU SHOULD KNOW IT IS IMPORTANT TO:

- Call the school when your child is absent.
- Arrange for homework to be sent home when your child is absent.
- Urge your child to keep up with school work when he/she is absent or when he/she has mild pain.
- Talk to the school social worker about tutoring and other services that may be available to your child.
- Talk to the school nurse about developing an individualized health plan (IHP) for your child.
**MEDICAL WORDS**

**Acute Chest Syndrome** - a pattern of chest or back pain, cough and difficulty breathing that may signal the start of a sickle cell crisis in the chest; a pneumonia.

**Anemia** - low red blood cell count.

**Aplastic Crisis** - bone marrow temporarily stops making red blood cells.

**Aseptic Necrosis** - gradual breakdown or destruction of bones usually involving the hips, knees or elbow joints.

**Bilirubin** - yellow substance which comes from the breakdown of red blood cells.

**Bone marrow** - the place inside your bones where blood cells are made.

**Carriers** - persons who carry a gene for a disease but do not have the disease.

**Chemotherapy** - medicines that are used to fight cancer.

**Chronic transfusions** - a blood transfusion that is given about once each month to hopefully prevent further complications from sickle cell disease.

**Crisis** - an emergency related to sickle cell disease.

**Dehydration** - loss of body fluids from sweating and urination.

**Diarrhea** - frequent and watery bowel movement.

**Genes** - message carriers which are passed on to children through the mother's egg and the father's sperm. Genes carry the messages for things like eye color, hair color, blood type, and the kind of hemoglobin a person will have.

**Genetic Centers** – Medical facilities where Genetic specialists can arrange for genetic tests and can answer questions on how genetic conditions may be passed down in families.

**Geneticist** - a doctor who specializes in hereditary conditions.

**Hematologist** - doctor who specializes in blood disorders.

**Hematuria** - blood in the urine.

**Hemoglobin** - part of blood that carries and delivers oxygen to all parts of the body.

**Hemoglobin SC Disease** - a variant of sickle cell disease.

**Hemoglobinopathy** - abnormal hemoglobin.

**Inherited** - passed on from the father and mother to their child.
Jaundice - yellow pigment (color) to eyes and skin.

Meningitis - brain infection.

Osteomyelitis - bone infection.

Platelets - part of the blood that helps make blood clot.

Pneumonia - lung infection.

Priapism - prolonged, unexpected painful erections.

Sepsis - serious blood infection.

Sickle beta-thalassemia - a variant (type) of sickle cell disease.

Sickle Cells - red blood cells that look sickle-shaped ( ) under a microscope. This shape can cause the cells to block the blood vessels and limit the amount of oxygen to the body.

Sickle Cell Anemia (SS) - most common sickling disease in the United States.

Sickle Cell Crisis - an emergency related to sickle cell disease.

Sickle Cell Disease - another term for Sickle Cell Anemia

Sickle Cell Trait - a normal hemoglobin gene is inherited from one parent and a sickle hemoglobin gene from the other.

Sickling Diseases - sickle cell anemia (SS), sickle- C (SC), and sickle beta-thalassemia (S-B-thalassemia).

Spleen - an organ on the left side of the belly. It acts as a filter for blood.

Splenic Sequestration - spleen overfills with blood.

Stroke - clogging of blood vessels to the brain.

Thalassemia major - an inherited disease with an abnormal hemoglobin that requires lifelong transfusions but does not put a child at risk for infection or crisis.

Thalassemia minor (trait) - an abnormal type of hemoglobin that is sometimes inherited along with sickle hemoglobin to produce sickle- beta thalassemia. Having the trait by itself, is not harmful.

Urination - passing water or peeing.

Vessels - the tubes that blood flows through such as veins, arteries, and capillaries.
SICKLE CELL DISEASE ON THE INTERNET

The Internet is a very useful source of information about sickle cell disease. There are hundreds of web sites devoted to this topic. Many of these sites provide information about sickle cell disease including commonly asked questions, complications of the disease, treatments and potential cures. There is information about the STOP study, hydroxyurea, and bone marrow transplant (see other sections of this manual). Some web sites will provide you with lists of organizations that have more information about sickle cell disease or can put you in contact with other persons who have sickle cell anemia.

If you want to go directly to a specific web site, here is an interesting one. Please be sure to type them exactly as they are written here.

- [www.sicklecelldisease.org](http://www.sicklecelldisease.org)  
  Sickle Cell Disease Association of America, Inc.

- [www.medicalhomeinfo.org/screening/SCDindex.html](http://www.medicalhomeinfo.org/screening/SCDindex.html)  
  American Academy of Pediatrics – Sickle Cell Disease Pages

If you want to explore on your own, use one of the search engines (e.g. EXCITE, YAHOO, WEBCRAWLER) and search the term: "Sickle Cell Anemia". You will come up with a number of matches that will start you on your search for topics related to sickle cell disease.

Web sites often share common information so that after searching a few sites, you will probably encounter much repeated information.

Some of the doctors and nurses at the Sickle Cell Treatment Centers can guide you through the steps needed to gain access to this service. Some public libraries, colleges and high schools have computers with Internet access and may be able to assist you in your search.
SICKLE CELL TREATMENT CENTERS

There are several Sickle Cell Treatment Centers located throughout the state of New Jersey for care of children, teenagers and young adults with sickle cell disease and other hemoglobinopathies. A list of centers is included at the end of this section.

Every member of the center’s health care team plays a role in helping you and your child. Each center is headed by a pediatric hematologist, a doctor that specializes in blood disorders of children. Other doctors, nurses, and social workers are also on staff to assist in treating these children and in helping families manage their care. The staff of the center will help you understand the information in this guide. They will also show you how to take a temperature and how to feel the spleen. They will examine your child during center visits. The social worker will help you deal with your concerns and issues about school and work. The staff can also arrange for you and your child or other family members to also be seen by other health professionals including: geneticist or genetic counselor, psychologist, and nutritionist. See list of genetic centers at the end of this section.

EMERGENCIES OR PROBLEMS

When your child has an emergency or problem, call the Sickle Cell Treatment Center or take your child to the nearest hospital emergency room.

APPOINTMENTS

Children with sickle cell disease are seen frequently in the first year of life. After that first year, children are usually seen two to four times each year. More frequent appointments may be necessary if your child has experienced major problems related to having sickle cell disease.

These visits are very important to help keep your child well. If you cannot make an appointment, please let the staff know as soon as possible so the appointment can be rescheduled.

ROUTINE VISITS

Routine visits to a center are set up so the staff can do the following:

- Recognize problems early when they can be more easily treated;
- Make sure that proper medicines are being given in proper doses;
- Be sure that the proper immunizations have been given to help prevent infection (in addition to the normal baby shots, other specific immunizations are given to children with sickle cell disease to help prevent infections).
- Answer your questions and continue to teach you more about the disease.

MEDICINES

Children with sickle cell disease should take Penicillin by mouth twice a day unless they are allergic to penicillin. In that case, a different antibiotic will be given. This is to help protect your child against life-threatening infection.

Children with sickle cell disease should also take Folic Acid, 1 mg, each day. This vitamin is needed to help your body make hemoglobin. Since your child makes hemoglobin more rapidly than normal, he or she will use up this vitamin faster than normal and so your child requires more than the usual amount of this vitamin.

TRY TO LET THE CENTER STAFF KNOW BEFORE YOU RUN OUT OF MEDICINE OR VITAMINS.
Comprehensive Sickle Cell/Hemoglobinopathies
Treatment Centers in New Jersey
*SUPPORTED BY THE NEW JERSEY DEPARTMENT OF HEALTH AND SENIOR SERVICES

Children’s Hospital of New Jersey
Newark Beth Israel Medical Center
Valerie Fund Children’s Center
201 Lyons Avenue
Newark, NJ 07112-2094
Phone: (973) 926-7161
Fax: (973) 282-0395

The Children’s Hospital at St. Joseph’s Hospital and Medical Center
703 Main Street
Paterson, NJ 07503-2691
Phone: (973) 754-3230
Fax: (973) 754-3331

University Hospital/UMDNJ
Pediatric Hematology/Oncology
UH Room F342
150 Bergen Street, Suite 5400
Newark, NJ 07103-2499
Phone: (973) 972-5106 or 972-0658
Fax: (973) 972-1340

Bristol-Meyers Squibb Children’s Hospital
at Robert Wood Johnson University Hospital
Affiliated with UMDNJ/RWJ Medical School
Division of Pediatric Hematology/Oncology
195 Little Albany Street
New Brunswick, NJ 08903-0029
Phone: (732) 235-5437
Fax: (732) 235-6462

Children’s Hospital of Philadelphia
New Jersey Section of Hematology/Oncology
Specialty Care Center
1012 Laurel Oak Road
Voorhees, NJ 08043
Phone: (856) 435-7502
Fax: (856) 627-2183 (office)

Genetic Centers in New Jersey
*SUPPORTED BY THE NEW JERSEY DEPARTMENT OF HEALTH AND SENIOR SERVICES

*Children’s Hospital of New Jersey
Newark Beth Israel Medical Center
201 Lyons Avenue
Newark, NJ 07112
Phone: (973) 831-6020

*St. Joseph’s Hospital and Medical Center
Section of Genetics
703 Main Street
Paterson, NJ 07503-2691
Phone: (973) 754-2727
Outreach Clinic: Fairfield

*Hackensack University Medical Center
Genetics Service
Don Imus Pediatric Center-Rm. 258
30 Prospect Avenue
Hackensack, NJ 07601-1991
Phone (201) 996-5264
Outreach Clinics: Hoboken, Parsippany

*UMDNJ/NJ Medical School
Center for Human & Molecular Genetics
90 Bergen Street, Suite 5400
Newark, NJ 07103-2499
Phone: (973) 972-3300
Outreach Clinics: Pompton Plains, West New York

*Saint Peter’s University Hospital
Institute for Genetic Medicine
254 Easton Ave.
New Brunswick, NJ 08903
Phone: (732) 745-6678

*Cooper Hospital/University Medical Center
Division of Genetics
3 Cooper Plaza Suite 309
Camden, NJ 08103 -1400
Phone: (856) 968-7255
Outreach Clinic: Children’s Regional Center at Voorhees