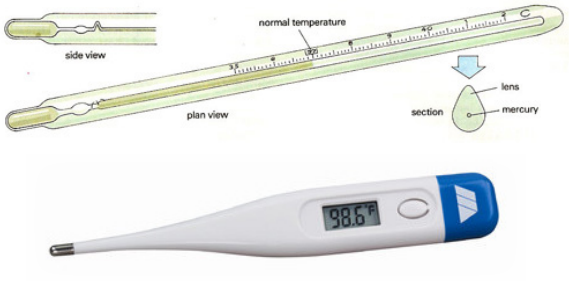
Calling the physician:

It is important to establish a relationship with your doctor and nurse so that you get to know and trust each other. This relationship will make it easier for the doctor to take care of your baby, whether in the home or hospital.

When calling the doctor, he or she will need to know:

1. Is there a fever? If so, how high is it? You must keep a thermometer on hand and learn how to read it so that you can give your doctor the exact temperature readings. This is very important for your baby's health.



Thermometer

2. Have you given your baby any medication? If so, what kind, how much and when was the last dose given?
3. Is the baby eating and/or drinking fluids? It is very important that people with sickle cell anemia drink lots of fluids. You should pay close attention to the amount of fluid intake. If your baby is not drinking fluids, please let the doctor or nurse know.
4. Is there vomiting?
5. Is there pain? If so, where? Have you given your baby any pain medication? If so, how much, and what kind? When was the last dosage?
6. Does your baby have any of the symptoms listed previously?

Preparing for emergency room or hospitalization:

If you need to take your baby to the emergency room or have your baby admitted to the hospital, here is what you will need:

Proof of insurance: Medicaid card, Medicare card, and/or private insurance card.

Clinic Card: LSU Health Science Center, Tulane, Sickle Cell Center or other.

Sweater or blanket: to keep you and your baby warm *(emergency rooms are usually cooler than home).*

For further information, contact the Sickle Cell Center in your area, your private physician or your local health clinic.

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For: State of Louisiana Genetic Diseases Program

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There are even more ways to get up-to-date health information, news and emergency updates from LDH.

www.facebook.com/LaDeptHealth

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Sickle Cell Anemia

(Hemoglobin SS Disease)

What Every Parent Should Know

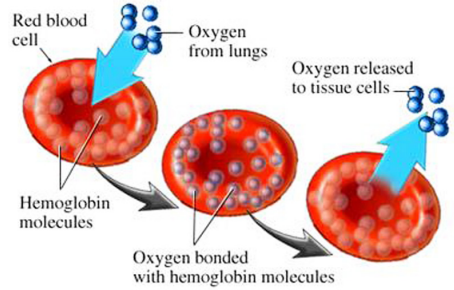
LOUISIANA DEPARTMENT OF HEALTH
Public Health

Genetic Diseases Program
1450 Poydras St., Suite 2046
New Orleans, LA 70112
(504) 568-8254
www.ldh.la.gov/genetics

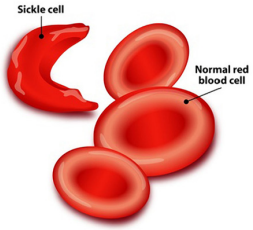
This disease is tested by newborn screening in Louisiana. FIND IT EARLY!

What is sickle cell anemia?

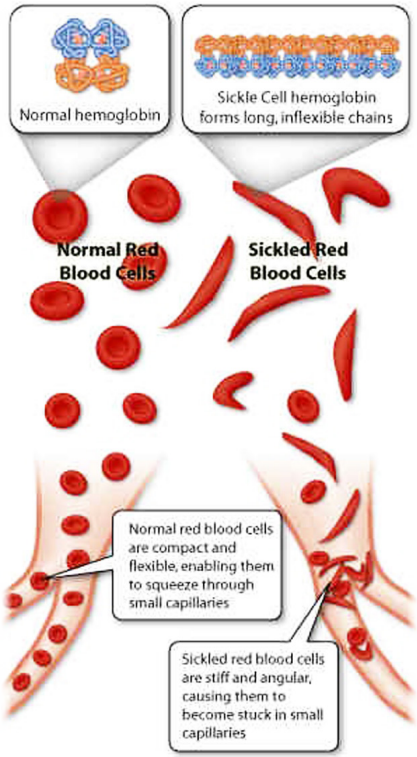
Sickle cell anemia, also known as hemoglobin SS (HSS) disease, is an inherited disorder of the hemoglobin in red blood cells. Hemoglobin is the oxygen-carrying portion of the red blood cell.



Hemoglobin Function



Red blood cells (RBCs) transport the oxygen from the air we breathe to all parts of our body. RBCs are usually round, soft and pliable.



In sickle cell anemia, RBCs can sometimes become distorted and block off blood vessels. They also do not live as long as RBCs in individuals without sickle cell anemia do.

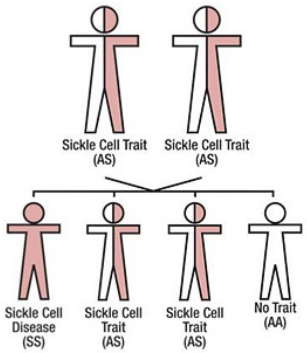
How did my baby get sickle cell anemia?

Sickle cell anemia is inherited from both parents. Like hair color, general body build and other physical characteristics, it is passed down from the parents through genes.

Genes come in pairs; for each characteristic there is one gene from the mother and one from the father. One pair of genes determine hemoglobin. If your child has sickle cell anemia, it means that the baby inherited a gene for the changed hemoglobin from both the mother and father. The baby did not inherit genes for normal hemoglobin A. Your baby therefore, has SS hemoglobin, or sickle cell anemia.

How it works:

Most people who carry genes for hemoglobin S do not have sickle cell anemia. They are carriers and have what is known as sickle cell trait. This means that they have inherited one gene for normal hemoglobin A and one for the changed hemoglobin. They have A hemoglobin. However, when a child is conceived, parents who both have AS hemoglobin can pass either the gene for A or the gene for S on to their child.



With each pregnancy there is a 25-percent chance that the child of carrier parents will inherit sickle cell anemia, a 50-percent chance that the child will be a sickle cell carrier (AS) and a 25-percent chance that the child will have normal hemoglobin (AA).

Caring for your baby with sickle cell anemia:

Even though your baby has a serious, chronic disease, he or she will behave like most babies who do not have a medical problem. As such, your baby should be cared for as any other newborn baby. However, there are some signs you should be aware of so that quick medical care can be obtained as soon as possible when needed.

Fever: If your baby feels warm, you should take his or her temperature. If the temperature is 100 degrees, you should immediately call the physician or nurse. If the temperature is 101 degrees or above, take the baby to your doctor or emergency room for treatment immediately. This may require that your baby be hospitalized and placed on intravenous antibiotics. Fever in babies with sickle cell anemia may indicate very serious, life-threatening infections.

Splenic sequestration: As blood is filtered through the spleen, sometimes sickled cells may become trapped in the spleen and keep the blood from moving out of the spleen. Symptoms of this are swollen stomach, unusual sleepiness or irritability (fretfulness). This is a serious, life-threatening problem. Go to an emergency room immediately. Have them call your baby's doctor. Your doctor or nurse can show you how to recognize this problem during your regular clinic appointment.

Swollen hands and feet: One of the first symptoms of sickle cell anemia in babies may be swollen hands and feet. The hands and feet feel warm and are sensitive to the touch. Contact your doctor for instructions on how to make your baby comfortable. This is NOT an emergency.

Anemia: Because sickle cells do not live as long as normal cells, your baby will be anemic. That is, your baby will have a low blood count. This is usually nothing to be concerned about; the baby's body will adjust to this lower blood level. However, the baby may have less energy than other babies. If the baby appears listless, contact the doctor immediately, as this may indicate a sudden and severe worsening of the anemia.

Jaundice: When cells are destroyed, they produce products in the blood that are carried off as waste. Sickle cells are destroyed at a much faster rate than regular red blood cells, so more of these waste products are found in the blood stream of individuals with HSS disease. These products can cause jaundice, giving a yellowish tint to the white of the eyes or an orange tint to the skin. Call your baby's doctor if this yellowish tint becomes more pronounced than usual.

Stroke: Strokes can occur in a small number of patients with sickle cell disease. These strokes can happen to children as young as 10 months of age. Any unusual drooling, weakness of muscles in the face (twisted or hanging mouth), inability to move arms or legs or dragging of the legs could be signs of stroke. **Go to an emergency room immediately**. Have them call your baby's doctor.

Skin problems: Breaks in the skin or untreated insect bites can sometimes lead to serious problems in patients with sickle cell disease. Sores, cuts and insect bites should be cleaned and treated with a mild antiseptic. If the area is not healing or begins to get swollen or reddish, contact the baby's doctor or nurse.