



HEALTH NEWS FROM THE DCF MEDICAL TEAM

SICKLE CELL DISEASE

What is Sickle Cell Disease?

Sickle Cell Disease is an inherited condition that affects the red blood cells and primarily affects those of African descent but can occur in individuals of Mediterranean, Indian, or Middle Eastern descent. The disease changes the shape of the red blood cells. When this occurs, the cells cannot readily flow through the blood vessels. The abnormally shaped cells cause an increase in blood thickness and decreased blood flow to the body. This reduced blood supply results in less oxygen getting to the body and can cause acute pain, cell destruction, and organ damage.

How does a child inherit Sickle Cell Disease?

- ◆ Sickle Cell Disease is a genetic condition that is present at birth.
- ◆ If one parent is a carrier, each child has a 25% chance of inheriting the trait. If both parents are carriers, each child has a 50% chance of inheriting the trait and a 25% chance of inheriting the disease.
- ◆ Genetic counseling should be provided to those with the disease or trait during adolescence and before each pregnancy.

What are the symptoms of Sickle Cell Disease?

The first symptoms do not usually appear until the first 4-6 months of life. People with Sickle Cell Disease usually have anemia, and also at times have other symptoms:

Common Symptoms
Abdominal pain
Bone pain
Breathlessness
Delayed growth and puberty
Fatigue
Fever
Paleness
Rapid heart rate

Other Symptoms
Chest pain
Excessive thirst/ Frequent urination
Problems during pregnancy
Painful and prolonged erection (10 - 40% of men with SCD)
Poor eyesight/blindness
Strokes
Skin ulcers
Enlarged Spleen

What is the treatment for Sickle Cell Disease?

- ◆ The goal of treatment is to manage and control symptoms and to limit the frequency of crises.
- ◆ People with Sickle Cell Disease need ongoing treatment, even when they are not having a painful crisis. They should take supplements of folic acid and avoid factors that can lead to a Sickle Cell Crisis: cold, dehydration, stress, overexertion and acute illness.
- ◆ Antibiotics and vaccines are given to prevent infections, which are common in children with sickle cell disease.
- ◆ Hydroxyurea is a drug that may be used to reduce the number of painful episodes.
- ◆ Blood transfusions are used to treat a sickle cell crisis and may also be used regularly to help prevent strokes.
- ◆ During a Sickle Cell Crisis, pain is treated with pain medicines and by drinking plenty of fluids and sometimes intravenous fluids. It is important to treat the pain. Non-narcotic medications may be effective, but sometimes large doses of narcotics are prescribed.

When is Sickle Cell Disease an Emergency?

“Sickle Cell Crisis” is a serious exacerbation of the disease. Most episodes last from 5 to 7 days. This can be life threatening and requires immediate medical attention. This condition can result in decreased blood flow to the body, painful enlargement of the spleen, worsening of anemia and chest pain. Call the child’s healthcare provider if there are signs of possible Sickle Cell Crisis or infection such as fever, body aches and fatigue.

References: www.CDC.gov; www.ncbi.nlm.nih.gov/pubmedhealth/PMH0001554; “Guidelines for the Treatment of People with Sickle Cell disease”, Sickle Cell Advisory Committee of New York, Puerto Rico and The Virgin Islands, March 2002.