PARENT FACT SHEET

DISORDER

Hemoglobin S/C Disease (Hb S/C)

CAUSE

Sickle cell disease is an inherited blood disorder characterized by defective hemoglobin (a protein in red blood cells that carries oxygen to the tissues of the body).

Sickle cells only live for about 15 days, while normal hemoglobin can live up to 120 days. Also, sickle cells risk being destroyed by the spleen because of their shape and stiffness. The spleen is an organ that helps filter the blood of infections and sickled cells get stuck in this filter and die. Due to the decreased number of hemoglobin cells circulating in the body, a person with sickle cell disease is chronically anemic. The spleen also suffers damage from the sickled cells blocking healthy oxygen carrying cells. Without a normal functioning spleen, these individuals are more at risk for infections. Infants and young children are at risk for life-threatening infections.

Sickle cell - hemoglobin C disease - The child has both HbS and HbC. This is often referred to as HbSC. Hemoglobin C causes red blood cells, called target cells, to develop. Having just some hemoglobin C and normal hemoglobin, a person will not have any symptoms of anemia. However, if the sickle hemoglobin S is combined with the target cell, some mild to moderate anemia may occur. These children often suffer some of the complications associated with HbSS, sickle cell disease, but to a milder degree. Vaso-occlusive crises (the flow of blood is blocked because the sickled cells have become stuck in the blood vessels), organ damage from repeated sickling and anemia, and high risk for infection are all similar traits for HbSS and HbSC.

TREATMENT OPTIONS

- Pain medications (for sickle cell crises).
- Drinking plenty of water daily (8 to 10 glasses) or receiving fluid intravenously (to prevent and treat pain crises).
- Blood transfusions (for anemia, and to prevent stroke; transfusions are also used to dilute the HbS with normal hemoglobin to treat chronic pain, acute chest syndrome, splenic sequestration and other emergencies.)
- Penicillin (to prevent infections).
- Folic acid (to help prevent severe anemia).
- Hydroxyurea (a medication recently developed that may help reduce the frequency of pain crises and acute chest syndrome; it may also help decrease the need for frequent blood transfusions. The long-term effects of the medication are unknown.)
- Bone marrow transplant (has been effective in curing some children with sickle cell disease; the decision to undergo this procedure is based on the severity of the disease and a suitable bone marrow donor. These decisions need to be discussed with your child's physician.)

IF TREATED

With early detection and comprehensive medical care, most people with sickle cell disease are in fairly good health most of the time. Most individuals can be expected to live well into adulthood, enjoying an improved quality of life including the ability to choose a variety of education, career, and family planning options for themselves.