SICKLE CELL DISEASE

Description

Sickle cell disease is a genetically inherited disorder of the red blood cells (RBCs), the cells responsible for carrying oxygen to the rest of the body. Oxygen is carried by the RBC on a molecule called hemoglobin. In sickle cell disease the normal hemoglobin is replaced by an abnormal one, and in conditions of low oxygen and stress, such as strenuous exercise or traveling to high altitudes, the abnormal RBCs become rigid and form a sicklelike shape. The sickle cell also has a chemical on its surface that sticks to blood vessel walls. As a result, these abnormally shaped, rigid, and sticky RBCs cannot squeeze through the small blood vessels, causing blockage and depriving tissues and organs of oxygen. Sickled blood cells also have a shorter life span compared with normal RBCs. This means the bone marrow must work harder to produce new RBCs and often cannot keep up with the rate of cell death, leading to anemia (low blood count). The severity of sickle cell disease generally depends on three factors: the degree of oxygen deprivation; the concentration of hemoglobin within the cell (the lower the better); and the amount of a protective molecule called hemoglobin F (for *fetal*). People with the sickle cell gene who continue to carry some fetal hemoglobin are better protected and suffer less severe forms of the disease.

Risk Factors

- Family members with sickle cell disease (both parents have to have the abnormal gene)
- Family origins in areas with high incidence of malaria (Africa, India, the Mediterranean, South and Central America, the Caribbean, and the Middle East)
- Physical stress
- Illness
- Exertion
- Dehydration
- High altitude

Common Signs and Symptoms

- Fever
- Swelling of the hands and feet
- Enlargement of the belly (heart, liver, and spleen)
- Frequent lung infections
- Fatigue
- Irritability
- Yellowing of the skin (jaundice)
- Severe bone and joint pain
- Delayed puberty
- Shortness of breath
- Pain in the belly, especially in the upper right side of the abdomen
- Nausea
- Prolonged, sometimes painful erections (priapism)
- Rapid or labored breathing
- Frequent infections

Prevention of Sickle Cell Crisis

- There are no proven methods for preventing sickle cell crises.
- Regularly follow up with your doctor.
- Avoid dehydration—drink plenty of fluids.
- Avoid high-altitude travel, especially rapid increases in altitude.
- Avoid stress.
- Get plenty of rest.
- Stay warm.
- Female patients may dink cranberry juice to help prevent urinary tract infections.
- Maintain good nutrition by eating green, red, and yellow vegetables; fruits; and juices that are rich in antioxidants and other important nutrients.
- Eat fish and soy products that are high in omega-three fatty acids.
- Make sure you consume enough folic acid, zinc, vitamin E, vitamin C, and L-glutamine.
- Blood transfusions.
- Immunizations, particularly against the flu and some bacteria, may reduce the risk of infection and thus sickle crisis.

General Treatment Considerations

For mild pain relief, common medications such as acetaminophen (Tylenol) or the class of drugs known as nonsteroidal anti-inflammatory drugs (NSAIDs) are often sufficient. Stress reduction techniques and relaxation methods appear to be helpful. The basic objectives for managing a sickle cell crisis are control of pain and rehydration by administration of fluids. Oxygen is typically for patients with difficulty breathing. High-risk children may be given blood transfusions to prevent sickle crisis. Other treatments include medications to help the body produce protective hemoglobin. You should discuss this with your doctor to see if it is right for you.

Notes: (Up to 4400 characters only)

Notes and suggestions