Sickle Cell Disease

What is Sickle Cell Disease?

A genetic disease, most prevalent in the African-American community, sickle cell disease (also known as "sickle cell anemia") is a disease in which red blood cells are an abnormal crescent shape. Red blood cells are normally shaped like a disc. They clump together, blocking blood vessels and creating intense pain. The disease's other "crises" can be divided into three groups:

- Hemolytic crisis occurs when damaged red blood cells break down rapidly
- Splenic sequestration crisis is when the spleen enlarges and traps the blood cells
- Aplastic crisis results when an infection causes the bone marrow to stop producing red blood cells

Symptoms

Common symptoms include:

- Attacks of abdominal pain
- Bone pain
- Breathlessness
- Delayed growth and puberty
- Fatigue
- Fever
- Jaundice
- Paleness
- Rapid heart rate
- Susceptibility to infections
- Ulcers on the lower legs (in adolescents and adults)
- Strokes

Diagnosis

Tests commonly performed to diagnose and monitor patients with sickle cell anemia include:

- Complete blood count (CBC)
- Hemoglobin electrophoresis
- Sickle cell test
- Sonograms to look for abnormal blood flow in the head
Treatments

Common treatments for sickle cell disease include:

- Pain medicines to treat painful crisis
- Fluids
- Oxygen
- Antibiotics to prevent and treat crises
- Immunizations
- Hydroxyurea (a drug that increases fetal hemoglobin and prevents painful crisis)
- Transfusions
- Bone Marrow Transplantation

Sickle Cell Disease is treated by our Division of Pediatric Hematology.