All About:
Sickle Cell Disease

What is sickle cell disease?

Sickle cell disease (SCD) is a group of inherited red blood cell disorders.

- Hemoglobin is the protein in red blood cells that carries oxygen throughout the body.
- Each person inherits two hemoglobin genes, one from each parent. A normal gene will make normal hemoglobin (A). A specific change in the hemoglobin gene will make sickled hemoglobin (S).
- In SCD, the red blood cells become hard and sticky and look like a C-shaped farming tool called a sickle.
- Sickle cells can get stuck in small blood vessels and block the flow of blood and oxygen to organs within the body. These blockages can cause many problems.
- The most common types of sickle cell disease are hemoglobin SS, hemoglobin SC, and sickle beta thalassemia.

What causes sickle cell disease?

SCD is inherited in the same way that people get the color of their eyes, skin, and hair.

- A person with SCD is born with it.
- People cannot catch SCD from being around a person who has it.
- If a person has sickle cell disease, they inherit a sickle hemoglobin from each parent.

What are the potential health problems of sickle cell disease?

Infections
The spleen helps the body fight infections. Sickle cells may get caught in the spleen, preventing it from working as well as it should. As a result, people with sickle cell disease are more likely to get infections. Children are put on antibiotics until their immune system matures.

Anemia
Sickle cells do not live as long as normal red blood cells. This causes anemia, or a low blood count. Anemia can cause weakness and fatigue.

Pain
Sickle cells that get caught in the small blood vessels of the body cause the interruption of oxygen and blood flow; these sickling crises can be very painful. This includes pain and swelling of the hands and feet in small children.

Acute Chest Syndrome
Blockage of the flow of blood to the lungs can cause acute chest syndrome (ACS). ACS is similar to pneumonia; symptoms include chest pain, coughing, difficulty breathing, and fever. It can be life threatening and should be treated in a hospital.

Stroke
Sickle cells can clog blood flow to the brain and cause a stroke. A stroke can result in lifelong disabilities and learning problems. Children under 16 are at the highest risk for stroke.

Jaundice
This is a yellowing of the eyes and skin. It is painless and occurs because of rapid breakdown and death of sickled red blood cells.

Leg Ulcers
Poor circulation due to blockage of blood vessels by sickle cells can lead to sores that are difficult to heal. These often occur around the ankles.

Complications During Pregnancy
A woman with sickle cell disease can have a healthy baby. However, risks are involved; both she and the baby should be closely monitored by a healthcare provider. Prenatal care is very important!

Other problems include:

- chest pain and trouble breathing
- organ damage
- gallstones
- blood in the urine
- eye disease
- painful erections in men

How is sickle cell disease managed?

Individuals with sickle cell disease should receive regular medical care from a hematologist and a general medical care provider.

Routine care is very important!

Treatments may include a medication called hydroxyurea which increases the body's level of fetal hemoglobin which prevents sickling, as well as antibiotics for infections or prevention of infections, and medicines for painful events.

Everyone should eat a balanced diet, and receive regular check-ups and immunizations. Children with sickle cell disease require a few special immunizations. A yearly flu shot is also recommended.

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