

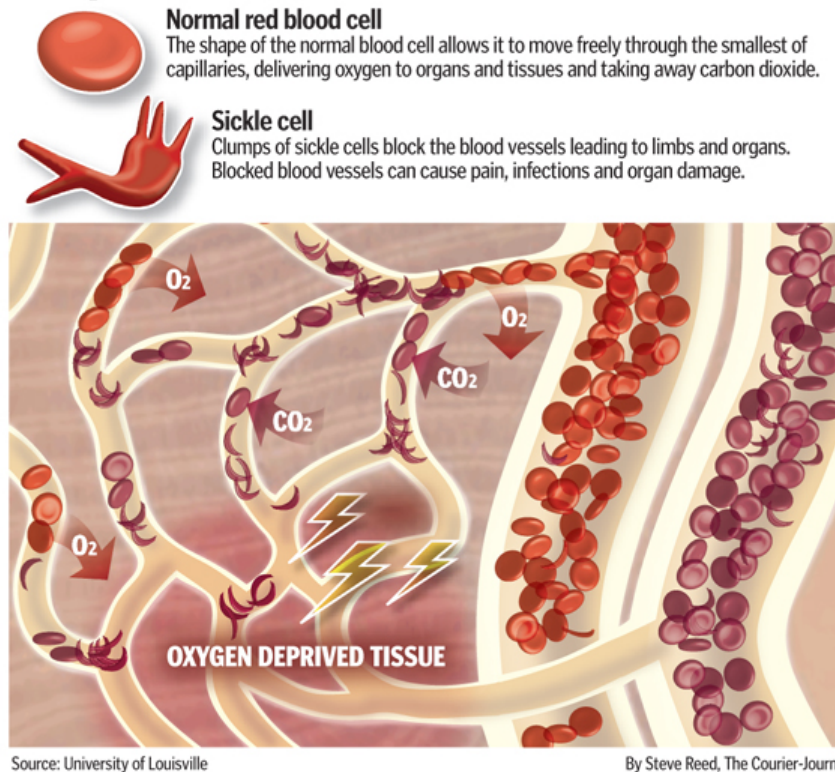
## Graphic: The Problem with Sickle Cells

February 21, 2010

**What is it?:** A disease in which the body makes sickle-shaped red blood cells instead of normal doughnut-shaped cells. Clumps of sickle cells block blood flow in the vessels leading to the limbs and organs, causing pain, infections and organ damage.

**Causes:** Those who have the disease inherit two recessive genes, one from each parent. Carriers without the disease are considered to have sickle cell “trait,” which is thought to be protective against a deadly type of malaria. It developed in areas where malaria is endemic.

### The problems with sickle cells



**Who's at Risk?:** It is common in people whose families come from areas prone to this malaria, such as Africa, South or Central America (especially Panama), the Caribbean islands, Mediterranean countries, India and Saudi Arabia. In the United States, it affects about 70,000 people, mainly African Americans.

**Signs and Symptoms:** Commonly fatigue, shortness of breath, dizziness, headache, coldness in the hands and feet, pale skin, chest pain. Sudden pain throughout the body is also common and is called a “sickle cell crisis,” which occurs when sickle blood cells form clumps in the bloodstream.

**Complications:** Hand-foot syndrome, which causes fever and pain and swelling in hands and feet; splenic crisis, which causes the spleen to shrink and may require blood transfusions; infections; acute chest syndrome, similar to pneumonia; pulmonary arterial hypertension, which makes it hard for the heart to pump blood through the lungs; delayed growth and puberty in children; stroke; eye problems; gallstones; ulcers on the legs; and multiple organ failure.

**Diagnosis:** Newborns are screened for the disease, and if the test shows a problem, a second blood test is done.

**Treatment:** Among others, bone marrow transplants; medicines and fluids for pain; a medicine called hydroxyurea to help prevent painful crises; blood transfusions.

**Prognosis:** In the past, sickle cell patients often died from organ failure between ages 20 and 40. Now some patients can live into their 50s and beyond – though half still die by 40.

Sources: National Heart, Lung and Blood Institute; National Institutes of Health; Dr. Suzanne Ildstad, University of Louisville.