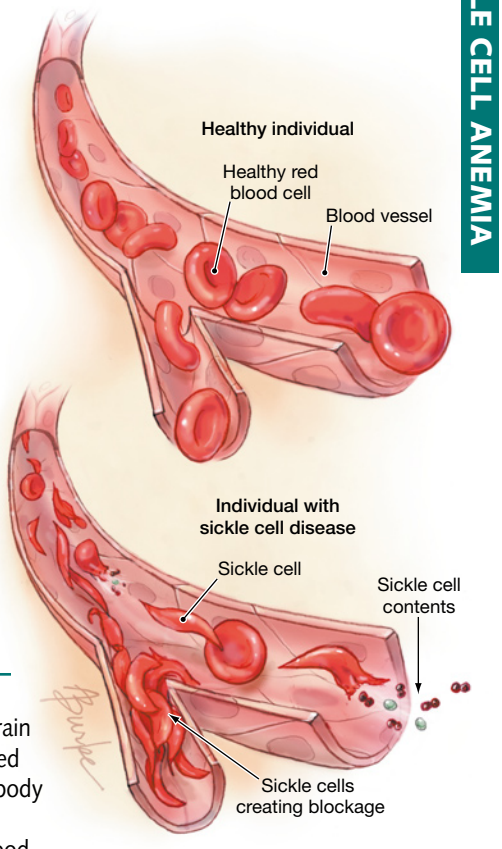
Sickle Cell Vasculopathy

Sickle cell disease affects the red blood cells and is one of the most common **genetic** (inherited) diseases. It affects 70 000 people in the United States. The normal red blood cell carries oxygen and delivers it to the tissues. Sickle-shaped red blood cells have difficulty traveling through the blood vessels to deliver oxygen to the tissues. These sickle cells are more stiff and can get stuck inside the blood vessels. People with sickle cell disease often have problems caused by blockages of these sickle cells within their blood vessels. Compared with normal red blood cells, sickle cells are more fragile and are often broken up within the blood vessels. When red blood cells are broken, the affected person becomes **anemic** (low blood count) and may develop a yellow discoloration in the whites of the eyes called **jaundice**. Pain is a major consequence of red blood cells sickling within the blood vessels. When pain is severe, it is called a **sickle cell pain crisis**. The contents of the sickle cell released during its breakage can damage the blood vessel wall, resulting in **vasculopathy** (disorder of the blood vessel wall that further restricts blood circulation to tissues or organs). The December 10, 2008, issue of *JAMA* includes an article about a person with sickle cell vasculopathy.



SIGNS AND SYMPTOMS

- Pain in the affected area of the body
- Fever
- **Acute chest syndrome**—pain in the chest associated with cough, shortness of breath, and fever resembling pneumonia
- Jaundice
- Lower leg ulcers that are often slow to heal
- **Stroke** when a blood vessel in the brain is affected, resulting in lost or reduced function in the affected part of the body
- **Priapism**—painful erection that can last more than 4 hours when the blood vessels in the penis are affected
- **Pulmonary hypertension**—elevated blood pressure in the artery that carries the blood from the heart to the lungs

TREATMENT

- Medication to relieve pain
- Intravenous fluid hydration
- Oxygen therapy
- Wound care for ulcers
- Blood transfusions
- Hydroxyurea—a drug used to decrease the frequency of pain crises and need for blood transfusions
- Medications for pulmonary hypertension

PREVENTION

Individuals with sickle cell disease should

- Drink plenty of fluids
- Avoid extreme exertion
- Avoid extremes of temperature
- Avoid high altitudes
- Get appropriate vaccinations as recommended by your physician, including an annual influenza vaccine and the pneumococcal vaccine
- Discuss with your doctor whether hydroxyurea treatment is appropriate for prevention of sickle cell complications

FOR MORE INFORMATION

- Centers for Disease Control and Prevention
  www.cdc.gov/features/sicklecell
- Sickle Cell Disease Association of America
  www.sicklecelldisease.org

INFORM YOURSELF

To find this and other JAMA Patient Pages, go to the Patient Page link on *JAMA*'s Web site at www.jama.com. Many are available in English, Spanish, and French. A Patient Page on facts about sickle cell disease was published in the May 12, 1999, issue; one on pulmonary hypertension in the January 23, 2008, issue; and one on blood transfusions in the October 6, 2004, issue.

Source: Centers for Disease Control and Prevention

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