

**Genitalia:** Priapism (painful penile erection)

### Sequestration Crisis

Pooling of large amounts of blood

Hepatomegaly

Splenomegaly

Circulatory collapse

### Effects of Chronic Vasoocclusive Phenomena

**Heart:** Cardiomegaly, systolic murmurs

**Lungs:** Altered pulmonary function, susceptibility to infections, pulmonary insufficiency

**Kidneys:** Inability to concentrate urine, enuresis, progressive renal failure

**Liver:** Hepatomegaly, cirrhosis, intrahepatic cholestasis

**Spleen:** Splenomegaly, susceptibility to infection, functional reduction in splenic activity progressing to autosplenectomy

**Eyes:** Intraocular abnormalities with visual disturbances; sometimes progressive retinal detachment and blindness

**Extremities:** Avascular necrosis of hip or shoulder; skeletal deformities, especially lordosis and kyphosis; chronic leg ulcers; susceptibility to osteomyelitis

**Central nervous system (CNS):** Hemiparesis, seizures

The clinical manifestations of SCA vary greatly in severity and frequency. The most acute symptoms of the disease occur during periods of exacerbation called **crises**. There are several types of