

episodic crises, including vasoocclusive, acute splenic sequestration, aplastic, hyperhemolytic, cerebrovascular accident (CVA), chest syndrome, and infection. The crises may occur individually or concomitantly with one or more other crises. The **vasoocclusive crisis (VOC)**, preferably called a “painful episode,” is characterized by ischemia causing mild to severe pain that may last from minutes to days or longer. **Sequestration crisis** is a pooling of a large amount of blood usually in the spleen and infrequently in the liver that causes a decreased blood volume and ultimately shock. **Aplastic crisis** is diminished RBC production, usually triggered by viral infection that may result in profound anemia. **Hyperhemolytic crisis** is an accelerated rate of RBC destruction characterized by anemia, jaundice, and reticulocytosis.

Another serious complication is **acute chest syndrome (ACS)**, which is clinically similar to pneumonia. It is the presence of a new pulmonary infiltrate and may be associated with chest pain, fever, cough, tachypnea, wheezing, and hypoxia. A **cerebrovascular accident (CVA, stroke)** is a sudden and severe complication, often with no related illnesses. Sickled cells block the major blood vessels in the brain, resulting in cerebral infarction, which causes variable degrees of neurologic impairment. The current treatment for SCD children who have experienced a stroke is chronic transfusion therapy. Repeat CVAs causing progressively greater brain damage occur in approximately 70% of untreated children who have experienced one stroke ([Heeney and Dover, 2009](#); [Wang and Dwan, 2013](#)).

## Diagnostic Evaluation

Universal screening of newborns for SCD has become standard in all 50 United States and territories ([McCavit, 2012](#); [McGann, Nero, and Ware, 2013](#); [Meier and Miller, 2012](#)). However, global newborn screening varies by country and is not a common practice in most countries where SCD is a public health concern ([Aygun and Odame, 2012](#); [Huttle, Maestre, Lantigua, et al, 2015](#)). The screening provides early identification of these children before complications develop. At birth, infants have up to 80% of HbF, which does not carry the defect. Because levels of HbS are low at birth, Hgb electrophoresis or other tests that measure Hgb concentrations are indicated. Early diagnosis (before 3 months of age) enables