initiation of appropriate interventions to minimize complications. The family is taught to administer prophylactic antibiotics and identify early signs of infection to seek medical therapy as soon as possible.

Although SCD is usually reported during the prenatal or neonatal periods, it may not be recognized until the toddler and preschool period during a crisis precipitated by an acute respiratory tract or GI infection. However, early diagnosis (before the age of 2 months) facilitates initiation of appropriate interventions to minimize complications. There are several specific tests that detect the presence of the abnormal Hgb in the heterozygous or the homozygous form of SCD. For screening purposes, the sickle-turbidity test (Sickledex) is used because it can be performed on blood from a finger or heel stick and yields accurate results in 3 minutes. If the test result is positive, however, Hgb electrophoresis is necessary to distinguish between children with the trait and those with the disease. **Hemoglobin electrophoresis** referred to as "fingerprinting" of the protein is a specially prepared blood test that separates various hemoglobins by high-voltage. The blood test is accurate, rapid, and specific for detecting the homozygous and heterozygous forms of the disease, as well as the percentages of the various types of Hgb. The hemoglobin electrophoresis is used as the initial screening test increasingly in centers within the United States.

Therapeutic Management

The aims of therapy are to prevent the sickling phenomena, which are responsible for the pathologic sequelae, and treat the medical emergencies of sickle cell crisis. The successful achievement of the aims depends on prompt nursing interventions, medical therapies, patient and family preventive measures, and use of innovative treatments.

Medical management of a crisis is usually directed toward supportive, symptomatic and specific treatments. The main objectives are to provide (1) rest to minimize energy expenditure and to improve oxygen utilization; (2) hydration through oral and IV therapy; (3) electrolyte replacement because hypoxia results in metabolic acidosis, which also promotes sickling; (4) analgesia for the severe pain from vasoocclusion; (5) blood replacement to treat