although the basic defect in Hgb synthesis remains unaffected. A major postsplenectomy complication is severe and overwhelming infection. Therefore, these children are often on prophylactic antibiotics with close medical supervision for many years and should receive the pneumococcal and meningococcal vaccines in addition to the regularly scheduled immunizations (see Immunizations, Chapter 7).

Nursing Tip

Ensure that the family and patient understand the need to notify the health professional of all fevers of 38.5° C (101.3° F) or greater because of the risk of sepsis in a child with asplenia.

Prognosis

Most children treated with blood transfusion and early chelation therapy survive well into adulthood. The most common causes of death are heart disease, postsplenectomy sepsis, and multiple-organ failure secondary to hemochromatosis (Cunningham, Sankaran, Nathan, et al, 2009; Yaish, 2015). A curative treatment for some children is HSCT. Children younger than 16 years old who undergo allogeneic HSCT have a high rate of complication-free survival; approximately 80% to 97% of these children are cured (Isgro, Gaziev, Sodani, et al, 2010; Lucarelli, Isgro, Sodani, et al, 2012).

Nursing Care Management

The objectives of nursing care are to (1) promote compliance with transfusion and chelation therapy, (2) assist the child in coping with the anxiety-provoking treatments and the effects of the illness, (3) foster the child's and family's adjustment to a chronic illness, and (4) observe for complications of multiple blood transfusions. Basic to each of these goals is explaining to parents and older children the defect responsible for the disorder, its effect on RBCs, and the potential effects of untreated iron overload (e.g., delayed growth and maturation and heart disease). Because this condition is prevalent among families of Mediterranean descent, the nurse also inquires about the family's previous knowledge about thalassemia. All families with a child with thalassemia should be tested for the