

manifestations of SCD that may appear in the first 2 years of life (dactylitis, severe anemia, leukocytosis) can be predictors of disease severity (Miller, Sleeper, Pegelow, et al, 2000). The nurse should care for the family as for any family with a child who has a chronic and life-threatening illness and give consideration to the siblings' reactions, the stress on the marital relationship, and the childrearing attitudes displayed toward the child (see Chapter 17). Several resources are available to families with a sickling disorder.\*

The nurse advises parents to inform all treating personnel of the child's condition. The use of medical identification, such as a bracelet, is another way of ensuring awareness of the disease.

If family members have the SCD trait or SCA, genetic counseling is necessary. A primary consideration in genetic counseling is informing parents of the 25% chance with each pregnancy of having a child with the disease when both parents carry the trait.

## Beta-Thalassemia (Cooley Anemia)

Worldwide, thalassemia is a common genetic disorder, affecting as many as 15 million people (Yaish, 2015). The term **thalassemia**, which is derived from the Greek word *thalassa*, meaning “sea,” is applied to a variety of inherited blood disorders characterized by deficiencies in the rate of production of specific globin chains in Hgb. The name appropriately refers to people living near the Mediterranean Sea, namely Italians, Greeks, Syrians, Asians, Africans, and their descendants. Evidence suggests that the high incidence of the disorders among these groups is a result of the selective advantage the trait in protecting against malaria, as is postulated in SCD. The disorder has a wide geographic distribution, probably as a result of genetic migration through intermarriage or possibly as a result of spontaneous mutation.

Beta-thalassemia is the most common of the thalassemias and occurs in four forms:

- Two heterozygous forms, **thalassemia minor**, an asymptomatic silent carrier, and **thalassemia trait**, which produces a mild microcytic anemia
- **Thalassemia intermedia**, which may involve either homozygous or heterozygous abnormalities and is manifested as splenomegaly and moderate to severe anemia