Systemic Lupus Erythematosus

Systemic lupus erythematosus (SLE) is a severe chronic autoimmune disease that results in inflammation and multi-organ system damage. Other forms of lupus include discoid lupus, which is limited to the skin, and neonatal lupus, which occurs when maternal autoantibodies cause a transient lupus-like syndrome in a newborn with the potential serious complication of heart block. The remaining discussion focuses on SLE.

The Lupus Foundation of America (2015) and National Kidney Foundation (2015) estimates that 1.5 million individuals have lupus, and 10% to 15% of these adults were diagnosed with SLE as children or adolescents. SLE in children tends to be more severe at onset and has more aggressive clinical course than adult-onset type (Mina and Brunner, 2013).

SLE is more common in girls, with an approximate 4 : 3 female-to-male predominance before 10 years old and 4 : 1 in the second decade, indicating a potential hormonal trigger with maturation. There is a familial tendency, although many newly diagnosed patients are unaware of other affected family members. SLE has been reported in all cultures, but within the United States, there has been a disproportionately higher incidence in African-American, Asian, and Hispanic children.

The cause of SLE is not known. It appears to result from a complex interaction of genetics with an unidentified trigger that activates the disease. Suspected triggers include exposure to ultraviolet (UV) light, estrogen, pregnancy, infections, and drugs. Genetic predisposition to SLE is evidenced in an increased concordance rate in twins (tenfold), increased incidence within family members (10% to 16%), and increased frequency of certain gene alleles in population-based studies.

Clinical Manifestations and Diagnostic Evaluation

The child with SLE may have any clinical manifestation with mild to life-threatening severity (Box 29-10). The diagnosis is established when four of the 11 diagnostic criteria are met (Box 29-11). Kidney involvement heralds progressive disease and the need for rigorous therapeutic management.