Table 9-1 International League of Associations for Rheumatology Classification of Juvenile Idiopathic Arthritis (JIA)

Subtype	% of JIA Cases	Sex	% of Patients With Uveitis
Oligoarticular	40–50	F > M	30
Polyarticular			
RF-negative subtype	20–25	F > M	15
RF-positive subtype	5	F > M	<1
Systemic	5–10	F = M	<1
Psoriatic	5–10	F > M	10
Enthesitis-related	5–10	M > F	7

The last 2 subtypes, *psoriatic arthritis* and *enthesitis-related arthritis*, were added to help in categorizing patients who did not fit into more common entities. Uveitis can occur in both of these subtypes.

Systemic Lupus Erythematosus

Systemic lupus erythematosus (SLE) is a heterogeneous autoimmune disease of undetermined cause that presents with a wide range of clinical manifestations and can involve any organ. It is characterized by remissions and relapses, from mild to severe. The disease is estimated to affect 1.5 million people in the United States. Women are affected far more frequently than men, and the median age of onset is between 35 and 50 years. Individuals of African, Asian, and Native American heritage are more likely than White individuals to develop SLE. It is associated with B-cell hyperactivity, hypergammaglobulinemia, and a plethora of autoantibodies. These include antinuclear antibodies (ANAs) as well as antibodies to DNA and cytoplasmic components. SLE has classically been considered an immune complex disease that leads to an inflammatory response and tissue damage.

Signs and Symptoms

Patients may present with single-organ involvement, such as nephritis, or with a multisystem disease. The characteristic cutaneous manifestation of SLE is the *butterfly rash* (or *malar rash*) across the nose and cheeks, which appears in 70%–80% of patients (Fig 9-4). Other cutaneous manifestations include discoid lesions, which often lead to scarring, and alopecia. Mucosal lesions, usually painless oral or nasal ulcers, are present in up to 40% of patients. Acute or chronic photosensitivity occurs in many patients.

Approximately 90% of SLE patients experience articular disease, as either a polyarthralgia or a nondeforming migratory polyarthritis. Constitutional symptoms, such as fatigue, fever, myalgia, and weight loss, are common. Renal disease affects approximately 50% of patients; it can present with a range of manifestations from hematuria, proteinuria, and nephrotic syndrome to fulminant glomerulonephritis and renal failure.

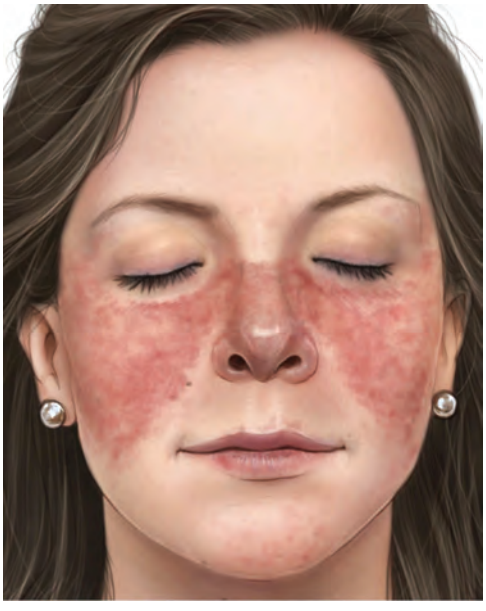

Figure 9-4 Malar "butterfly" rash in a patient with systemic lupus erythematosus. *(Used with permission from Mayo Foundation for Medical Education and Research. All rights reserved.)*

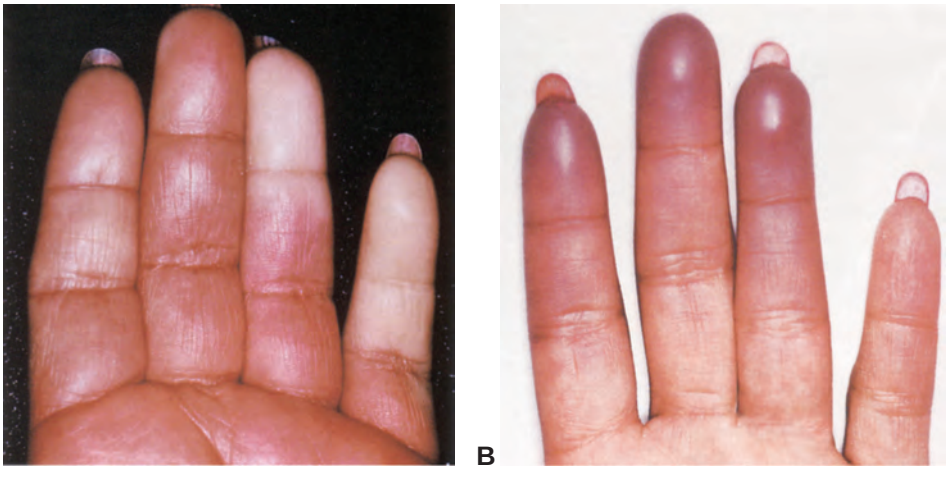

Figure 9-5 Raynaud phenomenon. **A,** Sharply demarcated pallor resulting from the closure of digital arteries. **B,** Digital cyanosis of the fingertips in a patient with primary Raynaud phenomenon. *(Reproduced with permission from Wigley FM. Clinical practice. Raynaud's phenomenon.* N Engl J Med. *2002;347(13):1001. Copyright ©2002 Massachusetts Medical Society.)*

Raynaud phenomenon occurs in up to 50% of patients with SLE (Fig 9-5). Anemia of chronic disease is common, along with reduction in leukocytes and platelets. The rate of coronary artery disease is significantly greater in SLE patients than in unaffected individuals. Other, less common cardiac manifestations include pericarditis, myocarditis, and Libman-Sacks endocarditis. Valvular disease has been reported in more than 50%