TABLE 79-3 SYSTEMIC LUPUS INTERNATIONAL COLLABORATING CLINICS (SLICC) CLASSIFICATION CRITERIA OF SYSTEMIC LUPUS ERYTHEMATOSUS

CLINICAL CRITERIA*	EXAMPLES
1. Acute cutaneous lupus	Bullous lupus Lupus malar rash (not malar discoid) Maculopapular lupus rash Photosensitive lupus rash (in the absence of dermatomyositis) Subacute cutaneous lupus Toxic epidermal necrolysis variant of SLE
2. Chronic cutaneous lupus	Classic discoid rash Localized (above the neck) Generalized (above and below the neck) Chilblains lupus Discoid lupus/lichen planus overlap Hypertrophic (verrucous) lupus Lupus erythematosus tumidus Lupus panniculitis (profundus) Mucosal lupus
3. Oral ulcers	Palate, buccal, tongue, *or* nasal ulcers (in the absence of other causes: vasculitis, Behçet's disease, infection, inflammatory bowel disease, reactive arthritis, and acidic foods)
4. Nonscarring alopecia	Diffuse thinning *or* hair fragility with visible broken hairs (in the absence of other causes: alopecia areata, drugs, iron deficiency, and androgenic alopecia)
5. Synovitis (≥2 joints)	Characterized by swelling or effusion *or* tenderness with ≥30 minutes of morning stiffness
6. Serositis	Typical pleurisy for >1 day *or* pleural effusions *or* pleural rub Typical pericardial pain for >1 day *or* pericardial effusion *or* pericardial rub *or* pericarditis by ECG (in the absence of other causes: infection, uremia, and Dressler's pericarditis)
7. Renal	Urine protein/creatinine (*or* 24-hr protein) ≥500 mg of protein/24 hr *or* red blood cell casts
8. Neurologic	Acute confusional state (in the absence of other causes: toxic-metabolic, uremia, and drugs) Mononeuritis multiplex (in the absence of other known causes: primary vasculitis) Myelitis Peripheral or cranial neuropathy (in the absence of other known causes: primary vasculitis, infection, and diabetes mellitus) Psychosis Seizure
9. Hemolytic anemia	
10. Leukopenia	<4000 cells/mm^3 detected at least once (in the absence of other known causes: Felty's syndrome, drugs, and portal hypertension)
or Lymphopenia	<1000 cells/mm^3 detected at least once (in the absence of other known causes: corticosteroids, drugs, and infection)
11. Thrombocytopenia	<100,000 cells/mm^3 detected at least once (in the absence of other known causes: drugs, portal hypertension, and thrombotic thrombocytopenic purpura)
IMMUNOLOGIC CRITERIA	
1. ANAs	Above laboratory reference range
2. Anti-double stranded DNA	Above laboratory reference range, except ELISA: two times greater than laboratory reference range
3. Anti-Smith	
4. Antiphospholipid	Any of the following: lupus anticoagulant, false-positive RPR, medium- or high-titer anticardiolipin (IgA, IgG, or IgM), or anti-β_2 glycoprotein I (IgA, IgG, or IgM)
5. Low complement	Low C3 Low C4 Low CH50
6. Direct Coombs test	In the absence of hemolytic anemia

Modified from Petri M, Orbai AM, Alarcón GS, et al: Derivation and validation of Systemic Lupus International Collaborating Clinics classification criteria for systemic lupus erythematosus, Arthritis Rheum 64:2677–2686, 2012.

ANAs, Antinuclear antibodies; ECG, electrocardiogram; ELISA, enzyme-linked immunosorbent assay; Ig, immunoglobulin; RPR, rapid plasma reagin; SLE, systemic lupus erythematosus.

*Criteria are cumulative. A patient is classified as having SLE using lupus nephritis as a stand-alone criterion (in the setting of ANAs or anti-dsDNA antibodies) *or* four criteria (with at least one of the clinical criteria and one of the immunologic criteria).

TABLE 79-4 PREVALENCE OF AUTOANTIBODIES IN SYSTEMIC LUPUS ERYTHEMATOSUS

TARGET AUTOANTIGEN	POSITIVE (%)
Nuclear antigens	>95
Double-stranded DNA	30-60
Smith	10-44
Ribonucleoprotein (U1-RNP)	25-40
SSA/Ro	30-40
SSB/La	38
Phospholipids	16-60
Ribosomal P	5-10
Histone	21-90

Data from Wallace D, Hahn BH: Other clinical laboratory tests in SLE. In Dubois' lupus erythematosus and related syndromes, ed 8, Philadelphia, 2013, Saunders, pp 526–531.

used. These patients typically are early in their disease course and eventually develop a specific autoimmune disease.

TREATMENT

No cure for SLE has been identified, and treatment is aimed at educating patients, reducing inflammation, suppressing the immune system, and closely monitoring patients to identify disease manifestations as early as possible. Treatment with glucocorticoids and immunosuppressive agents has reduced the morbidity and mortality of patients with SLE, although the treatments themselves are associated with extensive toxicity. Physicians caring for SLE patients must carefully weigh the benefits of therapy against the known risks of treatment.