

## 7 Table. 1997 Revised Criteria for Diagnosis of SLE\*

### I. Criterion Definition(1)

A. Definite SLE - 4 or more criteria, in general including a positive ANA  
Probable SLE - 3 criteria

1. Malar Rash Fixed erythema, flat or raised, over the malar eminences, tending to spare the nasolabial folds
2. Discoid rash Erythematous raised patches with adherent keratotic scaling and follicular plugging; atrophic scarring may occur in older lesions
3. Photosensitivity Skin rash as a result of unusual reaction to sunlight, by patient history or physician observation
4. Oral ulcers Oral or nasopharyngeal ulceration, usually painless, observed by physician
5. Nonerosive Arthritis Involving 2 or more peripheral joints, characterized by tenderness, swelling, or effusion
6. Pleuritis or Pericarditis
  - a. Pleuritis--convincing history of pleuritic pain or rubbing heard by a physician or evidence of pleural effusion
  - OR
  - b. Pericarditis--documented by electrocardiogram or rub or evidence of pericardial effusion
7. Renal Disorder
  - a. Persistent proteinuria > 0.5 grams per day or > than 3+ if quantitation not performed
  - OR
  - b. Cellular casts--may be red cell, hemoglobin, granular, tubular, or mixed
8. Neurologic Disorder
  - a. Seizures--in the absence of offending drugs or known metabolic derangements; e.g., uremia, ketoacidosis, or electrolyte imbalance
  - b. Psychosis--in the absence of offending drugs or known metabolic derangements, e.g., uremia, ketoacidosis, or electrolyte imbalance
9. Hematologic Disorder
  - a. Hemolytic anemia--with reticulocytosis

OR

b. Leukopenia--< 4,000/mm<sup>3</sup> on ≥ 2 occasions

OR

c. Lymphopenia--< 1,500/ mm<sup>3</sup> on ≥ 2 occasions

OR

d. Thrombocytopenia--<100,000/ mm<sup>3</sup> in the absence of offending drugs

#### 10. Immunologic Disorder

a. Anti-DNA: antibody to native DNA in abnormal titer

OR

b. Anti-Sm: presence of antibody to Sm nuclear antigen

OR

c. Positive finding of antiphospholipid antibodies on:

1. an abnormal serum level of IgG or IgM anticardiolipin antibodies,

2. a positive test result for lupus anticoagulant using a standard method, or

3. a false-positive test result for at least 6 months confirmed by Treponema pallidum immobilization or fluorescent treponemal antibody absorption test

11. Positive Antinuclear Antibody An abnormal titer of antinuclear antibody by immunofluorescence or an equivalent assay at any point in time and in the absence of drugs

### II. Further Points about diagnosis

A. So, when you get a positive ANA...order an ANA profile - which should include *anti-double stranded DNA, anti-Smith, anti-SSA and anti-SSB, and anti-RNP.*

B. Anti-dsDNA and anti-Sm are virtually 100% (96-100) specific for SLE (they are ONLY positive in pts with SLE). However, their sensitivity is not as good - 52% for Crithidida anti-dsDNA and 73% for Farr anti-dsDNA, and 18-31% for anti-Smith.

C. If ANA is *negative* and clinical signs strongly suggest SLE, check for *anti-SSA(Ro)* antibodies. If this is positive, the patient probably has "*ANA-negative*" SLE (rare). As many as 62% of patients with "*ANA-negative*" SLE have anti-SSA antibodies.

Complement levels can also be helpful diagnostically - total serum hemolytic complement (**CH50**) and individual complement components (**C3 and C4**) may be low in patients with active SLE due to the presence of immune complexes; low sensitivity (40%) but high specificity (90%).

1. Tan, E., A. Cohen, J. Fries, A. Masi, D. Mcshane, N. Rothfield, J. Schaller, N. Talal, and R. Winchester. 1982. The 1982 revised criteria for the classification of systemic lupus erythematosus. *Arthritis Rheum* 25:1271-1277.