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Appendix

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Appendix to: Golder V, Hoi A. What's new in systemic lupus erythematosus. *Med J Aust* 2017; 206: 215-220. doi: 10.5694/mja16.01229.

Appendix: Detailed classification criteria for systemic lupus erythematosus

Systemic Lupus International Collaborating Clinics (SLICC) Criteria[1]	American College of Rheumatology (ACR) Criteria[2]
Clinical criteria	
1. Acute cutaneous lupus o Lupus malar rash (do not count if malar discoid) o Bullous lupus o Toxic epidermal necrolysis variant of SLE o Maculopapular lupus rash o Photosensitive lupus rash in the absence of dermatomyositis o Subacute cutaneous lupus (nonindurated psoriasiform and/or annular polycyclic lesions that resolve without scarring, although occasionally with post inflammatory dyspigmentation or telangiectasias)	**1. Malar rash** o Fixed erythema, flat or raised, over the malar eminences, tending to spare the nasolabial folds
2. Chronic cutaneous lupus o Classic discoid rash o Localized (above the neck) o Generalized (above and below the neck) o Hypertrophic (verrucous) lupus o Lupus panniculitis (profundus) o Mucosal lupus o Lupus erythematosus tumidus o Chillblains lupus o Discoid lupus/lichen planus overlap	**2. Discoid rash** o Erythematous raised patches with adherent keratotic scaling and follicular plugging; atrophic scarring may occur in older lesions
3. Oral ulcers o Palate, buccal, tongue or nasal ulcers in the absence of other causes, such as vasculitis, Behcet's disease, infection (herpes virus), inflammatory bowel disease, reactive arthritis, and acidic foods	**3. Oral ulcers** o Oral or nasopharyngeal ulceration, usually painless, observed by physician
4. Nonscarring alopecia o Diffuse thinning or hair fragility with visible broken hairs, in the absence of other causes such as alopecia areata, drugs, iron deficiency, and androgenic alopecia	**4. Photosensitivity** o Skin rash as a result of unusual reaction to sunlight, by patient history or physician observation
5. Synovitis o Involving 2 or more joints, characterized by swelling or effusion o OR tenderness in 2 or more joints and at least 30 minutes of morning stiffness	**5. Non erosive Arthritis** o Involving 2 or more peripheral joints, characterized by tenderness, swelling or effusion
6. Serositis o Typical pleurisy for more than 1 day OR pleural effusions OR pleural rub o Typical pericardial pain (pain with recumbency improved by sitting forward) for more than 1 day, OR pericardial effusion, OR pericardial rub, OR pericarditis by	**6. Serositis** o Pleuritis--convincing history of pleuritic pain or rubbing heard by a physician or evidence of pleural effusion, OR o Pericarditis--documented by electrocardiogram or rub or evidence of pericardial effusion

electrocardiography; in the absence of other causes, such as infection, uremia, and Dressler's pericarditis	
7. Renal o Urine protein–to-creatinine ratio (or 24-hour urine protein) representing 500 mg protein/24 hours o OR red blood cell casts	**7. Renal disorder** o Persistent proteinuria > 0.5 grams per day or > than 3+ if quantitation not performed, OR o Cellular casts--may be red cell, haemoglobin, granular, tubular, or mixed
8. Neurologic o Seizures o Psychosis o Mononeuritis multiplex in the absence of other known causes such as primary vasculitis o Myelitis o Peripheral or cranial neuropathy in the absence of other known causes such as primary vasculitis, infection, and diabetes mellitus o Acute confusional state in the absence of other causes, including toxic/metabolic, uremia, drugs	**8. Neurologic disorder** o Seizures--in the absence of offending drugs or known metabolic derangements; e.g., uremia, ketoacidosis, or electrolyte imbalance o Psychosis--in the absence of offending drugs or known metabolic derangements, e.g., uremia, ketoacidosis, or electrolyte imbalance
9. Haemolytic anaemia	**9. Haematologic disorder** o Haemolytic anaemia--with reticulocytosis. OR o Leukopaenia--< 4,000/mm3 on ≥ 2 occasions, OR o Lymphopaenia--< 1,500/ mm3 on ≥ 2 occasions, OR o Thrombocytopaenia--<100,000/ mm3 in the absence of offending drugs
10. Leukopaenia o Leukopaenia (<4,000/mm3 at least once) in the absence of other known causes such as Felty's syndrome, drugs, and portal hypertension. OR o Lymphopaenia (<1,000/mm3 at least once)	
11.Thrombocytopaenia o (<100,000/mm3), At least once in the absence of other known causes, ie drugs, portal hypertension, and TTP.	
Laboratory criteria	
1. Antinuclear antibody o Level above laboratory reference range	**1. Antinuclear antibody** o An abnormal titre of antinuclear antibody by immunofluorescence or an equivalent assay at any point in time and in the absence of drugs known to be associated with "drug-induced lupus"
2. Anti-double stranded DNA antibody o Level above laboratory reference range (or >2-fold the reference range if tested by ELISA)	**2. Immunological disorders** o Anti-DNA: antibody to native DNA in abnormal titre OR o Anti-Sm: presence of antibody to Sm nuclear antigen OR o Positive finding of antiphospholipid antibodies on: o an abnormal serum level of IgG or IgM anticardiolipin antibodies, o a positive test result for lupus anticoagulant using a standard
3. Anti-Sm antibody o Presence of antibody to Sm nuclear antigen	
4. Antiphospholipid antibody positivity As determined by any of the following: o Positive test result for lupus anticoagulant o False-positive test result for rapid plasma	

reagin ○ Medium- or high-titer anticardiolipin antibody level (IgA, IgG, or IgM) ○ Positive test result for anti–β2 -glycoprotein I (IgA, IgG, or IgM)	method, or ○ a false-positive test result for at least 6 months confirmed by Treponema pallidum immobilization or fluorescent treponemal antibody absorption test
5. Low complement ○ Low C3 OR ○ Low C4 OR ○ Low CH50	
6. Direct Coombs' test ○ Positive test in the absence of haemolytic anaemia	
Requirement for diagnosis	
Meets 4 SLICC criteria (with at least one criterion being clinical and at least one criterion being immunological) **Or, alternatively in the presence of biopsy proven lupus nephritis and at least one immunological criterion**	**Meets 4 of 11 ACR criteria**

References

1 Petri M, Orbai AM, Alarcon GS, et al. Derivation and validation of the Systemic Lupus International Collaborating Clinics classification criteria for systemic lupus erythematosus. *Arthritis Rheum* 2012; 64: 2677-2686.

2 Hochberg MC. Updating the American College of Rheumatology revised criteria for the classification of systemic lupus erythematosus. *Arthritis Rheum* 1997; 40: 1725.