2019 European Alliance of Associations for Rheumatology (EULAR)/American College of Rheumatology (ACR) classification criteria for systemic lupus erythematosus

The entry criterion is necessary to classify SLE.

Entry criterion:

- ANA at a titer of ≥1:80 on HEp-2 cells or an equivalent positive test (ever).*

At least 1 clinical criterion required to classify SLE. Additional additive (clinical or immunology) criteria are counted toward the total score.

Additive criteria:

- Do not count a criterion if there is a more likely explanation than SLE.
- Occurrence of a criterion on ≥1 occasion is sufficient.
- Criteria need not occur simultaneously.
- Within each domain (eg, mucocutaneous, complement proteins), only the highest-weighted criterion is counted toward the total score if more than 1 is present.¶

Clinical domains and criteria	Weight
Constitutional	
Fever	2
Hematologic	
Leukopenia	3
Thrombocytopenia	4
Autoimmune hemolysis	4
Neuropsychiatric	
Delirium	2
Psychosis	3
Seizure	5
Mucocutaneous	
Nonscarring alopecia	2

Oral ulcers	2
Subacute cutaneous or discoid lupus	4
Acute cutaneous lupus	6
Serosal	
Pleural or pericardial effusion	5
Acute pericarditis	6
Musculoskeletal	
Joint involvement	6
Renal	
Proteinuria >0.5 g per 24 hours	4
Renal biopsy Class II or V lupus nephritis	8
Renal biopsy Class III or IV lupus nephritis	10
Immunology domains and criteria	**Weight**
Antiphospholipid antibodies	
Anti-cardiolipin antibodies **or** anti-beta-2GP1 antibodies **or** lupus anticoagulant	2
Complement proteins	
Low C3 **or** low C4	3
Low C3 **and** low C4	4
SLE-specific antibodies	
Anti-dsDNA antibody$^{\Delta}$ **or** anti-Smith antibody	6
A total score of ≥10 and ≥1 clinical criterion are required to classify SLE.	
Total score	

SLE: systemic lupus erythematosus; ANA: antinuclear antibody; HEp-2: human epithelial type 2; anti-beta-2GP1: anti-beta-2 glycoprotein 1; anti-dsDNA: anti-double-stranded DNA.

* If ANA is absent, do **not** classify as SLE.

¶ Additional criteria within the same domain will not be counted.

Δ In an assay with 90% specificity against relevant disease controls.

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Definitions of SLE classification criteria

Criteria	Definition
Antinuclear antibodies (ANA)	ANA at a titer of ≥1:80 on HEp-2 cells or an equivalent positive test at least once. Testing by immunofluorescence on HEp-2 cells or a solid phase ANA screening immunoassay with at least equivalent performance is highly recommended.
Fever	Temperature >38.3°C.
Leucopenia	White blood cell count $<4.0 \times 10^9$/L.
Thrombocytopenia	Platelet count $<100 \times 10^9$/L.
Autoimmune hemolysis	Evidence of hemolysis, such as reticulocytosis, low haptoglobin, elevated indirect bilirubin, elevated lactate dehydrogenase (LDH) AND positive Coombs (direct antiglobulin) test.
Delirium	Characterized by (1) change in consciousness or level of arousal with reduced ability to focus, (2) symptom development over hours to <2 days, (3) symptom fluctuation throughout the day, (4) either (4a) acute/subacute change in cognition (eg, memory deficit or disorientation), or (4b) change in behavior, mood, or affect (eg, restlessness, reversal of sleep/wake cycle).
Psychosis	Characterized by (1) delusions and/or hallucinations without insight and (2) absence of delirium.
Seizure	Primary generalized seizure or partial/focal seizure.
Nonscarring alopecia	Nonscarring alopecia observed by a clinician.*
Oral ulcers	Oral ulcers observed by a clinician.*
Subacute cutaneous or discoid lupus	Subacute cutaneous lupus erythematosus observed by a clinician*: Annular or papulosquamous (psoriasiform) cutaneous eruption, usually photodistributed. Discoid lupus erythematosus observed by a clinician*: Erythematous-violaceous cutaneous lesions with secondary changes of atrophic scarring, dyspigmentation, often follicular hyperkeratosis/hematological (scalp), leading to scarring alopecia on the scalp. If skin biopsy is performed, typical changes must be present. Subacute cutaneous lupus: interface vacuolar dermatitis consisting of a perivascular

	lymphohistiocytic infiltrate, often with dermal mucin noted. Discoid lupus: interface vacuolar dermatitis consisting of a perivascular and/or periappendageal lymphohistiocytic infiltrate. In the scalp, follicular keratin plugs may be seen. In longstanding lesions, mucin deposition and basement membrane thickening may be noted.
Acute cutaneous lupus	Malar rash or generalized maculopapular rash observed by a clinician. If skin biopsy is performed, typical changes must be present: interface vacuolar dermatitis consisting of a perivascular lymphohistiocytic infiltrate, often with dermal mucin noted. Perivascular neutrophilic infiltrate may be present early in the course.
Pleural or pericardial effusion	Imaging evidence (such as ultrasound, radiograph, CT scan, MRI) of pleural or pericardial effusion, or both.
Acute pericarditis	≥2 of (1) pericardial chest pain (typically sharp, worse with inspiration, improved by leaning forward), (2) pericardial rub, (3) electrocardiogram (EKG) with new widespread ST-elevation or PR depression, (4) new or worsened pericardial effusion on imaging (such as ultrasound, radiograph, CT scan, MRI).
Joint involvement	EITHER (1) synovitis involving 2 or more joints characterized by swelling or effusion OR (2) tenderness in 2 or more joints and at least 30 minutes of morning stiffness.
Proteinuria >0.5 g/24 hours	Proteinuria >0.5 g/24 hours by 24 hours urine or equivalent spot urine protein-to-creatinine ratio.
Class II or V lupus nephritis on renal biopsy according to ISN/RPS 2003 classification	Class II: mesangial proliferative lupus nephritis: purely mesangial hypercellularity of any degree or mesangial matrix expansion by light microscopy, with mesangial immune deposit. A few isolated subepithelial or subendothelial deposits may be visible by immune-fluorescence or electron microscopy, but not by light microscopy. Class V: membranous lupus nephritis: global or segmental subepithelial immune deposits or their morphologic sequelae by light microscopy and by immunofluorescence or electron microscopy, with or without mesangial alterations.
Class III or IV lupus nephritis on renal biopsy according to International Society of Nephrology/ Renal Pathology Society	Class III: focal lupus nephritis: active or inactive focal, segmental or global endocapillary or extracapillary glomerulonephritis involving <50% of all glomeruli, typically with focal subendothelial immune deposits, with or without mesangial alterations. Class IV: diffuse lupus nephritis: active or inactive diffuse, segmental or global endocapillary or extracapillary glomerulonephritis involving ≥50% of

(ISN/RPS) 2003	all glomeruli, typically with diffuse subendothelial immune deposits, with or without mesangial alterations. This class includes cases with diffuse wire loop deposits but with little or no glomerular proliferation.
Positive antiphospholipid antibodies	Anticardiolipin antibodies (IgA, IgG, or IgM) at medium or high titer (>40 A phospholipids [APL], GPL or MPL units, or >the 99th percentile) or positive anti-beta-2GP1 antibodies (IgA, IgG, or IgM) or positive lupus anticoagulant.
Low C3 OR low C4	C3 OR C4 below the lower limit of normal.
Low C3 AND low C4	Both C3 AND C4 below their lower limits of normal.
Anti-dsDNA antibodies OR anti-Smith (Sm) antibodies	Anti-dsDNA antibodies in an immunoassay with demonstrated ≥90% specificity for SLE against relevant disease controls OR anti-Sm antibodies.

SLE: systemic lupus erythematosus; HEp-2: human epithelial type 2; CT: computed tomography; MRI: magnetic resonance imaging; GP: glycoprotein; dsDNA: double-stranded DNA.
* This may include physical examination or review of a photograph.

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