

**2019 European League Against Rheumatism/American College of Rheumatology Classification Criteria for Systemic Lupus Erythematosus**

**Supplementary Table 1.** Multiphase development of SLE classification criteria

<b>Phase 1b Delphi Exercise</b>	<b>Phase 1c Early cohort study</b>	<b>Phase 1d Patient survey (all with a prevalence of &gt;50%)</b>	<b>Phase 2b Consensus meeting using nominal group technique (recommended definition)</b>	<b>Phase 3a Criteria refining</b>	<b>Final criteria</b>
ANA on HEp2 cells with a pattern compatible with SLE, titer $\geq$ 1:160	ANA	ANA (95%)	ANA $\geq$ 1:80 (HEp2 IF)	ANA $\geq$ 1:80 (HEp2 IF)	ANA $\geq$ 1:80 HEp2 IF or an equivalent positive test
ANA positive $\geq$ 1:160 (any pattern)					
ANA positive by HEp 2 IF					
	fever***	fever (54%)	fever (to be defined)	fever $>$ 38.3°C	fever $>$ 38.3°C
	fatigue (neg)	fatigue (89%)			
	weight loss				
Raynaud's phenomenon	Raynaud's phenomenon (neg)***	Raynaud's phenomenon (52%)			
	dry eyes (neg)***				
	dry mouth (neg)***				
	dysphagia (neg)				
lymphopenia ( $<$ 1500/mm <sup>3</sup> on 2 or more occasions)					
lymphopenia ( $<$ 1000/mm <sup>3</sup> on 2 or more occasions)					
leukopenia ( $<$ 4000/mm <sup>3</sup> on 2 or more occasions)	leukopenia		leukopenia( $<$ 4000/mm <sup>3</sup> on 2 or more occasions)	leukopenia (to be tested)	leukopenia ( $<$ 4000/mm <sup>3</sup> once)
thrombocytopenia			thrombocytopenia	thrombocytopenia	thrombocytopenia

thrombocytopenia (severe)			(<100,000twice)	(<100,000 once)	(<100,000 once)
autoimmune hemolytic anemia	Coombs test		autoimmune hemolytic anemia	autoimmune hemolytic anemia	autoimmune hemolytic anemia
<b>Phase 1b Delphi Exercise</b>	<b>Phase 1c Early cohort study</b>	<b>Phase 1d Patient survey (all with a prevalence of &gt;50%)</b>	<b>Phase 2b Consensus meeting using nominal group technique (provisional definition)</b>	<b>Phase 3a Criteria refining</b>	<b>Final criteria</b>
CNS dysfunction (seizures, psychosis, chorea or acute confusional state)	neurological involvement (ACR)		CNS manifestations (seizures, psychosis, chorea, myelitis, optic neuritis, stroke or acute confusional state)	delirium	delirium
				psychosis	psychosis
				seizure	seizure
	CNS at least one			mononeuropathy	
			cranial neuropathy		
alopecia with associated scalp inflammation	alopecia***	alopecia (51%)	alopecia with associated scalp inflammation	non-scarring alopecia	non-scarring alopecia
oral mucosal lesions on the hard palate	oral ulcer***		oral mucosal lesions on the hard palate	oral ulcers	oral ulcers
photosensitivity	photosensitivity***	photosensitivity (79%)			
photosensitive rash					
acute, subacute, or chronic lupus rash		skin rash (71%)			
lupus profundus			included in chronic cutaneous lupus		
discoid rash			chronic cutaneous lupus	SCLE or discoid lupus	SCLE or discoid lupus
SCLE			included in ACLE		
malar rash	malar rash***		acute cutaneous lupus	malar rash or generalized maculopapular rash	malar rash or generalized maculopapular rash
rash with dermoepidermal interface changes and Ig and/or complement			rash with dermo-epidermal interface changes and/or Ig	included into ACLE, SCLE, discoid lupus	included into ACLE, SCLE, discoid lupus

deposition on IF			and/or complement deposition on IF		
Phase 1b Delphi Exercise	Phase 1c Early cohort study	Phase 1d Patient survey (all with a prevalence of >50%)	Phase 2b Consensus meeting using nominal group technique (provisional definition)	Phase 3a Criteria refining	Final criteria
serositis (clinical signs, or pleural or pericardial effusion by imaging)	serositis***		serositis (pleural, pericardial effusion, pleurisy, pericarditis, peritonitis)	pleural or pericardial effusion	pleural or pericardial effusion
pleural effusion				acute pericarditis	acute pericarditis
pleuritis					
pericardial effusion	pericarditis***				
		myalgia (76%)			
arthritis	inflammatory arthritis***	joint pain (87%)	arthritis (to be defined)	arthritis (to be tested)	joint involvement
	arthralgia (negative)				
	kidney involvement***				
	hypertension				
persistent proteinuria (>0.5g/day)			persistent proteinuria (≥0.5g/day)	proteinuria >0.5 g/24h	proteinuria >0.5 g/24h
	peripheral edema				
urine cellular casts					
urinary RBC casts (≥ 1 cast/hpf)					
dysmorphic urinary RBC or urinary RBC casts					
active urine sediment (without UTI)			active urine sediment (without UTI)		
lupus nephritis by renal biopsy with immune			lupus nephritis by renal biopsy with immune	lupus nephritis by ISN/RPS	ISN/RPS class III or IV nephritis

deposits			deposits		ISN/RPS class II or V nephritis
Phase 1b Delphi Exercise	Phase 1c Early cohort study	Phase 1d Patient survey (all with a prevalence of >50%)	Phase 2b Consensus meeting using nominal group technique (recommended definition)	Phase 3a Criteria refining	Final criteria
APS (clinical signs/history + antibodies)					
antiphospholipid antibodies (lupus anti-coagulant, anti-cardiolipin, anti-beta2GP1, or prolonged RVVT)	anti-Cardiolipin IgM***		anti-phospholipid antibodies (lupus anti-coagulant, anti-cardiolipin, anti-beta2GP1, or prolonged RVVT)	anti-cardiolipin IgG (>40 IgG phospholipid units) or anti-beta2GP1 IgG (>40 units) or lupus anticoagulant positive	Anti-Cardiolipin antibodies (IgA, IgG, or IgM) at medium or high titer or positive anti-beta2GP1 antibodies (IgA, IgG, or IgM) or positive lupus anticoagulant.
positive lupus anticoagulant panel	anti-beta2GP1				
low C3	hypocomplementemia***		low C3 and/or C4	low C3 OR low C4	low C3 OR low C4
low C3 and C4				low C3 AND C4	low C3 AND C4
anti-dsDNA antibody	anti-dsDNA***	anti-dsDNA (78%)	anti-dsDNA antibody	anti-dsDNA antibodies	anti-dsDNA antibodies
anti-Sm antibody	anti-Sm***		anti-Sm antibody	anti-Sm antibodies	anti-Sm antibodies
	anti-RNP***				
presence of multiple autoantibodies			presence of multiple autoantibodies (to be defined)		

\*\*\* significant in univariate analysis

ACLE acute cutaneous lupus erythematosus

ACR American College of Rheumatology

ANA Antinuclear antibodies

anti-beta2GP1 anti-beta 2 glycoprotein 1 antibody

anti-dsDNA anti-double stranded DNA antibody

APS antiphospholipid antibody syndrome

CNS Central nervous system

HEp-2 IF Indirect immunofluorescence on human epithelial cells,

hpf high power field

Ig immunoglobulin

ISN/RPS International Society of Nephrology/Renal Pathology Society

(neg) denotes negative association, e.g. presence of fatigue is more common in control than SLE subjects

RVVT Russel viper venom time

SCLE subacute cutaneous lupus erythematosus

UTI urinary tract infection