

BILAG2004 INDEX GLOSSARY

- all features must be attributable to SLE and refer to the last 4 weeks compared with the previous 4 weeks
- in some manifestations, it may be difficult to differentiate SLE from other causes as there may not be any specific test and the decision would then lie with the physician's judgement on the balance of probabilities
- definition of improvement: (a) the amount of improvement is sufficient for consideration of reduction in therapy
(b) improvement must be present ≥ 2 weeks of the previous 4 weeks
- most are self-explanatory but definitions are available for most descriptors
- ophthalmic manifestations need to be assessed by ophthalmologist
- for descriptors that are based on measurements (in renal and haematology systems), it is important to indicate if these are not due to lupus (for consideration of scoring) as they are usually recorded routinely into a database

CONSTITUTIONAL

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| 1. Pyrexia | temperature $> 37.5^{\circ}\text{C}$ documented |
| 2. Unintentional weight loss $> 5\%$ | |
| 3. Lymphadenopathy | palpable lymph node more than 1 cm diameter |
| 4. Fatigue or malaise or lethargy | |
| 5. Anorexia | |

MUCOCUTANEOUS

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| 6. Severe eruption | $> 18\%$ body surface area or bullous
includes discoid lesion |
| | body surface area (BSA) is defined using the
rules of nines (used to assess extent of burns) as
follows: |
| | palm(excluding fingers) = 1% BSA
each lower limb = 18% BSA
each upper limb = 9% BSA
torso (front) = 18% BSA
torso (back) = 18% BSA |

head = 9% BSA
genital (male) = 1% BSA

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| 7. Mild eruption | ≤ 18% body surface area
includes discoid lesion |
| 8. Angio-oedema | potentially life-threatening eg: stridor |
| 9. Severe mucosal ulceration | disabling extensive &/or deep ulceration |
| 10. Mild mucosal ulceration | localised non-disabling ulceration |
| 11. Severe panniculitis | any one:
affecting the face
> 9% body surface area
threatens integrity of epithelium &/or
subcutaneous tissue |
| 12. Mild panniculitis | ≤ 9% body surface area and does not fulfil any
criteria for severe panniculitis |
| 13. Cutaneous vasculitis/thrombosis | resulting in gangrene or ulceration or skin
infarction |
| 14. Digital infarct/nodular vasculitis | localised single or multiple infarct(s) over
digit(s) or tender erythematous nodule(s) |
| 15. Severe alopecia | clinically detectable diffuse or patchy hair loss
with scalp inflammation |
| 16. Mild alopecia | not clinically detectable and little/no scalp
inflammation (may be diffuse & must be
spontaneous) |
| 17. Peri-ungual erythema or chilblains | |
| 18. Splinter haemorrhages | |

NEUROPSYCHIATRIC

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| 19. Aseptic meningitis | criteria (all): acute/subacute onset
headache
photophobia
neck stiffness
fever
signs of meningeal irritation
abnormal CSF but negative cultures

exclude CNS/meningeal infection, intracranial
haemorrhage |
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20. Cerebral vasculitis	should be present with features of vasculitis in another system and supportive imaging &/or biopsy findings
21. Demyelinating syndrome	discrete white matter lesion with associated neurological deficit not recorded elsewhere there must have been at least one previously recorded event exclude multiple sclerosis
22. Myelopathy	acute onset of rapidly evolving paraparesis or quadriparesis and/or sensory level exclude intramedullary and extramedullary space occupying lesion
23. Acute confusional state	acute disturbance of consciousness or level of arousal with reduced ability to focus, maintain or shift attention includes hypo- and hyperaroused states and encompasses the spectrum from delirium to coma
24. Psychosis	delusion or hallucinations does not occur exclusively during course of a delirium exclude drugs, substance abuse, primary psychotic disorder
25. Acute inflammatory demyelinating polyradiculoneuropathy	criteria: progressive polyradiculoneuropathy loss of reflexes symmetrical involvement increased CSF protein without pleocytosis supportive abnormal nerve conduction study
26. Mononeuropathy (single/multiplex)	nerve conduction study not essential
27. Cranial neuropathy	except optic neuropathy which is classified elsewhere
28. Plexopathy	disorder of brachial or lumbosacral plexus resulting in neurological deficit not corresponding to territory of single root or nerve

	positive electrophysiology study required
29. Polyneuropathy	<p>symmetrical distal sensory and/or motor deficit</p> <p>positive electrophysiology study required</p>
30. Seizure disorder	independent description of seizure by reliable witness
31. Status epilepticus	a seizure or series of seizures lasting ≥ 30 minutes without full recovery to baseline
32. Cerebrovascular disease (not due to vasculitis)	<p>any one with supporting imaging:</p> <ul style="list-style-type: none"> stroke syndrome transient ischaemic attack intracranial haemorrhage <p>exclude hypoglycaemia, cerebral sinus thrombosis, vascular malformation, tumour, abscess</p> <p>cerebral sinus thrombosis not included as definite thrombosis not considered part of lupus activity</p>
33. Cognitive dysfunction	<p>significant deficits in any cognitive functions:</p> <ul style="list-style-type: none"> simple attention complex attention memory visual-spatial processing language reasoning/problem solving psychomotor speed executive functions <p>neuropsychological testing should be done if possible or corroborating history from third party that it is interfering with daily activities</p> <p>exclude substance abuse</p>
34. Movement disorder	exclude drugs
35. Autonomic disorder	<p>any one:</p> <ul style="list-style-type: none"> fall in blood pressure to standing $> 30/15$ mm Hg (systolic/diastolic) increase in heart rate to standing ≥ 30 bpm

	loss of heart rate variation with respiration (max – min < 15 bpm, expiration:inspiration ratio < 1.2, Valsalva ratio < 1.4)
	loss of sweating over body and limbs (anhidrosis) by sweat test
	exclude drugs and diabetes mellitus
36. Cerebellar ataxia	
37. Severe headache (unremitting)	disabling headache unresponsive to narcotic analgesia & lasting ≥ 3 days
	exclude intracranical space occupying lesion and CNS infection
38. Migraine with/without aura	recurrent attacks of headache lasting 4 - 72 hours
	may be preceded by neurological aura (lasting up to 1 hour)
39. Tension headache	recurrent episodes of headaches lasting minutes to days
40. Cluster headache	attacks of severe unilateral headache lasting 15 - 180 minutes
	attacks at least once every other day and up to 8 times a day
	attacks occur in clusters (series of weeks or months) separated by remissions of usually months or years
41. Headache from IC hypertension	exclude cerebral sinus thrombosis
42. Mood disorder (depression/mania)	prominent & persistent disturbance in mood characterised by depressed mood or markedly diminished interest or pleasure in almost all activities or elevated, expansive or irritable mood
	should result in significant distress or impaired functioning
43. Anxiety disorder	prominent anxiety, panic disorder, panic attacks or obsessions or compulsions resulting in clinically significant distress or impaired functioning

MUSCULOSKELETAL

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| 44. Definite myositis | ≥ 3 Bohan & Peter criteria:
proximal muscle weakness
elevated muscle enzymes
positive muscle biopsy
abnormal EMG |
| 45. Incomplete myositis | 2 Bohan & Peter criteria |
| 46. Severe polyarthritis | observed active synovitis ≥ 2 joints with
significant impairment of activities of daily
living and has been present on several days
(cumulatively) over the last 4 weeks |
| 47. Arthritis or Tendonitis | tendonitis or active synovitis ≥ 1 joint with some
impairment of function, which has been present
on several days over the last 4 weeks |
| 48. Arthralgia or Myalgia | inflammatory joint or muscle pain which does
not fulfil the above criteria for arthritis or
myositis |

CARDIORESPIRATORY

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| 49. Mild myocarditis | inflammation of myocardium with raised
cardiac enzymes &/or ECG changes and without
resulting cardiac failure, arrhythmia or valvular
dysfunction |
| 50. Cardiac failure | cardiac failure due to myocarditis or non-
infective inflammation of endocardium or
cardiac valves (endocarditis) |
| 51. Arrhythmia | arrhythmia (except sinus tachycardia) due to
myocarditis or non-infective inflammation of
endocardium or cardiac valves (endocarditis) |
| 52. New valvular dysfunction | new cardiac valvular dysfunction due to
myocarditis or non-infective inflammation of
endocardium or cardiac valves (endocarditis) |
| 53. Mild serositis (pleuro-pericardial pain) | in absence of cardiac tamponade or pleural
effusion with dyspnoea |

54. Cardiac tamponade	
55. Pleural effusion with dyspnoea	
56. Pulmonary haemorrhage/vasculitis	inflammation of pulmonary vasculature with haemoptysis &/or dyspnoea &/or pulmonary hypertension
	supporting imaging &/or histological diagnosis
57. Interstitial alveolitis/pneumonitis	radiological features of alveolar infiltration not due to infection or haemorrhage
	reduced corrected gas transfer Kco (< 70% normal)
58. Shrinking lung syndrome	reduced lung volumes (< 70% predicted) in presence of normal corrected gas transfer Kco with dysfunctional diaphragmatic movements
59. Aortitis	inflammation of aorta with or without dissection with supporting imaging abnormalities
	accompanied by > 10 mm Hg difference in BP between arms &/or claudication of extremities &/or vascular bruits
60. Coronary vasculitis	inflammation of coronary vessels with radiographic evidence of non-atheromatous narrowing, obstruction or aneurismal changes

GASTROINTESTINAL

61. Peritonitis	serositis presenting as acute abdomen with rebound/guarding
62. Serositis	not presenting as acute abdomen
63. Lupus enteritis or colitis	vasculitis or inflammation of small or large bowel with supportive imaging &/or biopsy findings
64. Malabsorption	diarrhoea with abnormal D- xylose absorption test or increased faecal fat excretion after exclusion of coeliac's disease (poor response to gluten-free diet) and gut vasculitis
65. Protein-losing enteropathy	diarrhoea with hypoalbuminaemia or increased fecal excretion of iv radiolabeled albumin after exclusion of gut vasculitis

66. Intestinal pseudo-obstruction	subacute intestinal obstruction due to intestinal hypomotility
67. Hepatitis	raised transaminases in absence of autoantibodies specific to autoimmune hepatitis (eg: anti-smooth muscle, anti-liver cytosol 1) &/or biopsy appearance of chronic active hepatitis
68. Acute cholecystitis	after exclusion of gallstones and infection
69. Acute pancreatitis	usually associated multisystem involvement

OPHTHALMIC

70. Orbital inflammation	
71. Severe keratitis	sight threatening includes: corneal melt peripheral ulcerative keratitis
72. Mild keratitis	not sight threatening
73. Anterior uveitis	
74. Severe posterior uveitis &/or retinal vasculitis	sight-threatening &/or retinal vasculitis not due to vaso-occlusive disease
75. Mild posterior uveitis &/or retinal vasculitis	not sight-threatening not due to vaso-occlusive disease
76. Episcleritis	
77. Severe scleritis	necrotising anterior scleritis anterior &/or posterior scleritis requiring systemic steroids/immunosuppression &/or not responding to NSAIDs
78. Mild scleritis	anterior &/or posterior scleritis not requiring systemic steroids excludes necrotising anterior scleritis
79. Retinal/choroidal vaso-occlusive disease	includes: retinal arterial & venous occlusion serous retinal &/or retinal pigment epithelial detachments secondary to choroidal vasculopathy
80. Isolated cotton-wool spots	also known as cytoid bodies

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| 81. Optic neuritis | excludes anterior ischaemic optic neuropathy |
| 82. Anterior ischaemic optic neuropathy | visual loss with pale swollen optic disc due to occlusion of posterior ciliary arteries |

RENAL

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| 83. Systolic blood pressure | |
| 84. Diastolic blood pressure | |
| 85. Accelerated hypertension | blood pressure rising to > 170/110 mm Hg within 1 month with grade 3 or 4 Keith-Wagener-Barker retinal changes (flame-shaped haemorrhages or cotton-wool spots or papilloedema) |
| 86. Urine dipstick | |
| 87. Urine albumin-creatinine ratio | on freshly voided urine sample |
| 88. Urine protein-creatinine ratio | on freshly voided urine sample |
| 89. 24 hour urine protein | |
| 90. Nephrotic syndrome | <p>criteria:</p> <p>heavy proteinuria (> 50 mg/kg/day or > 3.5 g/day or protein-creatinine ratio > 350 mg/mmol or albumin-creatinine ratio > 350 mg/mmol)</p> <p>hypoalbuminaemia</p> <p>oedema</p> |
| 91. Plasma/Serum creatinine | |
| 92. GFR | <p>MDRD formula:</p> $\text{GFR} = 170 \times [\text{serum creatinine(mg/dl)}]^{-0.999} \times [\text{age}]^{-0.176} \times [\text{serum urea(mg/dl)}]^{-0.17} \times [\text{serum albumin(g/dl)}]^{0.318} \times [0.762 \text{ if female}] \times [1.180 \text{ if black}]$ <p>conversion:</p> <p>serum creatinine - mg/dl = (μmol/l)/88.5</p> <p>serum urea - mg/dl = (mmol/l) x 2.8</p> <p>creatinine clearance not recommended as it is not reliable</p> |
| 93. Active urinary sediment | Uncentrifuged specimen: pyuria (> 5 WCC/hpf), haematuria (> 5 RBC/hpf) or red cell casts in absence of other causes |
| 94. Histology of active nephritis | WHO Class III, IV or V |

within last 3 months or since previous assessments if seen less than 3 months ago

glomerular sclerosis without inflammation not counted

HAEMATOLOGY

95. Haemoglobin

96. White cell count

97. Neutrophil count

98. Lymphocyte count

99. Platelet count

100. Evidence of active haemolysis

positive Coomb's test & evidence of haemolysis (raised bilirubin or raised reticulocyte count or reduced haptoglobulins)

101. Isolated positive Coomb's test

ADDITIONAL ITEMS

These items are required mainly for calculation of GFR

i. Date of Birth

ii. Weight

iii. Black

iv. Serum urea

v. Serum albumin