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 lies 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

1. Pyrexia	temperature > 37.5˚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

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
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

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

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	symmetrical distal sensory and/or motor deficit positive electrophysiology study required
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)	any one with supporting imaging: stroke syndrome transient ischaemic attack intracranial haemorrhage exclude hypoglycaemia, cerebral sinus thrombosis, vascular malformation, tumour, abscess cerebral sinus thrombosis not included as definite thrombosis not considered part of lupus activity
33. Cognitive dysfunction	significant deficits in any cognitive functions: simple attention complex attention memory visual-spatial processing language reasoning/problem solving psychomotor speed executive functions neuropsychological testing should be done if possible or corroborating history from third party that it is interfering with daily activities exclude substance abuse
34. Movement disorder	exclude drugs
35. Autonomic disorder	any one: 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

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

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

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

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	criteria: 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) hypoalbuminaemia oedema
91. Plasma/Serum creatinine	
92. GFR	MDRD formula: GFR = 170 x [serum creatinine(mg/dl)]$^{-0.999}$ x [age]$^{-0.176}$ x [serum urea(mg/dl]$^{-0.17}$ x [serum albumin(g/dl)]$^{0.318}$ x [0.762 if female] x [1.180 if black] conversion: serum creatinine - mg/dl = (μmol/l)/88.5 serum urea - mg/dl = (mmol/l) x 2.8 creatinine clearance not recommended as it is not reliable
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