

◆ **Only record manifestations/items due to SLE Disease Activity**

◆ **Assessment refers to manifestations occurring in the last 4 weeks (compared with the previous 4 weeks)**

◆ **TO BE USED WITH THE GLOSSARY**

Record: ND Not Done

0 Not present

1 Improving

2 Same

3 Worse

4 New

Yes/No OR Value (where indicated)

*Y/N Confirm this is due to SLE activity (Yes/No)

CONSTITUTIONAL

- 1. Pyrexia - documented > 37.5°C ()
- 2. Weight loss - unintentional > 5% ()
- 3. Lymphadenopathy/splenomegaly ()
- 4. Anorexia ()

MUCOCUTANEOUS

- 5. Skin eruption - severe ()
- 6. Skin eruption - mild ()
- 7. Angio-oedema - severe ()
- 8. Angio-oedema - mild ()
- 9. Mucosal ulceration - severe ()
- 10. Mucosal ulceration - mild ()
- 11. Panniculitis/Bullous lupus - severe ()
- 12. Panniculitis/Bullous lupus - mild ()
- 13. Major cutaneous vasculitis/thrombosis ()
- 14. Digital infarcts or nodular vasculitis ()
- 15. Alopecia - severe ()
- 16. Alopecia - mild ()
- 17. Peri-ungual erythema/chilblains ()
- 18. Splinter haemorrhages ()

NEUROPSYCHIATRIC

- 19. Aseptic meningitis ()
- 20. Cerebral vasculitis ()
- 21. Demyelinating syndrome ()
- 22. Myelopathy ()
- 23. Acute confusional state ()
- 24. Psychosis ()
- 25. Acute inflammatory demyelinating polyradiculoneuropathy ()
- 26. Mononeuropathy (single/multiplex) ()
- 27. Cranial neuropathy ()
- 28. Plexopathy ()
- 29. Polyneuropathy ()
- 30. Seizure disorder ()
- 31. Status epilepticus ()
- 32. Cerebrovascular disease (not due to vasculitis) ()
- 33. Cognitive dysfunction ()
- 34. Movement disorder ()
- 35. Autonomic disorder ()
- 36. Cerebellar ataxia (isolated) ()
- 37. Lupus headache - severe unremitting ()
- 38. Headache from IC hypertension ()

MUSCULOSKELETAL

- 39. Myositis - severe ()
- 40. Myositis - mild ()
- 41. Arthritis (severe) ()
- 42. Arthritis (moderate)/Tendonitis/Tenosynovitis ()
- 43. Arthritis (mild)/Arthralgia/Myalgia ()

CARDIORESPIRATORY

- 44. Myocarditis - mild ()
- 45. Myocarditis/Endocarditis + Cardiac failure ()
- 46. Arrhythmia ()
- 47. New valvular dysfunction ()
- 48. Pleurisy/Pericarditis ()
- 49. Cardiac tamponade ()
- 50. Pleural effusion with dyspnoea ()
- 51. Pulmonary haemorrhage/vasculitis ()
- 52. Interstitial alveolitis/pneumonitis ()
- 53. Shrinking lung syndrome ()
- 54. Aortitis ()
- 55. Coronary vasculitis ()

GASTROINTESTINAL

- 56. Lupus peritonitis ()
- 57. Abdominal serositis or ascites ()
- 58. Lupus enteritis/colitis ()
- 59. Malabsorption ()
- 60. Protein losing enteropathy ()
- 61. Intestinal pseudo-obstruction ()
- 62. Lupus hepatitis ()
- 63. Acute lupus cholecystitis ()
- 64. Acute lupus pancreatitis ()

OPHTHALMIC

- 65. Orbital inflammation/myositis/proptosis ()
- 66. Keratitis - severe ()
- 67. Keratitis - mild ()
- 68. Anterior uveitis ()
- 69. Posterior uveitis/retinal vasculitis - severe ()
- 70. Posterior uveitis/retinal vasculitis - mild ()
- 71. Episcleritis ()
- 72. Scleritis - severe ()
- 73. Scleritis - mild ()
- 74. Retinal/choroidal vaso-occlusive disease ()
- 75. Isolated cotton-wool spots (cytoid bodies) ()
- 76. Optic neuritis ()
- 77. Anterior ischaemic optic neuropathy ()

RENAL

- 78. Systolic blood pressure (mm Hg) value () Y/N*
- 79. Diastolic blood pressure (mm Hg) value () Y/N*
- 80. Accelerated hypertension Yes/No ()
- 81. Urine dipstick protein (+=1, ++=2, +++=3) () Y/N*
- 82. Urine albumin-creatinine ratio mg/mmol () Y/N*
- 83. Urine protein-creatinine ratio mg/mmol () Y/N*
- 84. 24 hour urine protein (g) value () Y/N*
- 85. Nephrotic syndrome Yes/No ()
- 86. Creatinine (plasma/serum) µmol/l () Y/N*
- 87. GFR (calculated) ml/min/1.73 m² () Y/N*
- 88. Active urinary sediment Yes/No ()
- 89. Active nephritis Yes/No ()

HAEMATOLOGICAL

- 90. Haemoglobin (g/dl) value () Y/N*
- 91. Total white cell count (x 10⁹/l) value () Y/N*
- 92. Neutrophils (x 10⁹/l) value () Y/N*
- 93. Lymphocytes (x 10⁹/l) value () Y/N*
- 94. Platelets (x 10⁹/l) value () Y/N*
- 95. TTP ()
- 96. Evidence of active haemolysis Yes/No ()
- 97. Coombs' test positive (isolated) Yes/No ()

Weight (kg):	Serum urea (mmol/l):
African ancestry: Yes/No	Serum albumin (g/l):

BILAG-2004 INDEX GLOSSARY

INSTRUCTIONS

- only record features that are **attributable to SLE disease activity and not due to damage, infection, thrombosis (in absence of inflammatory process) or other conditions**
- assessment refers to manifestations occurring in the **last 4 weeks compared with the previous 4 weeks**
- activity refers to disease process which is reversible while damage refers to permanent process/scarring (irreversible)
- damage due to SLE should be considered as a cause of features that are fixed/persistent (SLICC/ACR damage index uses persistence ≥ 6 months to define damage)
- in some manifestations, it may be difficult to differentiate SLE from other conditions as there may not be any specific test and the decision would then lie with the **physician's judgement on the balance of probabilities**
- ophthalmic manifestations usually need to be assessed by an ophthalmologist and these items would need to be recorded after receiving the response from the ophthalmologist
- guidance for scoring:

(4) NEW

- manifestations are recorded as new when it is a new episode occurring in the last 4 weeks (compared to the previous 4 weeks) that has not improved and this includes new episodes (recurrence) of old manifestations
- new episode occurring in the last 4 weeks but also satisfying the criteria for improvement (below) would be classified as improving instead of new

(3) WORSE

- this refers to manifestations that have deteriorated/worsened significantly in the last 4 weeks compared to the previous 4 weeks, sufficient for consideration of increase in therapy

(2) SAME

- this refers to manifestations that have been present for the last 4 weeks and the previous 4 weeks without significant improvement or deterioration (from the previous 4 weeks)
- this also applies to manifestations that have improved over the last 4 weeks compared to the previous 4 weeks but do not meet the criteria for improvement

(1) IMPROVING

- definition of **improvement**: (a) the amount of improvement is sufficient for

consideration of reduction in therapy and would not justify escalation in therapy

AND

(b) improvement must be **present currently and for at least 2 weeks** out of the last 4 weeks

OR

manifestation that has **completely resolved and remained absent** over the **whole of last 1 week**

(0) NOT PRESENT

(ND) NOT DONE

- it is important to indicate if a test has not been performed (particularly laboratory investigations) so that this will be recorded as such in the database & not as normal or absent (which is the default)

☐ INDICATE (TICK) IF NOT DUE TO SLE ACTIVITY

- for descriptors that are based on measurements (in renal and haematology systems), it is important to indicate if these are not due to lupus disease activity (for consideration of scoring) as they are usually recorded routinely into a database

CHANGE IN SEVERITY CATEGORY

- there are several items in the index which have been divided into categories of mild and severe (depending on definition). It is essential to record mild and severe items appropriately if the manifestations fulfil both criteria during the last 4 weeks
- if a mild item deteriorated to the extent that it fulfilled the definition of severe category (ie changed into severe category) within the last 4 weeks:
severe item scored as new (4)
AND mild item scored as worsening (3)
- if a severe item improved (fulfilling the improvement criteria) to the extent that it no longer fulfilled the definition of severe category (ie changed into mild category) within the last 4 weeks:
severe item scored as not present (0) if criteria for severe category has not been met over last 4 weeks
or as improving (1) if criteria for severe category has been met at some point over last 4 weeks

AND

mild item scored as improving (1) if it is improving over last 4 weeks
or as the same (2) if it has remained stable over last 4 weeks

CONSTITUTIONAL

1. Pyrexia

temperature > 37.5°C documented

2. Unintentional weight loss > 5%

3. Lymphadenopathy

lymph node more than 1 cm diameter

exclude infection

4. Anorexia

MUCOCUTANEOUS

5. Severe eruption

> 18% body surface area

any lupus rash except panniculitis, bullous lesion & angio-oedema

body surface area (BSA) is estimated using the rules of nines (used to assess extent of burns) as follows:

palm(excluding fingers) = 1% BSA

each lower limb = 18% BSA

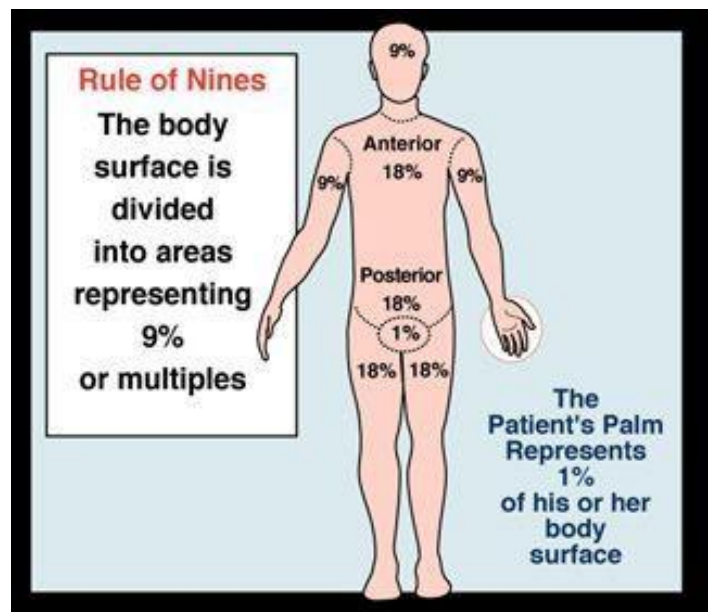
each upper limb = 9% BSA

torso (front) = 18% BSA

torso (back) = 18% BSA

head = 9% BSA

genital (male) = 1% BSA



6. Mild eruption

≤ 18% body surface area

	any lupus rash except panniculitis, bullous lesion & angio-oedema
	malar rash must have been observed by a physician and has to be present continuously (persistent) for at least 1 week to be considered significant (to be recorded)
7. Severe angio-oedema	potentially life-threatening eg: stridor
	angio-oedema is a variant form of urticaria which affects the subcutaneous, submucosal and deep dermal tissues
8. Mild angio-oedema	not life threatening
9. Severe mucosal ulceration	disabling (significantly interfering with oral intake), extensive & deep ulceration
	must have been observed by a physician
10. Mild mucosal ulceration	localised &/or non-disabling ulceration
11. Severe panniculitis or bullous lupus	any one: > 9% body surface area facial panniculitis panniculitis that is beginning to ulcerate panniculitis that threatens integrity of subcutaneous tissue (beginning to cause surface depression) on > 9% body surface area
	panniculitis presents as a palpable and tender subcutaneous induration/nodule
	note that established surface depression and atrophy alone is likely to be due to damage
12. Mild panniculitis or bullous lupus	≤ 9% body surface area does not fulfil any criteria for severe panniculitis (for panniculitis)
13. Major cutaneous vasculitis/thrombosis	resulting in extensive gangrene or ulceration or skin infarction
14. Digital infarct or 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 (redness over scalp)

16. Mild alopecia
diffuse or patchy hair loss without scalp inflammation (clinically detectable or by history)
17. Peri-ungual erythema or chilblains
chilblains are localised inflammatory lesions (may ulcerate) which are precipitated by exposure to cold
18. Splinter haemorrhages

NEUROPSYCHIATRIC

19. Aseptic meningitis
criteria (all): acute/subacute onset
headache
fever
abnormal CSF (raised protein &/or lymphocyte predominance) but negative cultures

preferably photophobia, neck stiffness and meningeal irritation should be present as well but are not essential for diagnosis

exclude CNS/meningeal infection, intracranial haemorrhage
20. Cerebral vasculitis
should be present with features of vasculitis in another system

supportive imaging &/or biopsy findings
21. Demyelinating syndrome
discrete white matter lesion with associated neurological deficit not recorded elsewhere

ideally there should have been at least one previously recorded event

supportive imaging required

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 electrophysiology study
26. Mononeuropathy (single/multiplex)	supportive electrophysiology study required
27. Cranial neuropathy	except optic neuropathy which is classified under ophthalmic system
28. Plexopathy	disorder of brachial or lumbosacral plexus resulting in neurological deficit not corresponding to territory of single root or nerve supportive electrophysiology study required
29. Polyneuropathy	acute symmetrical distal sensory and/or motor deficit supportive 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 (ability to register & maintain information)
complex attention
memory (ability to register, recall & recognise information eg learning, recall)
visual-spatial processing (ability to analyse, synthesise & manipulate visual-spatial information)
language (ability to comprehend, repeat & produce oral/written material eg verbal fluency, naming)
reasoning/problem solving (ability to reason & abstract)
psychomotor speed
executive functions (eg planning, organising, sequencing)

in absence of disturbance of consciousness or level of arousal

sufficiently severe to interfere with daily activities

neuropsychological testing should be done or corroborating history from third party if possible

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 \geq 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

cerebellar ataxia in isolation of other CNS features

usually subacute presentation

37. Severe lupus headache (unremitting) disabling headache unresponsive to narcotic analgesia & lasting ≥ 3 days

exclude intracranial space occupying lesion and CNS infection

38. Headache from IC hypertension exclude cerebral sinus thrombosis

MUSCULOSKELETAL

39. Severe myositis significantly elevated serum muscle enzymes with significant muscle weakness

exclude endocrine causes and drug-induced myopathy

electromyography and muscle biopsy are used for diagnostic purpose and are not required to determine level of activity

40. Mild myositis significantly elevated serum muscle enzymes with myalgia but without significant muscle weakness

asymptomatic elevated serum muscle enzymes not included

exclude endocrine causes and drug-induced myopathy

electromyography and muscle biopsy are used for diagnostic purpose and are not required to determine level of activity

41. Severe arthritis observed active synovitis ≥ 2 joints with marked loss of functional range of movements and significant impairment of activities of daily living, that has been present on several days (cumulatively) over the last 4 weeks

42. Moderate arthritis or Tendonitis or Tenosynovitis tendonitis/tenosynovitis or active synovitis ≥ 1 joint (observed or through history) with some loss of functional range of movements, that has been present on several days over the last 4 weeks

43. Mild arthritis or Arthralgia or Myalgia inflammatory type of pain (worse in the morning with stiffness, usually improves with activity & not brought on by activity) over joints/muscle

inflammatory arthritis which does not fulfil the above criteria for moderate or severe arthritis

CARDIORESPIRATORY

44. Mild myocarditis

inflammation of myocardium with raised cardiac enzymes &/or ECG changes and without resulting cardiac failure, arrhythmia or valvular dysfunction

45. Cardiac failure

cardiac failure due to myocarditis or non-infective inflammation of endocardium or cardiac valves (endocarditis)

cardiac failure due to myocarditis is defined by left ventricular ejection fraction $\leq 40\%$ & pulmonary oedema or peripheral oedema

cardiac failure due to acute valvular regurgitation (from endocarditis) can be associated with normal left ventricular ejection fraction

diastolic heart failure is not included

46. Arrhythmia

arrhythmia (except sinus tachycardia) due to myocarditis or non-infective inflammation of endocardium or cardiac valves (endocarditis)

confirmation by electrocardiogram required (history of palpitations alone inadequate)

47. New valvular dysfunction

new cardiac valvular dysfunction due to myocarditis or non-infective inflammation of endocardium or cardiac valves (endocarditis)

supportive imaging required

48. Pleurisy/Pericarditis

convincing history &/or physical findings that you would consider treating

in absence of cardiac tamponade or pleural effusion with dyspnoea

do not score if you are unsure whether or not it is pleurisy/pericarditis

49. Cardiac tamponade

supportive imaging required

50. Pleural effusion with dyspnoea

supportive imaging required

51. Pulmonary haemorrhage/vasculitis

inflammation of pulmonary vasculature with

	haemoptysis &/or dyspnoea &/or pulmonary hypertension
	supportive imaging &/or histological diagnosis required
52. Interstitial alveolitis/pneumonitis	radiological features of alveolar infiltration not due to infection or haemorrhage required for diagnosis
	corrected gas transfer Kco reduced to < 70% normal or fall of > 20% if previously abnormal
	on-going activity would be determined by clinical findings and lung function tests, and repeated imaging may be required in those with deterioration (clinically or lung function tests) or failure to respond to therapy
53. Shrinking lung syndrome	acute reduction (> 20% if previous measurement available) in lung volumes (to < 70% predicted) in the presence of normal corrected gas transfer (Kco) & dysfunctional diaphragmatic movements
54. Aortitis	inflammation of aorta (with or without dissection) with supportive imaging abnormalities
	accompanied by > 10 mm Hg difference in BP between arms &/or claudication of extremities &/or vascular bruits
	repeated imaging would be required to determine on-going activity in those with clinical deterioration or failure to respond to therapy
55. Coronary vasculitis	inflammation of coronary vessels with radiographic evidence of non-atheromatous narrowing, obstruction or aneurysmal changes
<u>GASTROINTESTINAL</u>	
56. Lupus peritonitis	serositis presenting as acute abdomen with rebound/guarding
57. Serositis	not presenting as acute abdomen
58. Lupus enteritis or colitis	vasculitis or inflammation of small or large bowel with supportive imaging &/or biopsy findings
59. 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
60. Protein-losing enteropathy	diarrhoea with hypoalbuminaemia or increased faecal excretion of iv radiolabeled albumin after exclusion of gut vasculitis and malabsorption
61. Intestinal pseudo-obstruction	subacute intestinal obstruction due to intestinal hypomotility
62. Lupus hepatitis	raised transaminases absence of autoantibodies specific to autoimmune hepatitis (eg: anti-smooth muscle, anti-liver cytosol 1) &/or biopsy appearance of chronic active hepatitis hepatitis typically lobular with no piecemeal necrosis exclude drug-induced and viral hepatitis
63. Acute lupus cholecystitis	after exclusion of gallstones and infection
64. Acute lupus pancreatitis	usually associated multisystem involvement

OPHTHALMIC

65. Orbital inflammation	orbital inflammation with myositis &/or extra-ocular muscle swelling &/or proptosis supportive imaging required
66. Severe keratitis	sight threatening includes: corneal melt peripheral ulcerative keratitis
67. Mild keratitis	not sight threatening
68. Anterior uveitis	
69. Severe posterior uveitis &/or retinal vasculitis	sight-threatening &/or retinal vasculitis not due to vaso-occlusive disease
70. Mild posterior uveitis &/or retinal vasculitis	not sight-threatening not due to vaso-occlusive disease
71. Episcleritis	
72. Severe scleritis	necrotising anterior scleritis

73. Mild scleritis
- anterior &/or posterior scleritis requiring systemic steroids/immunosuppression &/or not responding to NSAIDs
- anterior &/or posterior scleritis not requiring systemic steroids
- excludes necrotising anterior scleritis
74. Retinal/choroidal vaso-occlusive disease
- includes: retinal arterial & venous occlusion serous retinal &/or retinal pigment epithelial detachments secondary to choroidal vasculopathy
75. Isolated cotton-wool spots
- also known as cytoid bodies
76. Optic neuritis
- excludes anterior ischaemic optic neuropathy
77. Anterior ischaemic optic neuropathy
- visual loss with pale swollen optic disc due to occlusion of posterior ciliary arteries

RENAL

78. Systolic blood pressure
79. Diastolic blood pressure
80. 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)
81. Urine dipstick
82. Urine albumin-creatinine ratio
- on freshly voided urine sample
- conversion: 1 mg/mg = 113 mg/mmol
- it is important to exclude other causes (especially infection) when proteinuria is present
83. Urine protein-creatinine ratio
- on freshly voided urine sample
- conversion: 1 mg/mg = 113 mg/mmol
- it is important to exclude other causes (especially infection) when proteinuria is present
84. 24 hour urine protein
- it is important to exclude other causes (especially infection) when proteinuria is present
85. Nephrotic syndrome
- criteria:
- heavy proteinuria (≥ 3.5 g/day or protein-

creatinine ratio ≥ 350 mg/mmol or albumin-creatinine ratio ≥ 350 mg/mmol)

hypoalbuminaemia
oedema

86. Plasma/Serum creatinine

exclude other causes for increase in creatinine (especially drugs)

87. GFR

MDRD formula:

$$\text{GFR} = 170 \times [\text{serum creatinine (mg/dl)}]^{-0.999} \times [\text{age}]^{-0.176} \times [\text{serum urea (mg/dl)}]^{-0.17} \times [\text{serum albumin (g/dl)}]^{0.318} \times [0.762 \text{ if female}] \times [1.180 \text{ if African ancestry}]$$

units = ml/min per 1.73 m^2

normal: male = 130 ± 40

female = 120 ± 40

conversion:

serum creatinine - mg/dl = ($\mu\text{mol/l}$)/88.5

serum urea - mg/dl = (mmol/l) x 2.8

serum albumin - g/dl = (g/l)/10

creatinine clearance not recommended as it is not reliable

exclude other causes for decrease in GFR (especially drugs)

88. Active urinary sediment

pyuria (> 5 WCC/hpf or > 10 WCC/ mm^3 (μl))

OR

haematuria (> 5 RBC/hpf or > 10 RBC/ mm^3 (μl))

OR

red cell casts

OR

white cell casts

exclude other causes (especially infection, vaginal bleed, calculi)

89. Histology of active nephritis

WHO Classification (1995): (any one)

Class III – (a) or (b) subtypes

Class IV – (a), (b) or (c) subtypes

Class V – (a), (b), (c) or (d) subtypes

Vasculitis

OR

ISN/RPS Classification (2003): (any one)

Class III – (A) or (A/C) subtypes

Class IV – (A) or (A/C) subtypes

Class V

Vasculitis

within last 3 months

glomerular sclerosis without inflammation not included

HAEMATOLOGICAL

90. Haemoglobin

exclude dietary deficiency & GI blood loss

91. White cell count

exclude drug-induced cause

92. Neutrophil count

exclude drug-induced cause

93. Lymphocyte count

94. Platelet count

exclude drug-induced cause

95. TTP

thrombotic thrombocytopenic purpura

clinical syndrome of micro-angiopathic haemolytic anaemia and thrombocytopenia in absence of any other identifiable cause

96. Evidence of active haemolysis

positive Coombs' test & evidence of haemolysis (raised bilirubin or raised reticulocyte count or reduced haptoglobulins or fragmented RBC or microspherocytes)

97. Isolated positive Coombs' test

ADDITIONAL ITEMS

These items are required mainly for calculation of GFR

- i. Weight
- ii. African ancestry
- iii. Serum urea
- iv. Serum albumin

References:

- 1) Rule of nines diagram. Burn Center, University of Utah Health Sciences Center (<http://uuhsc.utah.edu/burncenter/emergencycare/extent.html>)
- 2) Levey, A. S., Bosch, J. P., Lewis, J. B., Greene, T., Rogers, N., & Roth, D. A more accurate method to estimate glomerular filtration rate from serum creatinine: a new prediction equation. Modification of Diet in Renal Disease Study Group. *Ann.Intern.Med.* 1999; 130(6): 461-470.
- 3) Weening, J. J., D'Agati, V. D., Schwartz, M. M., Seshan, S. V., Alpers, C. E., Appel, G. B., Balow, J. E., Bruijn, J. A., Cook, T., Ferrario, F., Fogo, A. B., Ginzler, E. M., Hebert, L., Hill, G., Hill, P., Jennette, J. C., Kong, N. C., Lesavre, P., Lockshin, M., Looi, L. M., Makino, H., Moura, L. A., & Nagata, M. The classification of glomerulonephritis in systemic lupus erythematosus revisited. *J.Am.Soc.Nephrol.* 2004; 15(2): 241-250.

BILAG-2004 INDEX SCORING

- scoring based on the principle of physician's intention to treat

Category	Definition
A	<p>Severe disease activity requiring any of the following treatment:</p> <ol style="list-style-type: none"> 1. systemic high dose oral glucocorticoids (equivalent to prednisolone > 20 mg/day) 2. intravenous pulse glucocorticoids (equivalent to pulse methylprednisolone \geq 500 mg) 3. systemic immunomodulators (include biologicals, immunoglobulins and plasmapheresis) 4. therapeutic high dose anticoagulation in the presence of high dose steroids or immunomodulators eg: warfarin with target INR 3 - 4
B	<p>Moderate disease activity requiring any of the following treatment:</p> <ol style="list-style-type: none"> 1. systemic low dose oral glucocorticoids (equivalent to prednisolone \leq 20 mg/day) 2. intramuscular or intra-articular or soft tissue glucocorticoids injection (equivalent to methylprednisolone < 500mg) 3. topical glucocorticoids 4. topical immunomodulators 5. antimalarials or thalidomide or prasterone or acitretin 6. symptomatic therapy eg: NSAIDs for inflammatory arthritis
C	Mild disease
D	Inactive disease but previously affected
E	System never involved

CONSTITUTIONAL

Category A:

Pyrexia recorded as 2 (same), 3 (worse) or 4 (new) **AND**

Any 2 or more of the following recorded as 2 (same), 3 (worse) or 4 (new):

Weight loss
Lymphadenopathy/splenomegaly
Anorexia

Category B:

Pyrexia recorded as 2 (same), 3 (worse) or 4 (new) **OR**

Any 2 or more of the following recorded as 2 (same), 3 (worse) or 4 (new):

Weight loss
Lymphadenopathy/splenomegaly
Anorexia

BUT do not fulfil criteria for Category A

Category C

Pyrexia recorded as 1 (improving) **OR**

One or more of the following recorded as > 0:

Weight loss
Lymphadenopathy/Splenomegaly
Anorexia

BUT does not fulfil criteria for category A or B

Category D

Previous involvement

Category E

No previous involvement

MUCOCUTANEOUS

Category A

Any of the following recorded as 2 (same), 3 (worse) or 4 (new):

- Skin eruption - severe
- Angio-oedema - severe
- Mucosal ulceration - severe
- Panniculitis/Bullous lupus - severe
- Major cutaneous vasculitis/thrombosis

Category B

Any Category A features recorded as 1 (improving) **OR**

Any of the following recorded as 2 (same), 3 (worse) or 4 (new):

- Skin eruption - mild
- Panniculitis/Bullous lupus - mild
- Digital infarcts or nodular vasculitis
- Alopecia - severe

Category C

Any Category B features recorded as 1 (improving) **OR**

Any of the following recorded as > 0:

- Angio-oedema - mild
- Mucosal ulceration - mild
- Alopecia - mild
- Periungual erythema/chilblains
- Splinter haemorrhages

Category D

Previous involvement

Category E

No previous involvement

NEUROPSYCHIATRIC

Category A

Any of the following recorded as 2 (same), 3 (worse) or 4 (new):

- Aseptic meningitis
- Cerebral vasculitis
- Demyelinating syndrome
- Myelopathy
- Acute confusional state
- Psychosis
- Acute inflammatory demyelinating polyradiculoneuropathy
- Mononeuropathy (single/multiplex)
- Cranial neuropathy
- Plexopathy
- Polyneuropathy
- Status epilepticus
- Cerebellar ataxia

Category B

Any Category A features recorded as 1 (improving) **OR**

Any of the following recorded as 2 (same), 3 (worse) or 4 (new):

- Seizure disorder
- Cerebrovascular disease (not due to vasculitis)
- Cognitive dysfunction
- Movement disorder
- Autonomic disorder
- Lupus headache - severe unremitting
- Headache due to raised intracranial hypertension

Category C

Any Category B features recorded as 1 (improving)

Category D

Previous involvement

Category E

No previous involvement

MUSCULOSKELETAL

Category A

Any of the following recorded as 2 (same), 3 (worse) or 4 (new):

Severe Myositis
Severe Arthritis

Category B

Any Category A features recorded as 1 (improving) **OR**

Any of the following recorded as 2 (same), 3 (worse) or 4 (new):

Mild Myositis
Moderate Arthritis/Tendonitis/Tenosynovitis

Category C

Any Category B features recorded as 1 (improving) **OR**

Any of the following recorded as > 0:

Mild Arthritis/Arthralgia/Myalgia

Category D

Previous involvement

Category E

No previous involvement

CARDIORESPIRATORY

Category A

Any of the following recorded as 2 (same), 3 (worse) or 4 (new):

- Myocarditis/Endocarditis + Cardiac failure
- Arrhythmia
- New valvular dysfunction
- Cardiac tamponade
- Pleural effusion with dyspnoea
- Pulmonary haemorrhage/vasculitis
- Interstitial alveolitis/pneumonitis
- Shrinking lung syndrome
- Aortitis
- Coronary vasculitis

Category B

Any Category A features recorded as 1 (improving) **OR**

Any of the following recorded as 2 (same), 3 (worse) or 4 (new):

- Pleurisy/Pericarditis
- Myocarditis - mild

Category C

Any Category B features recorded as 1 (improving)

Category D

Previous involvement

Category E

No previous involvement

GASTROINTESTINAL

Category A

Any of the following recorded as 2 (same), 3 (worse) or 4 (new):

- Peritonitis
- Lupus enteritis/colitis
- Intestinal pseudo-obstruction
- Acute lupus cholecystitis
- Acute lupus pancreatitis

Category B

Any Category A feature recorded as 1 (improving) **OR**

Any of the following recorded as 2 (same), 3 (worse) or 4 (new):

- Abdominal serositis and/or ascites
- Malabsorption
- Protein losing enteropathy
- Lupus hepatitis

Category C

Any Category B features recorded as 1 (improving)

Category D

Previous involvement

Category E

No previous involvement

OPHTHALMIC

Category A

Any of the following recorded as 2 (same), 3 (worse) or 4 (new):

- Orbital inflammation/myositis/proptosis
- Keratitis - severe
- Posterior uveitis/retinal vasculitis - severe
- Scleritis - severe
- Retinal/choroidal vaso-occlusive disease
- Optic neuritis
- Anterior ischaemic optic neuropathy

Category B

Any Category A features recorded as 1 (improving) **OR**

Any of the following recorded as 2 (same), 3 (worse) or 4 (new):

- Keratitis - mild
- Anterior uveitis
- Posterior uveitis/retinal vasculitis - mild
- Scleritis - mild

Category C

Any Category B features recorded as 1 (improving) **OR**

Any of the following recorded as > 0:

- Episcleritis
- Isolated cotton-wool spots (cytoid bodies)

Category D

Previous involvement

Category E

No previous involvement

RENAL

Category A

Two or more of the following **providing 1, 4 or 5 is included:**

1. Deteriorating proteinuria (severe) defined as

- (a) urine dipstick increased by ≥ 2 levels (used only if other methods of urine protein estimation not available); **or**
- (b) 24 hour urine protein > 1 g that has not decreased (improved) by $\geq 25\%$; **or**
- (c) urine protein-creatinine ratio > 100 mg/mmol that has not decreased (improved) by $\geq 25\%$; **or**
- (d) urine albumin-creatinine ratio > 100 mg/mmol that has not decreased (improved) by $\geq 25\%$

2. Accelerated hypertension

3. Deteriorating renal function (severe) defined as

- (a) plasma creatinine > 130 $\mu\text{mol/l}$ and having risen to $> 130\%$ of previous value; **or**
- (b) GFR < 80 ml/min per 1.73 m^2 and having fallen to $< 67\%$ of previous value; **or**
- (c) GFR < 50 ml/min per 1.73 m^2 , and last time was > 50 ml/min per 1.73 m^2 or was not measured.

4. Active urinary sediment

5. Histological evidence of active nephritis within last 3 months

6. Nephrotic syndrome

Category B

One of the following:

1. One of the Category A feature

2. Proteinuria (that has not fulfilled Category A criteria)

- (a) urine dipstick which has risen by 1 level to at least 2+ (used only if other methods of urine protein estimation not available); **or**
- (b) 24 hour urine protein ≥ 0.5 g that has not decreased (improved) by $\geq 25\%$; **or**
- (c) urine protein-creatinine ratio ≥ 50 mg/mmol that has not decreased (improved) by $\geq 25\%$; **or**
- (d) urine albumin-creatinine ratio ≥ 50 mg/mmol that has not decreased (improved) by $\geq 25\%$

3. Plasma creatinine > 130 $\mu\text{mol/l}$ and having risen to $\geq 115\%$ but $\leq 130\%$ of previous value

Category C

One of the following:

1. Mild/Stable proteinuria defined as

- (a) urine dipstick $\geq 1+$ but has not fulfilled criteria for Category A & B (used only if other methods of urine protein estimation not available); **or**

- (b) 24 hour urine protein > 0.25 g but has not fulfilled criteria for Category A & B ; **or**
- (c) urine protein-creatinine ratio > 25 mg/mmol but has not fulfilled criteria for Category A & B; **or**
- (d) urine albumin-creatinine ratio > 25 mg/mmol but has not fulfilled criteria for Category A & B

2. Rising blood pressure (providing the recorded values are > 140/90 mm Hg) which has not fulfilled criteria for Category A & B, defined as

- (a) systolic rise of ≥ 30 mm Hg; **and**
- (b) diastolic rise of ≥ 15 mm Hg

Category D

Previous involvement

Category E

No previous involvement

Note: although albumin-creatinine ratio and protein-creatinine ratio are different, we use the same cut-off values for this index

HAEMATOLOGICAL

Category A

TTP recorded as 2 (same), 3 (worse) or 4 (new) **OR**

Any of the following:

Evidence of haemolysis and Haemoglobin < 8 g/dl
Platelet count < 25 x 10⁹/l

Category B

TTP recorded as 1 (improving) **OR**

Any of the following:

Evidence of haemolysis and Haemoglobin 8 - 9.9 g/dl
Haemoglobin < 8 g/dl (without haemolysis)
White cell count < 1.0 x 10⁹/l
Neutrophil count < 0.5 x 10⁹/l
Platelet count 25 - 49 x 10⁹/l

Category C

Any of the following:

Evidence of haemolysis and Haemoglobin ≥ 10g/dl
Haemoglobin 8 - 10.9 g/dl (without haemolysis)
White cell count 1 - 3.9 x 10⁹/l
Neutrophil count 0.5 - 1.9 x 10⁹/l
Lymphocyte count < 1.0 x 10⁹/L
Platelet count 50 - 149 x 10⁹/l
Isolated Coombs' test positive

Category D

Previous involvement

Category E

No previous involvement