**BILAG-2004 INDEX**  
**Centre:**  
**Date:**  
**Initials/Hosp No:**

- 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

<table>
<thead>
<tr>
<th>Record</th>
<th>Description</th>
<th>Value</th>
</tr>
</thead>
<tbody>
<tr>
<td>ND</td>
<td>Not Done</td>
<td></td>
</tr>
<tr>
<td>0</td>
<td>Not present</td>
<td></td>
</tr>
<tr>
<td>1</td>
<td>Improving</td>
<td></td>
</tr>
<tr>
<td>2</td>
<td>Same</td>
<td></td>
</tr>
<tr>
<td>3</td>
<td>Worse</td>
<td></td>
</tr>
<tr>
<td>4</td>
<td>New</td>
<td></td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Yes/No OR Value (where indicated)</th>
</tr>
</thead>
<tbody>
<tr>
<td><em>Y/N</em> Confirm this is due to SLE activity (Yes/No)</td>
</tr>
</tbody>
</table>

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

### HAEMATOLOGICAL

90. Haemoglobin (g/dl) value ( )
91. Total white cell count (x 10⁹/l) value ( )
92. Neutrophils (x 10⁹/l) value ( )
93. Lymphocytes (x 10⁹/l) value ( )
94. Platelets (x 10⁹/l) value ( )
95. TTP ( )
96. Evidence of active haemolysis Yes/No ( )
97. Coombs’ test positive (isolated) Yes/No ( )
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 \( \geq 6 \) 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 lies 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)
<p>| | |</p>
<table>
<thead>
<tr>
<th></th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td>16. Mild alopecia</td>
<td>diffuse or patchy hair loss without scalp inflammation (clinically detectable or by history)</td>
</tr>
<tr>
<td>17. Peri-ungual erythema or chilblains</td>
<td>chilblains are localised inflammatory lesions (may ulcerate) which are precipitated by exposure to cold</td>
</tr>
<tr>
<td>18. Splinter haemorrhages</td>
<td></td>
</tr>
</tbody>
</table>

**NEUROPSYCHIATRIC**

<p>| | |</p>
<table>
<thead>
<tr>
<th></th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td>19. Aseptic meningitis</td>
<td>criteria (all): acute/subacute onset headache fever abnormal CSF (raised protein &amp;/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</td>
</tr>
<tr>
<td>20. Cerebral vasculitis</td>
<td>should be present with features of vasculitis in another system supportive imaging &amp;/or biopsy findings</td>
</tr>
<tr>
<td>21. Demyelinating syndrome</td>
<td>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</td>
</tr>
<tr>
<td>22. Myelopathy</td>
<td>acute onset of rapidly evolving paraparesis or quadriplegia and/or sensory level exclude intramedullary and extramedullary space occupying lesion</td>
</tr>
<tr>
<td>23. Acute confusional state</td>
<td>acute disturbance of consciousness or level of arousal with reduced ability to focus, maintain or shift attention</td>
</tr>
</tbody>
</table>

Revision: 1/Sep/2009
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 ≥ 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 ≤ 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

GASTROINTESTINAL

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 extraocular 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
anterior &/or posterior scleritis requiring systemic steroids/immunosuppression &/or not responding to NSAIDs

73. Mild scleritis
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

**RENAI**

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

units = ml/min per 1.73 m²
normal: male = 130 ± 40
female = 120 ± 40

conversion:

serum creatinine - mg/dl = (μmol/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/mm³ (μl))

OR

haematuria (> 5 RBC/hpf or > 10 RBC/mm³ (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 exclude drug-induced cause
94. Platelet count thrombotic thrombocytopenic purpura
95. TTP 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)


### BILAG-2004 INDEX SCORING

- Scoring based on the principle of physician’s intention to treat

<table>
<thead>
<tr>
<th>Category</th>
<th>Definition</th>
</tr>
</thead>
</table>
| A | Severe disease activity requiring any of the following treatment:  
1. systemic high dose oral glucocorticoids (equivalent to prednisolone > 20 mg/day)  
2. intravenous pulse glucocorticoids (equivalent to pulse methylprednisolone ≥ 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 | Moderate disease activity requiring any of the following treatment:  
1. systemic low dose oral glucocorticoids (equivalent to prednisolone ≤ 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 ≥ 25%; or
   (c) urine protein-creatinine ratio > 100 mg/mmol that has not decreased (improved) by ≥ 25%; or
   (d) urine albumin-creatinine ratio > 100 mg/mmol that has not decreased (improved) by ≥ 25%

2. Accelerated hypertension
3. Deteriorating renal function (severe) defined as
   
   (a) plasma creatinine > 130 μmol/l and having risen to > 130% of previous value; or
   (b) GFR < 80 ml/min per 1.73 m² and having fallen to < 67% of previous value; or
   (c) GFR < 50 ml/min per 1.73 m², and last time was > 50 ml/min per 1.73 m² 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 ≥ 25%; or
   (c) urine protein-creatinine ratio ≥ 50 mg/mmol that has not decreased (improved) by ≥ 25%; or
   (d) urine albumin-creatinine ratio ≥ 50 mg/mmol that has not decreased (improved) by ≥ 25%

3. Plasma creatinine > 130 μmol/l and having risen to ≥ 115% but ≤ 130% of previous value

Category C
One of the following:

1. Mild/Stable proteinuria defined as
   
   (a) urine dipstick ≥ 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 10^9/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 10^9/l
   Neutrophil count < 0.5 x 10^9/l
   Platelet count 25 - 49 x 10^9/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 10^9/l
   Neutrophil count 0.5 - 1.9 x 10^9/l
   Lymphocyte count < 1.0 x 10^9/L
   Platelet count 50 - 149 x 10^9/l
   Isolated Coombs’ test positive

Category D
Previous involvement

Category E
No previous involvement