

Appendix 2 Classic BILAG Index

All features must be attributable to SLE and refer to last 4 weeks compared with prior disease activity

Indicate features which are present: 1) Improving

2) Same

3) Worse

4) New

Or Y/N or value (where indicated)

(default is 0 = not present)

GENERAL

- | | |
|-------------------------------------|-----|
| 1. Pyrexia (documented) | () |
| 2. Weight loss - unintentional > 5% | () |
| 3. Lymphadenopathy/splenomegaly | () |
| 4. Fatigue/malaise/lethargy | () |
| 5. Anorexia/nausea/vomiting | () |

MUCOCUTANEOUS

- | | |
|---|---------|
| 6. Maculopapular eruption - severe, active (or discoid/bullous) | () |
| 7. Maculopapular eruption - mild | () |
| 8. Active discoid lesions – generalised or extensive | () |
| 9. Active discoid lesions - localised (including lupus profundus) | () |
| 10. Alopecia (severe, active) | () |
| 11. Alopecia (mild) | () |
| 12. Panniculitis (severe) | () |
| 13. Angio-oedema | () |
| 14. Extensive mucosal ulceration | () |
| 15. Small mucosal ulcers | () |
| 16. Malar erythema | () |
| 17. Subcutaneous nodules | () |
| 18. Perniotic skin lesions | () |
| 19. Peri-ungual erythema | () |
| 20. Swollen fingers | Y/N () |
| 21. Sclerodactyly | Y/N () |
| 22. Calcinosis | Y/N () |
| 23. Telangiectasia | Y/N () |

NEUROLOGICAL

- | | |
|--|-----|
| 24. Deteriorating level of consciousness | () |
| 25. Acute psychosis or delirium or confusional state | () |
| 26. Seizures | () |
| 27. Stroke or stroke syndrome | () |
| 28. Aseptic meningitis | () |
| 29. Mononeuritis multiplex | () |
| 30. Ascending or transverse myelitis | () |
| 31. Peripheral or cranial neuropathy | () |
| 32. Disc swelling/cytoid bodies | () |

- 33. Chorea ()
- 34. Cerebellar ataxia ()
- 35. Headache severe, unremitting ()
- 36. Organic depressive illness ()
- 37. Organic brain syndrome including pseudotumor cerebri ()
- 38. Episodic migraine headaches ()

MUSCULOSKELETAL

- 39. Definite myositis (Bohan & Peter) ()
- 40. Severe polyarthritis - with loss of function ()
- 41. Arthritis ()
- 42. Tendonitis ()
- 43. Mild chronic myositis ()
- 44. Arthralgia ()
- 45. Myalgia ()
- 46. Tendon contractures and fixed deformity Y/N ()
- 47. Aseptic necrosis Y/N ()

CARDIOVASCULAR & RESPIRATORY

- 48. Pleuropericardial pain ()
- 49. Dyspnoea ()
- 50. Cardiac failure ()
- 51. Friction rub ()
- 52. Effusion (pericardial or pleural) ()
- 53. Mild or intermittent chest pain ()
- 54. Progressive Chest X-ray changes-lung fields Y/N ()
- 55. Progressive Chest X-ray changes-heart size Y/N ()
- 56. ECG evidence of pericarditis or myocarditis Y/N ()
- 57. Cardiac arrhythmias including tachycardia > 100 in absence of fever Y/N ()
- 58. Pulmonary function fall by > 20% Y/N ()
- 59. Cytohistological evidence of inflammatory lung disease Y/N ()

VASCULITIS

- 60. Major cutaneous vasculitis including ulcers ()
- 61. Major abdominal crisis due to vasculitis ()
- 62. Recurrent thromboembolism (excluding strokes) ()
- 63. Raynaud's ()
- 64. Livedo reticularis ()
- 65. Superficial phlebitis ()
- 66. Minor cutaneous vasculitis(nail fold vasculitis, digital vasculitis, purpura, urticaria) ()
- 67. Thromboembolism (excluding stroke) - 1st episode Y/N ()

RENAL

- 68. Systolic blood pressure (mm Hg) value ()
- 69. Diastolic blood pressure (mm Hg) value ()
- 70. Accelerated hypertension Y/N ()

71. Urine dipstick protein (+ = 1, ++ = 2, +++ = 3)	<i>value</i>	()
72. 24 hour urinary protein (g)	<i>value</i>	()
73. Newly documented proteinuria > 1g / 24hrs	<i>Y/N</i>	()
74. Nephrotic syndrome	<i>Y/N</i>	()
75. Creatinine (plasma/serum)	<i>value</i>	()
76. Creatinine clearance/GFR ml/min	<i>value</i>	()
77. Active urinary sediment	<i>Y/N</i>	()
78. Histological evidence of active nephritis - within 3 months	<i>Y/N</i>	()

HAEMATOLOGY

79. Haemoglobin g/dl	<i>value</i>	()
80. Total white cell count x 10 ⁹ /l	<i>value</i>	()
81. Neutrophils x 10 ⁹ /L	<i>value</i>	()
82. Lymphocytes x 10 ⁹ /L	<i>value</i>	()
83. Platelets x 10 ⁹ /L	<i>value</i>	()
84. Evidence of active haemolysis	<i>Y/N</i>	()
85. Coomb's test positive	<i>Y/N</i>	()
86. Evidence of circulating anticoagulant	<i>Y/N</i>	()

CLASSIC BILAG GLOSSARY

It is implicit in this scoring system that all features scored are considered to be due to active lupus.

GENERAL

1. Pyrexia: temperature > 37.5°C documented
2. Weight loss unintentional more than 5%
3. Lymphadenopathy: palpable lymph nodes > 1cm in diameter
4. Fatigue or malaise or lethargy
5. Anorexia or nausea or vomiting

MUCOCUTANEOUS

6. Maculopapular eruption – severe: active maculopapular or bullous eruption; extensive (> 18% of body surface area), scarring or causing disability.
7. Maculopapular eruption – mild (\leq 18% body surface area), non-scarring, non-disabling
8. Active discoid lesions – generalised or extensive, > 18% surface area
9. Active discoid lesions – localised ie \leq 18% surface area
10. Alopecia – severe, active: abnormal diffuse hair loss which is clinically detectable with scalp inflammation.
11. Alopecia (mild): limited relatively inactive: abnormal diffuse hair loss with little or no detectable scalp inflammation.
12. Panniculitis: extensive, painful, erythematous subcutaneous nodules associated with fat necrosis resolve with scarring, or lupus profundus: erythematous elevated plaques with an overlying discoid skin lesion
13. Angio-oedema: potentially life threatening eg stridor
14. Extensive mucosal ulceration: severe, deep, disabling ulcers.

NEUROLOGICAL

24. Acute deteriorating level of consciousness by any accepted clinical criteria
25. Acute psychosis or delirium or confusional state: severe disturbance in the perception of reality characterised by: delusions, hallucinations, incoherence, marked illogical thinking, bizarre or catatonic behaviour
35. Headache severe, unremitting: continuous headache not relieved by narcotic analgesia
36. Organic depressive illness: associated with somatic symptoms and severe enough to merit treatment with anti-depressive medication
37. Organic brain syndrome: impaired orientation, memory or other intellectual function in the absence of metabolic, psychiatric or pharmacological causes
Clinical features develop over a short period (usually hours to days) and tend to fluctuate over the course of the day:
 - a. Clouding of consciousness with reduced capacity to focus and sustain attention to environment.
 - b.
 - i. Perceptual disturbance: misinterpretations, illusions or hallucinations
 - ii. incoherent speech
 - iii. insomnia or daytime drowsiness

- iv. increased or decreased psychomotor activity
- c. Disorientation and recent memory impairment

MUSCULOSKELETAL

- 39. Myositis: at least three of – proximal muscle weakness, elevated muscle enzymes, positive muscle biopsy and abnormal EMG.
- 40. Polyarthritis with loss of function: active joint inflammation with clinically significant loss of the functional range of movement of the involved joints.
- 41. Arthritis: active joint inflammation without loss of functional range of motion

CARDIO RESPIRATORY

- 48. Pleuropericardial pain: localised sharp or dull pain in the chest aggravated by respiration.
- 49. Dyspnoea: on exercise (not orthopnoea alone).
- 53. Mild intermittent chest pain: non-specific (not clearly pleuritic, pericardial, musculoskeletal or angina).
- 58. Pulmonary function fall by > 20%: >20% of expected (predicted for height, weight, sex and age) or > 20% fall in total lung capacity (forced vital capacity) and/or DLCO.

VASCULAR SYSTEM

- 59. Major cutaneous vasculitis including ulcers: extensive gangrene and/or ulceration
- 66. Minor cutaneous vasculitis eg digital vasculitis with nail fold infarcts

RENAL

- 70. Accelerated hypertension: BP rising to >170/110 (5th phase) within one month, if accompanied by Grade IV retinal changes (ie haemorrhage, exudates).
- 77. Active urinary sediment: on uncentrifuged specimen. Pyuria (> 5wc/hpf), haematuria (>5rbc/hpf) or red cell casts in the absence of infection or any other cause
- 78. Histological evidence of active nephritis: according to WHO criteria. Sclerosis alone (without inflammation) will not be regarded as active nephritis

HAEMATOLOGY

- 79. Evidence of circulating lupus anticoagulant or other antiphospholipid antibody.

CLASSIC BILAG INDEX SCORING

General

Category A

Pyrexia scoring 2 (same), 3 (worse) or 4 (new) **AND**
≥ 2 others scoring 2 (same), 3 (worse) or 4 (new)

Category B

Pyrexia scoring 2 (same), 3 (worse) or 4 (new) **OR**
≥ 2 others scoring 2 (same), 3 (worse) or 4 (new)

BUT do not fulfill criteria for Category A

Category C

Pyrexia scoring 1 (improving) **OR**
One or more of others scoring > 0

BUT do not fulfill criteria for Category A or B

Category D

Previous involvement

Category E

No previous involvement