SUPPLEMENTARY DATA

Search strategy
A systematic search was undertaken using PubMed and the Cochrane Database of Systematic Reviews to find original papers and systematic reviews with or without meta-analysis in the English language using the terms shown below in the supplementary table S1. The questions about the management of lupus developed by the guideline development group to be addressed by the literature review were:

i) What clinical and serological features should prompt consideration of a diagnosis of SLE?

ii) How should SLE patients be assessed?

iii) How should SLE patients be monitored in the non-acute setting?

iv) What is the evidence for the management of mild SLE?

v) What is the evidence for the management of moderate SLE?

vi) What is the evidence for the management of severe SLE?

Papers covering purely animal studies, pediatric studies, narrative review articles (except systematic reviews), commentaries, conference abstracts or statements, expert opinion statements and other guidelines were excluded (although such papers were checked manually for additional relevant references). We only reviewed papers that included the following numbers of patients (with search terms as described below): background, prevalence & prognosis a minimum 50 SLE patients, for diagnosis, assessment & monitoring a minimum 10 patients, for therapy a minimum 5 patients. Papers meeting these selection criteria were graded according to the SIGN revised grading system for recommendations in evidence based guidelines as shown in supplementary table S2 (1).
**Supplementary table S1: Search terms used in PubMed and Cochrane Database of Systematic Reviews for the literature review**

<table>
<thead>
<tr>
<th>Section of guideline</th>
<th>Topic</th>
<th>Search terms used in addition to SLE OR Systemic Lupus Erythematosus OR Lupus</th>
</tr>
</thead>
</table>
| Diagnosis and background | Clinical | Diagnosis  
Clinical manifestations/ Manifestations  
Clinical features  
Presentation  
Classification |
| | Serologic | Immunology/Immunological  
Antibody/auto-antibody/serological  
Anti-nuclear antibodies, ANA, anti-dsDNA, anti-Ro, anti-Sm, C3, C4, anti-phospholipid, antiphospholipid, anti-cardiolipin, anticardiolipin, lupus anticoagulant |
| | Lupus manifestations including differences between lupus in males and females | SLE activity  
Disease Damage  
Mortality  
Presentation  
Outcome  
ACR classification criteria  
Malar rash  
Discoid Rash  
Photosensitivity  
Oral Ulcers  
Nonerosive arthritis  
Pleuritis OR Pericarditis  
Proteinuria OR Cellular casts  
Neuropsychiatric  
Haemolytic anaemia OR Leucopenia/Leukopenia OR Lymphopenia OR Thrombocytopenia  
anti-double stranded DNA OR anti-Sm OR antiphospholipid antibodies OR anti-phospholipid antibodies OR ANA  
+- gender differences  
+- male/men/man |
| For assessment and monitoring | Lupus features | All above items AND  
Assess/ assessment  
Activity/ disease activity/BILAG/SLEDAI  
Monitoring  
Damage/ SLICC  
Prognosis  
Quality indicators  
Recommendations |
| | Neuro-psychiatric disease | Neuropsychiatric AND  
Prevalence  
Risk factors  
Screening  
Diagnosis  
Monitoring  
Prevention  
Prognosis |
| Malignancy                        | Cancer OR Malignancy AND Mortality  
|                                 | Lymphoma                        
|                                 | HPV OR cervical dysplasia OR cervical 
|                                 | Lung                            
|                                 | Prostate                        
|                                 | Endometrial                     
|                                 | Ovarian                         
|                                 | Screen                          
| Infection                       | Infection Risk AND/OR Death     
|                                 | Antibiotic prophylaxis vaccin*   
|                                 | Bacteria* Infections             
|                                 | CMV                             
|                                 | HPV                             
|                                 | Varicella Zoster virus           
|                                 | Hepatitis B AND C               
|                                 | Hepatitis vaccin*               
|                                 | Pneumocystis jiroveci           
|                                 | TB OR Tuberculosis               
| Treatment                       | Treatment or therapy or trial or study or management) AND 
|                                 | Therapy NAME AND/OR Mild or Moderate or Severe Activity or damage or flare BILAG or SLEDAI or ECLAM or SLAM or disease activity index Efficacy or safety or outcome Non-renal Constitutional Rash or mucocutaneous or dermatol* Vasculitis Arthritis or musculoskeletal Cardiac or respiratory or cardio-respiratory or gastro-intestinal Neuro-psychiatric or neuro*  
| Hydroxychloroquine/chloroquine/mepacrine | Methotrexate                     
|                                 | NSAIDs                          
|                                 | Sunscreen/sunblock              
|                                 | Prednisolone/prednisone/methylprednisolone/methylprednisone/triamcinolone/corticosteroid* Azathioprine Ciclosporin/cyclosporine/cyclosporine/tacrolimus Mycophenolate mofetil/mycophenolic acid Leflunomide Rituximab Belimumab Intra-venous immunoglobulin/intravenous immunoglobulin/IVIG Plasma exchange/plasmapharesis |
Supplementary table S2: SIGN revised grading system for recommendations in evidence based guidelines

<table>
<thead>
<tr>
<th>SIGN Levels of evidence</th>
<th>SIGN Grades of recommendations</th>
</tr>
</thead>
<tbody>
<tr>
<td>1++ High quality meta-analyses, systematic reviews of RCTs, or RCTs with a very low risk of bias</td>
<td>A At least one meta-analysis, systematic review, or RCT rated as 1++ and directly applicable to the target population or a systematic review of RCTs or a body of evidence consisting principally of studies rated as 1+ directly applicable to the target population and demonstrating overall consistency of results</td>
</tr>
<tr>
<td>1+ Well conducted meta-analyses, systematic reviews of RCTs, or RCTs with a low risk of bias</td>
<td>B A body of evidence including studies rated as 2++ directly applicable to the target population and demonstrating overall consistency of results or Extrapolated evidence from studies rated as 1++ or 1+</td>
</tr>
<tr>
<td>1− Meta-analyses, systematic reviews or RCTs, or RCTs with a high risk of bias</td>
<td>C A body of evidence including studies rated as 2+ directly applicable to the target population and demonstrating overall consistency of results or Extrapolated evidence from studies rated as 2++</td>
</tr>
<tr>
<td>2++ High quality systematic reviews of case-control or cohort studies or High quality case-control or cohort studies with a very low risk of confounding, bias, or chance and a high probability that the relationship is causal</td>
<td>D Evidence level 3 or 4 or Extrapolated evidence from studies rated as 2+</td>
</tr>
<tr>
<td>2+ Well conducted case-control or cohort studies with a low risk of confounding, bias, or chance and a moderate probability that the relationship is causal</td>
<td></td>
</tr>
<tr>
<td>2− Case-control or cohort studies with a high risk of confounding, bias, or chance and a significant risk that the relationship is not causal</td>
<td></td>
</tr>
<tr>
<td>3 Non-analytic studies, e.g. case reports, case series</td>
<td></td>
</tr>
<tr>
<td>4 Expert opinion</td>
<td></td>
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</table>

Supplementary Table S3: Cumulative incidence of SLE manifestations in lupus cohorts

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</thead>
<tbody>
<tr>
<td>Number of patients in cohort studied</td>
<td>(n=100)</td>
<td>(n=1214)</td>
<td>(n=600)</td>
<td>(n=1000)</td>
<td>(n=1156)</td>
<td>(n=500)</td>
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<td><strong>Constitutional</strong></td>
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<tr>
<td>Fever</td>
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<td>58</td>
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<td>28^a</td>
<td>16^e</td>
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<td>ANA</td>
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<td>99</td>
<td>-</td>
<td>82&lt;sup&gt;d&lt;/sup&gt;</td>
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<td>Anti-dsDNA</td>
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<td>90</td>
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<td>64&lt;sup&gt;e&lt;/sup&gt;</td>
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<td>10</td>
<td>64&lt;sup&gt;e&lt;/sup&gt;</td>
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<tr>
<td>Anticardiolipin IgG/IgM</td>
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<td>51/39</td>
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<td>-</td>
<td>64&lt;sup&gt;e&lt;/sup&gt;</td>
<td>21/9</td>
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<tr>
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<td>30</td>
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<td>Anti-Ro</td>
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<tr>
<td>Anti-RNP</td>
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<td>Low C3</td>
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<tr>
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<td>38</td>
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</table>

<sup>a</sup> combined incidence for items with same value. <sup>b</sup> confirmed arthritis only (usually non-erosive). <sup>c</sup> all neurological features associated with lupus combined. <sup>d</sup> possible failure of ascertainment but patients met ≥4 ACR criteria. <sup>e</sup> combined as met ACR criteria for immunological involvement. - not reported.
**CONSTITUTIONAL**
1. Pyrexia - documented > 37.5°C
2. Weight loss - unintentional > 5%
3. Lymphadenopathy/splenomegaly
4. Anorexia

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

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

**TOPIC:**
- Weight (kg): 
- Serum urea (mmol/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 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) (9) 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 \( \leq 18\% \) 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 \( \leq 9\% \) 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 quadriplegia 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 $\geq 30$ 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  
tendonitis/tenosynovitis or active synovitis ≥ 1

or Tenosynovitis 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
50. Pleural effusion with dyspnoea
51. Pulmonary haemorrhage/vasculitis

supportive imaging required

supportive imaging required

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

**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 (10):

\[
GFR = 170 \times [\text{serum creatinine (mg/dl)}]^{0.999} \times [\text{age}]^{0.116} \times [\text{serum urea (mg/dl)}]^{-0.17} \times [\text{serum albumin (g/dl)}]^{0.318} \times [0.762 \text{ if female}] \times [1.180 \text{ if African ancestry}]
\]

units = ml/min per 1.73 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

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) (11): (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 haptoglobin 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
## 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

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 $\geq 1+$ 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 $\geq 30$ mm Hg; and
   (b) diastolic rise of $\geq 15$ 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

**SLEDAI-2000 index**

**Name/ID:**

**Date of Birth:**

**Date of Assessment:**

**SLEDAI-2000 index data collection form**

(Circle in SLEDAI Score column if descriptor is present at the time of the visit or in the preceding 10 days)

<table>
<thead>
<tr>
<th><strong>SLEDAI Score</strong></th>
<th><strong>Descriptor</strong></th>
<th><strong>Definition</strong></th>
</tr>
</thead>
<tbody>
<tr>
<td>8</td>
<td>Seizure</td>
<td>Recent onset, exclude metabolic, infectious or drug causes</td>
</tr>
<tr>
<td>8</td>
<td>Psychosis</td>
<td>Altered ability to function in normal activity due to severe disturbance in the perception of reality. Include hallucinations, incoherence, marked loose associations, impoverished thought content, marked illogical thinking, bizarre, disorganised, or catatonic behaviour. Exclude uraemia and drug causes</td>
</tr>
<tr>
<td>8</td>
<td>Organic brain syndrome</td>
<td>Altered mental function with impaired orientation, memory, or other intellectual function, with rapid onset and fluctuating clinical features, inability to sustain attention to environment, plus at least 2 of the following: perceptual disturbance, incoherent speech, insomnia or daytime drowsiness, or increased or decreased psychomotor activity. Exclude metabolic, infectious or drug causes</td>
</tr>
<tr>
<td>8</td>
<td>Visual disturbance</td>
<td>Retinal changes of SLE. Include cytoid bodies, retinal hemorrhages, serous exudates or hemorrhages in the choroid, or optic neuritis. Exclude hypertension, infection, or drug causes</td>
</tr>
<tr>
<td>8</td>
<td>Cranial nerve disorder</td>
<td>New onset of sensory or motor neuropathy involving cranial nerves</td>
</tr>
<tr>
<td>8</td>
<td>Lupus headache</td>
<td>Severe, persistent headache; may be migrainous, but must be non-responsive to narcotic analgesia</td>
</tr>
<tr>
<td>8</td>
<td>CVA</td>
<td>New onset Cerebrovascular accident(s). Exclude arteriosclerosis</td>
</tr>
<tr>
<td>8</td>
<td>Vasculitis</td>
<td>Ulceration, gangrene, tender finger nodules, periungual infarction, splinter hemorrhages or biopsy or angiogram proof of vasculitis</td>
</tr>
<tr>
<td>4</td>
<td>Arthritis</td>
<td>$\geq 2$ joints with pain and signs of inflammation (i.e. tenderness, swelling or effusion)</td>
</tr>
<tr>
<td>4</td>
<td>Myositis</td>
<td>Proximal muscle aching/weakness, associated with elevated creatinine phosphokinase (CK)/aldolase, or EMG changes or a biopsy showing myositis</td>
</tr>
<tr>
<td>4</td>
<td>Urinary casts</td>
<td>Heme-granular or RBC casts</td>
</tr>
<tr>
<td>4</td>
<td>Hematuria</td>
<td>$&gt; 5$ RBC/high power field. Exclude stone, infection or other cause</td>
</tr>
<tr>
<td>4</td>
<td>Proteinuria</td>
<td>$&gt; 0.5$ gram/24 hours</td>
</tr>
<tr>
<td>4</td>
<td>Pyuria</td>
<td>$&gt; 5$ WBC/high power field. Exclude infection</td>
</tr>
<tr>
<td>2</td>
<td>Rash</td>
<td>Inflammatory type rash</td>
</tr>
<tr>
<td>2</td>
<td>Alopecia</td>
<td>Abnormal, patchy or diffuse loss of hair</td>
</tr>
<tr>
<td>2</td>
<td>Mucosal ulcers</td>
<td>Oral or nasal ulcerations</td>
</tr>
<tr>
<td>2</td>
<td>Pleurisy</td>
<td>Pleuritic chest pain with pleural rub or effusion, or pleural thickening</td>
</tr>
<tr>
<td>2</td>
<td>Pericarditis</td>
<td>Pericardial pain with at least 1 of the following: rub, effusion or ECG or echocardiogram confirmation</td>
</tr>
<tr>
<td>2</td>
<td>Low complement</td>
<td>Decrease in CH50, C3 or C4 below lower limit of normal for testing laboratory</td>
</tr>
<tr>
<td>2</td>
<td>Increased DNA binding</td>
<td>Increased DNA binding above normal range for testing laboratory</td>
</tr>
<tr>
<td>1</td>
<td>Fever</td>
<td>$&gt; 38^\circ C$. Exclude infectious cause</td>
</tr>
<tr>
<td>1</td>
<td>Thrombocytopenia</td>
<td>$&lt; 100 \times 10^9$ platelets/L, exclude drug causes</td>
</tr>
<tr>
<td>1</td>
<td>Leukopenia</td>
<td>$&lt; 3 \times 10^9$ WBC/L, exclude drug causes</td>
</tr>
</tbody>
</table>

**TOTAL SCORE:**

# SELENA version of SLEDAI

SELENA-SLEDAI index data collection form

*(Circle in SLEDAI Score column if descriptor is present at the time of the visit or in the preceding 4 weeks)*

<table>
<thead>
<tr>
<th>Item no.</th>
<th>SLEDAI SCORE</th>
<th>Descriptor</th>
<th>Definition</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>8</td>
<td>Seizure</td>
<td>Recent onset, exclude metabolic, infectious or drug causes</td>
</tr>
<tr>
<td>2</td>
<td>8</td>
<td>Psychosis</td>
<td>Altered ability to function in normal activity due to severe disturbance in the perception of reality. Include hallucinations, incoherence, marked loose associations, impoverished thought content, marked illogical thinking, bizarre, disorganised, or catatonic behaviour. Exclude uraemia and drug causes</td>
</tr>
<tr>
<td>3</td>
<td>8</td>
<td>Organic brain syndrome</td>
<td>Altered mental function with impaired orientation, memory, or other intellectual function, with rapid onset and fluctuating clinical features, inability to sustain attention to environment, plus at least 2 of the following: perceptual disturbance, incoherent speech, insomnia or daytime drowsiness, or increased or decreased psychomotor activity. Exclude metabolic, infectious or drug causes</td>
</tr>
<tr>
<td>4</td>
<td>8</td>
<td>Visual disturbance</td>
<td>Retinal changes of SLE. Include cytoid bodies, retinal hemorrhages, serous exudates or hemorrhages in the choroid, or optic neuritis, scleritis or episcleritis. Exclude hypertension, infection, or drug causes</td>
</tr>
<tr>
<td>5</td>
<td>8</td>
<td>Cranial nerve disorder</td>
<td>New onset of sensory or motor neuropathy involving cranial nerves</td>
</tr>
<tr>
<td>6</td>
<td>8</td>
<td>Lupus headache</td>
<td>Severe, persistent headache; may be migrainous, but must be non-responsive to narcotic analgesia</td>
</tr>
<tr>
<td>7</td>
<td>8</td>
<td>CVA</td>
<td>New onset cerebrovascular accident(s). Exclude arteriosclerosis</td>
</tr>
<tr>
<td>8</td>
<td>8</td>
<td>Vasculitis</td>
<td>Ulceration, gangrene, tender finger nodules, periungual infarction, splinter hemorrhages or biopsy or angiogram proof of vasculitis</td>
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<tr>
<td>9</td>
<td>4</td>
<td>Arthritis</td>
<td>&gt; 2 joints with pain and signs of inflammation (i.e. tenderness with swelling or effusion)</td>
</tr>
<tr>
<td>10</td>
<td>4</td>
<td>Myositis</td>
<td>Proximal muscle aching/weakness, associated with elevated creatinine phosphokinase (CK)/aldolase, or EMG changes or a biopsy showing myositis</td>
</tr>
<tr>
<td>11</td>
<td>4</td>
<td>Urinary casts</td>
<td>Heme-granular or RBC casts</td>
</tr>
<tr>
<td>12</td>
<td>4</td>
<td>Hematuria</td>
<td>&gt; 5 RBC/high power field. Exclude stone, infection or other cause</td>
</tr>
<tr>
<td>13</td>
<td>4</td>
<td>Proteinuria</td>
<td>New onset or recent increase of more than 0.5 gm/24 hours</td>
</tr>
<tr>
<td>14</td>
<td>4</td>
<td>Pyuria</td>
<td>&gt; 5 WBC/high power field. Exclude infection</td>
</tr>
<tr>
<td>15</td>
<td>2</td>
<td>Rash</td>
<td>Inflammatory type rash</td>
</tr>
<tr>
<td>16</td>
<td>2</td>
<td>Alopecia</td>
<td>Abnormal, patchy or diffuse loss of hair</td>
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<td>17</td>
<td>2</td>
<td>Mucosal ulcers</td>
<td>Oral or nasal ulcerations</td>
</tr>
<tr>
<td>18</td>
<td>2</td>
<td>Pleurisy</td>
<td>Pleuritic chest pain or pleural rub or effusion, or pleural thickening (does not require an objective component if medically convincing)</td>
</tr>
<tr>
<td>19</td>
<td>2</td>
<td>Pericarditis</td>
<td>Classic pericardial pain and/or rub, effusion or ECG or echocardiogram confirmation (does not require an objective component if medically convincing)</td>
</tr>
<tr>
<td>20</td>
<td>2</td>
<td>Low complement</td>
<td>Decrease in CH50, C3 or C4 &lt; lower limit of nl for testing laboratory</td>
</tr>
<tr>
<td>21</td>
<td>2</td>
<td>Increased DNA binding</td>
<td>Increased DNA binding above normal range for testing laboratory</td>
</tr>
<tr>
<td>22</td>
<td>1</td>
<td>Fever</td>
<td>&gt; 38°C. Exclude infectious cause</td>
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<tr>
<td>23</td>
<td>1</td>
<td>Thrombocytopenia</td>
<td>&lt; 100 x 10⁹ platelets/L, exclude drug causes</td>
</tr>
<tr>
<td>24</td>
<td>1</td>
<td>Leukopenia</td>
<td>&lt; 3 x 10⁹ WBC/L, exclude drug causes</td>
</tr>
</tbody>
</table>

*Total SCORE*

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References


