**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:

1. What clinical and serological features should prompt consideration of a diagnosis of SLE?
2. How should SLE patients be assessed?
3. How should SLE patients be monitored in the non-acute setting?
4. What is the evidence for the management of mild SLE?
5. What is the evidence for the management of moderate SLE?
6. 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 S 2 (1).

**Supplementary table S1: Search terms used in PubMed and Cochrane Database of Systematic Reviews for the literature review**

|  |  |  |
| --- | --- | --- |
|  | **Topic** | * + **Search terms used in addition to SLE OR Systemic Lupus Erythematosus OR Lupus** |
| **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** | Hydroxychloroquine/chloroquine/mepacrine  Methotrexate  NSAIDs  Sunscreen/sunblock  Prednisolone/prednisone/methylprednisolone/methylprednisone/triamcinolone/corticosteroid\*  Azathioprine  Ciclosporin/cyclosporine/cyclosporin/cyclosporine/tacrolimus  Mycophenolate mofetil/mycophenolic acid  Leflunomide  Rituximab  Belimumab  Intra-venous immunoglobulin/intravenous immunoglobulin/IVIG  Plasma exchange/plasmapharesis | 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\* |

**Supplementary table S2: SIGN revised grading system for recommendations in evidence based guidelines**

|  |  |
| --- | --- |
| **SIGN Levels of evidence** | **SIGN Grades of recommendations** |
| **1++** High quality meta-analyses, systematic reviews of RCTs, or RCTs with a very low risk of bias  **1+** Well conducted meta-analyses, systematic reviews of RCTs, or RCTs with a low risk of bias  **1−** Meta-analyses, systematic reviews or RCTs, or RCTs with a high risk of bias  **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  **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  **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  **3**  Non-analytic studies, e.g. case reports, case series  **4** Expert opinion | **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  **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+  **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++  **D** Evidence level 3 or 4 or  Extrapolated evidence from studies rated as 2+ |

Reproduced from A new system for grading recommendations in evidence based guidelines, Harbour R, Miller J, 323, 334-6, 2001withpermission from BMJ Publishing Group Ltd [1].

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

|  |  |  |  |  |  |  |
| --- | --- | --- | --- | --- | --- | --- |
| **Cumulative % incidence of SLE manifestations in lupus cohorts** | | | | | | |
| **Author**  **Year**  **Reference** | **Worral**  **1990**  **(2)** | **Pons**  **Estel**  **2004**  **(3)** | **Font**  **2004**  **(4)** | **Cervera**  **2009**  **(5)** | **Lim**  **2014**  **(6)** | **Isenberg**  **2010**  **(7)** |
| **Number of patients in cohort studied** | (n=100) | (n=1214) | (n=600) | (n=1000) | (n=1156) | (n=500) |
| **Constitutional**  Fever  Weight Loss  Lymphadenopathy | -  -  - | 57  27  15 | 42  -  1 | 17  -  - | -  -  - | -  -  - |
| **Cutaneous**  Alopecia  Oral/nasal ulcers  Photosensitivity  Malar rash  Discoid rash  Subacute cutaneous  Raynaud’s phenomenon | 27  36  48  90a  90a  -  - | 58  42  56  61  12  3  28 | 18  30  41  54  6  8  22 | -  13  23  31  8  -  16 | -  22  26  32  23  -  - | -  26  35  62a  62a  -  - |
| **Musculoskeletal**  Arthralgia/Arthritis  Myalgia/myositis | 94  - | 93  18 | 83  7 | 48b  4 | 67b  - | 94  - |
| **Cardiorespiratory**  Pericarditis  Pleurisy  Pneumonitis  Myocarditis  Endocarditis | 57 **a**  57 a  -  -  - | 17  22  2  3  3 | 28a  28a  4  2  8 | 16a  16a  -  -  - | 43a  43a  -  -  - | 43a  43a  -  -  - |
| **Neurological**  Seizures  Psychosis  Chorea  Transverse myelitis  Organic brain syndrome | 45c  45c  45c  45c  45c | 8  4  0.4  0.6  2 | 12a  12a  0.5  -  - | 19a  19a  -  -  - | 14a  14a  -  -  - | 21c  21c  21c  -  - |
| **Renal**  Proteinuria/sediment  Nephrotic Syndrome  ESRD | 29  -  - | 46  7  2 | 34  -  - | 28  -  - | 34  -  7 | 31  -  - |
| **Gastrointestinal**  Ascites  Liver | -  - | 1  - | -  0.3 | -  - | -  - | -  - |
| **Haematological**  Haemolytic anaemia  Leucopenia  Lymphopenia  Thrombocytopenia  Thrombosis | -  57  81  21  - | 12  42  59  19  6 | 8  66  82  31  7 | 5  -  -  13  9 | -  75a  75a  -  - | -  -  -  -  - |
| **Serological**  ANA  Anti-dsDNA  Anti-Smith  Anticardiolipin IgG/IgM  Lupus anticoagulant  Anti-Ro  Anti-RNP  Rheumatoid factor  Low C3  Low C4 | 99  55  -  34  19  -  -  27  -  - | 98  71  48  51/39  30  49  51  -  49  54 | 99  90  13  -  15  23  -  12  31  38 | -  -  10  -  -  25  13  18  -  - | 82d  64e  64e  64e  -  -  -  -  -  - | 95  64  13  21/9  14  37  27  25  44  - |

a combined incidence for items with same value. bconfirmed arthritis only (usually non-erosive). call neurological features associated with lupus combined. dpossible failure of ascertainment but patients met ≥4 ACR criteria. ecombined as met ACR criteria for immunological involvement. - not reported.

♦ Only record manifestations/items due to SLE Disease Activity

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

♦ TO BE USED WITH THE GLOSSARY

Record: **ND Not Done**

**0 Not present**

**1 Improving**

**2 Same**

**3 Worse**

**4 New**

**Yes/No OR Value (where indicated)**

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

**CONSTITUTIONAL**

1. Pyrexia - documented > 37.5ºC ( )

2. Weight loss - unintentional > 5% ( )

3. Lymphadenopathy/splenomegaly ( )

4. Anorexia ( )

**MUCOCUTANEOUS**

5. Skin eruption - severe ( )

6. Skin eruption - mild ( )

7. Angio-oedema - severe ( )

8. Angio-oedema - mild ( )

9. Mucosal ulceration - severe ( )

10. Mucosal ulceration - mild ( )

11. Panniculitis/Bullous lupus - severe ( )

12. Panniculitis/Bullous lupus - mild ( )

13. Major cutaneous vasculitis/thrombosis ( )

14. Digital infarcts or nodular vasculitis ( )

15. Alopecia - severe ( )

16. Alopecia - mild ( )

17. Peri-ungual erythema/chilblains ( )

18. Splinter haemorrhages ( )

**NEUROPSYCHIATRIC**

19. Aseptic meningitis ( )

20. Cerebral vasculitis ( )

21. Demyelinating syndrome ( )

22. Myelopathy ( )

23. Acute confusional state ( )

24. Psychosis ( )

25. Acute inflammatory demyelinating ( )

polyradiculoneuropathy

26. Mononeuropathy (single/multiplex) ( )

27. Cranial neuropathy ( )

28. Plexopathy ( )

29. Polyneuropathy ( )

30. Seizure disorder ( )

31. Status epilepticus ( )

32. Cerebrovascular disease (not due to vasculitis) ( )

33. Cognitive dysfunction ( )

34. Movement disorder ( )

35. Autonomic disorder ( )

36. Cerebellar ataxia (isolated) ( )

37. Lupus headache - severe unremitting ( )

38. Headache from IC hypertension ( )

**MUSCULOSKELETAL**

39. Myositis - severe ( )

40. Myositis - mild ( )

41. Arthritis ( severe) ( )

42. Arthritis (moderate)/Tendonitis/Tenosynovitis ( )

43. Arthritis (mild)/Arthralgia/Myalgia ( )

**Weight (kg): Serum urea (mmol/l):**

**African ancestry: Yes/No Serum albumin (g/l):**

**CARDIORESPIRATORY**

44. Myocarditis - mild ( )

45. Myocarditis/Endocarditis + Cardiac failure ( )

46. Arrhythmia ( )

47. New valvular dysfunction ( )

48. Pleurisy/Pericarditis ( )

49. Cardiac tamponade ( )

50. Pleural effusion with dyspnoea ( )

51. Pulmonary haemorrhage/vasculitis ( )

52. Interstitial alveolitis/pneumonitis ( )

53. Shrinking lung syndrome ( )

54. Aortitis ( )

55. Coronary vasculitis ( )

**GASTROINTESTINAL**

56. Lupus peritonitis ( )

57. Abdominal serositis or ascites ( )

58. Lupus enteritis/colitis ( )

59. Malabsorption ( )

60. Protein losing enteropathy ( )

61. Intestinal pseudo-obstruction ( )

62. Lupus hepatitis ( )

63. Acute lupus cholecystitis ( )

64. Acute lupus pancreatitis ( )

**OPHTHALMIC**

65. Orbital inflammation/myositis/proptosis ( )

66. Keratitis - severe ( )

67. Keratitis - mild ( )

68. Anterior uveitis ( )

69. Posterior uveitis/retinal vasculitis - severe ( )

70. Posterior uveitis/retinal vasculitis - mild ( )

71. Episcleritis ( )

72. Scleritis - severe ( )

73. Scleritis - mild ( )

74. Retinal/choroidal vaso-occlusive disease ( )

75. Isolated cotton-wool spots (cytoid bodies) ( )

76. Optic neuritis ( )

77. Anterior ischaemic optic neuropathy ( )

**RENAL**

78. Systolic blood pressure (mm Hg) value ( ) **Y/N\***

79. Diastolic blood pressure (mm Hg) value ( ) **Y/N\***

80. Accelerated hypertension Yes/No ( )

81. Urine dipstick protein (+=1, ++=2, +++=3) ( ) **Y/N\***

82. Urine albumin-creatinine ratio mg/mmol ( ) **Y/N\***

83. Urine protein-creatinine ratio mg/mmol ( ) **Y/N\***

84. 24 hour urine protein (g) value ( ) **Y/N\***

85. Nephrotic syndrome Yes/No ( )

86. Creatinine (plasma/serum) μmol/l ( ) **Y/N\***

87. GFR (calculated) ml/min/1.73 m2 ( ) **Y/N\***

88. Active urinary sediment Yes/No ( )

89. Active nephritis Yes/No ( )

**HAEMATOLOGICAL**

90. Haemoglobin (g/dl) value ( ) **Y/N\***

91. Total white cell count (x 109/l) value ( ) **Y/N\***

92. Neutrophils (x 109/l) value ( ) **Y/N\***

93. Lymphocytes (x 109/l) value ( ) **Y/N\***

94. Platelets (x 109/l) value ( ) **Y/N\***

95. TTP ( )

96. Evidence of active haemolysis Yes/No ( )

97. Coombs’ test positive (isolated) Yes/No ( )

**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

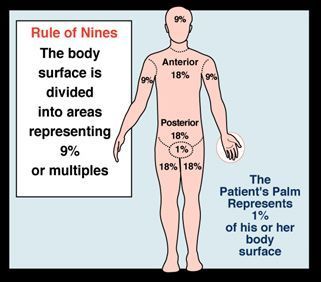
        each upper limb = 9% BSA

        torso (front) = 18% BSA

        torso (back) = 18% BSA

        head = 9% BSA

        genital (male) = 1% BSA



6. Mild eruption **≤** 18% body surface area

any lupus rash except panniculitis, bullous lesion

& angio-oedema

malar rash must have been observed by a

physician and has to be present continuously

(persistent) for at least 1 week to be considered

significant (to be recorded)

7. Severe angio-oedema potentially life-threatening eg: stridor

angio-oedema is a variant form of urticaria

which affects the subcutaneous, submucosal and

deep dermal tissues

8. Mild angio-oedema not life threatening

9. Severe mucosal ulceration disabling (significantly interfering with oral

intake), extensive & deep ulceration

must have been observed by a physician

10. Mild mucosal ulceration localised &/or non-disabling ulceration

11. Severe panniculitis or bullous lupus any one:

> 9% body surface area

facial panniculitis

panniculitis that is beginning to ulcerate

panniculitis that threatens integrity of

subcutaneous tissue (beginning to cause

surface depression) on > 9% body surface

area

panniculitis presents as a palpable and tender

subcutaneous induration/nodule

note that established surface depression and

atrophy alone is likely to be due to damage

12. Mild panniculitis or bullous lupus ≤ 9% body surface area

does not fulfil any criteria for severe panniculitis (for panniculitis)

13. Major cutaneous vasculitis/thrombosis resulting in extensive gangrene or ulceration or

skin infarction

14. Digital infarct or nodular vasculitis localised single or multiple infarct(s) over

digit(s) or tender erythematous nodule(s)

15. Severe alopecia clinically detectable (diffuse or patchy) hair loss

with scalp inflammation (redness over scalp)

16. Mild alopecia diffuse or patchy hair loss without scalp inflammation (clinically detectable or by history)

17. Peri-ungual erythema or chilblains chilblains are localised inflammatory lesions

(may ulcerate) which are precipitated by

exposure to cold

18. Splinter haemorrhages

**NEUROPSYCHIATRIC**

19. Aseptic meningitis criteria (all): acute/subacute onset

headache

fever

abnormal CSF (raised protein &/or

lymphocyte predominance) but negative

cultures

preferably photophobia, neck stiffness and

meningeal irritation should be present as well but

are not essential for diagnosis

exclude CNS/meningeal infection, intracranial

haemorrhage

20. Cerebral vasculitis should be present with features of vasculitis

in another system

supportive imaging &/or biopsy findings

21. Demyelinating syndrome discrete white matter lesion with associated

neurological deficit not recorded elsewhere

ideally there should have been at least one previously recorded event

supportive imaging required

exclude multiple sclerosis

22. Myelopathy acute onset of rapidly evolving paraparesis or

quadriparesis and/or sensory level

exclude intramedullary and extramedullary

space occupying lesion

23. Acute confusional state acute disturbance of consciousness or level of

arousal with reduced ability to focus, maintain or shift attention

includes hypo- and hyperaroused states and encompasses the spectrum from delirium to coma

24. Psychosis delusion or hallucinations

does not occur exclusively during course of a

delirium

exclude drugs, substance abuse, primary

psychotic disorder

25. Acute inflammatory demyelinating criteria:

polyradiculoneuropathy 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 any one with supporting imaging:

(not due to vasculitis) 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 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 extra-

ocular muscle swelling &/or proptosis

supportive imaging required

66. Severe keratitis sight threatening

includes: corneal melt

peripheral ulcerative keratitis

67. Mild keratitis not sight threatening

68. Anterior uveitis

69. Severe posterior uveitis &/or retinal sight-threatening &/or retinal vasculitis

vasculitis not due to vaso-occlusive disease

70. Mild posterior uveitis &/or retinal not sight-threatening

vasculitis

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 includes: retinal arterial & venous occlusion

disease 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 x [serum creatinine (mg/dl)]-0.999 x

[age]-0.176 x [serum urea (mg/dl]-0.17 x

[serum albumin (g/dl)]0.318 x [0.762 if

female] x [1.180 if 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) [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 thrombocytopaenic purpura

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

96. Evidence of active haemolysis positive Coombs’ test & evidence of haemolysis (raised bilirubin or raised reticulocyte count or reduced haptoglobulins or fragmented RBC or microspherocytes)

97. Isolated positive Coombs’ test

**ADDITIONAL ITEMS**

These items are required mainly for calculation of GFR

i. Weight

ii. African ancestry

iii. Serum urea

iv. Serum albumin

**BILAG-2004 INDEX SCORING**

• scoring based on the principle of physician’s intention to treat

|  |  |
| --- | --- |
| **Category** | **Definition** |
| **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 andhaving risen to ≥ 115% but ≤ 130% of previous value

**Category C**

One of the following:

1. Mild/Stable proteinuria defined as

1. 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 ≥ 15mm 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 data collection form**

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

**preceding 10 days**)

|  |  |  |
| --- | --- | --- |
| **SLEDAI SCORE** | **Descriptor** | **Definition** |
| 8 | Seizure | Recent onset, exclude metabolic, infectious or drug causes |
| 8 | Psychosis | 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 |
| 8 | Organic brain syndrome | 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 |
| 8 | Visual disturbance | 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 |
| 8 | Cranial nerve disorder | New onset of sensory or motor neuropathy involving cranial nerves |
| 8 | Lupus headache | Severe, persistent headache; may be migrainous, but must be non-responsive to narcotic analgesia |
| 8 | CVA | New onset Cerebrovascular accident(s). Exclude arteriosclerosis |
| 8 | Vasculitis | Ulceration, gangrene, tender finger nodules, periungual infarction, splinter hemorrhages or biopsy or angiogram proof of vasculitis |
| 4 | Arthritis | ≥ 2 joints with pain and signs of inflammation (i.e. tenderness, swelling or effusion) |
| 4 | Myositis | Proximal muscle aching/weakness, associated with elevated creatinine phosphokinase (CK)/aldolase, or EMG changes or a biopsy showing myositis |
| 4 | Urinary casts | Heme-granular or RBC casts |
| 4 | Hematuria | > 5 RBC/high power field. Exclude stone, infection or other cause |
| 4 | Proteinuria | > 0.5 gram/24 hours |
| 4 | Pyuria | > 5 WBC/high power field. Exclude infection |
| 2 | Rash | Inflammatory type rash |
| 2 | Alopecia | Abnormal, patchy or diffuse loss of hair |
| 2 | Mucosal ulcers | Oral or nasal ulcerations |
| 2 | Pleurisy | Pleuritic chest pain with pleural rub or effusion, or pleural thickening |
| 2 | Pericarditis | Pericardial pain with at least 1 of the following: rub, effusion or ECG or echocardiogram confirmation |
| 2 | Low complement | Decrease in CH50, C3 or C4 below lower limit of normal for testing laboratory |
| 2 | Increased DNA binding | Increased DNA binding above normal range for testing laboratory |
| 1 | Fever | > 38ºC. Exclude infectious cause |
| 1 | Thrombocytopenia | < 100 x 109 platelets/L, exclude drug causes |
| 1 | Leukopenia | < 3 x 109 WBC/L, exclude drug causes |

**TOTAL SCORE:**

Reprinted with permission from The Journal of Rheumatology, Gladman DD et al. Systemic lupus erythematosus disease activity index 2000. J Rheumatol 2002;29(2):288. All rights reserved [12].

**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)***

|  |  |  |  |
| --- | --- | --- | --- |
| **Item no.** | **SLEDAI SCORE** | **Descriptor** | **Definition** |
| 1 | **8** | **Seizure** | Recent onset, exclude metabolic, infectious or drug causes |
| 2 | **8** | **Psychosis** | 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 |
| 3 | **8** | **Organic brain syndrome** | 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 |
| 4 | **8** | **Visual disturbance** | 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 |
| 5 | **8** | **Cranial nerve disorder** | New onset of sensory or motor neuropathy involving cranial nerves |
| 6 | **8** | **Lupus headache** | Severe, persistent headache; may be migrainous, but must be non-responsive to narcotic analgesia |
| 7 | **8** | **CVA** | New onset cerebrovascular accident(s). Exclude arteriosclerosis |
| 8 | **8** | **Vasculitis** | Ulceration, gangrene, tender finger nodules, periungual infarction, splinter hemorrhages or biopsy or angiogram proof of vasculitis |
| 9 | **4** | **Arthritis** | > 2 joints with pain and signs of inflammation (i.e. tenderness with swelling or effusion) |
| 10 | **4** | **Myositis** | Proximal muscle aching/weakness, associated with elevated creatinine phosphokinase (CK)/aldolase, or EMG changes or a biopsy showing myositis |
| 11 | **4** | **Urinary casts** | Heme-granular or RBC casts |
| 12 | **4** | **Hematuria** | > 5 RBC/high power field. Exclude stone, infection or other cause |
| 13 | **4** | **Proteinuria** | New onset or recent increase of more than 0.5 gm/24 hours |
| 14 | **4** | **Pyuria** | > 5 WBC/high power field. Exclude infection |
| 15 | **2** | **Rash** | Inflammatory type rash |
| 16 | **2** | **Alopecia** | Abnormal, patchy or diffuse loss of hair |
| 17 | **2** | **Mucosal ulcers** | Oral or nasal ulcerations |
| 18 | **2** | **Pleurisy** | Pleuritic chest pain or pleural rub or effusion, or pleural thickening (does not require an objective component if medically convincing) |
| 19 | **2** | **Pericarditis** | Classic pericardial pain and/or rub, effusion or ECG or echocardiogram confirmation (does not require an objective component if medically convincing) |
| 20 | **2** | **Low complement** | Decrease in CH50, C3 or C4 < lower limit of nl for testing laboratory |
| 21 | **2** | **Increased DNA binding** | Increased DNA binding above normal range for testing laboratory |
| 22 | **1** | **Fever** | > 38ºC. Exclude infectious cause |
| 23 | **1** | **Thrombocytopenia** | < 100 x 109 platelets/L, exclude drug causes |
| 24 | **1** | **Leukopenia** | < 3 x 109 WBC/L, exclude drug causes |

**\_\_\_\_\_\_\_\_\_\_\_\_\_\_ Total SCORE**

Republished with permission of Elsevier, from Systemic lupus erythematosus: a companion to Rheumatology, G Tsokos et al., 1st Edition, © 2007; permission conveyed through Copyright Clearance Center, Inc [13].

**References**

1. Harbour R, Miller J. A new system for grading recommendations in evidence based guidelines. BMJ 2001; 323(7308):334-6.
2. Worrall JG, Snaith ML, Batchelor JR, Isenberg DA. SLE: a rheumatological view. Analysis of the clinical features, serology and immunogenetics of 100 SLE patients during long-term follow-up. Q J Med 1990; 74(275):319-30.
3. Pons-Estel BA, Catoggio LJ, Cardiel MH, Soriano ER, Gentiletti S, Villa AR et al. The GLADEL multinational Latin American prospective inception cohort of 1,214 patients with systemic lupus erythematosus: ethnic and disease heterogeneity among "Hispanics". Medicine (Baltimore) 2004; 83(1):1-17.
4. Font J, Cervera R, Ramos-Casals M, Garcia-Carrasco M, Sents J, Herrero C et al. Clusters of clinical and immunologic features in systemic lupus erythematosus: analysis of 600 patients from a single center. Semin Arthritis Rheum 2004; 33(4):217-30.
5. Cervera R, Khamashta MA, Hughes GR. The Euro-lupus project: epidemiology of systemic lupus erythematosus in Europe. Lupus 2009; 18(10):869-74.
6. Lim SS, Bayakly AR, Helmick CG, Gordon C, Easley KA, Drenkard C. The incidence and prevalence of systemic lupus erythematosus, 2002-2004: The Georgia Lupus Registry. Arthritis Rheumatol 2014; 66(2):357-68.
7. Isenberg D. Thirty years, five hundred patients: some lessons learned from running a lupus clinic. Lupus 2010; 19(6):667-74.
8. Yee CS, Cresswell L, Farewell V, Rahman A, Teh LS, Griffiths B et al. Numerical scoring for the BILAG-2004 index. Rheumatology (Oxford) 2010; 49(9):1665-9.
9. UW Medicine . Quick reference card: burns stabilization: rule of nines for adults.. (http://www.uwmedicine.org/airlift-nw/Documents/burn-pocket-card-final.pdf)
10. Levey AS., Bosch JP, Lewis JB, Greene T, Rogers N, Roth D. A more accurate method to estimate glomerular filtration rate from serum creatinine: a new prediction equation. Modification of Diet in Renal Disease Study Group. Ann Intern Med 1999; 130(6): 461-470.
11. Weening JJ, D'Agati VD, Schwartz MM., Seshan SV, Alpers CE., Appel GB, et al. The classification of glomerulonephritis in systemic lupus erythematosus revisited. J Am Soc Nephrol2004; 15(2): 241-250.
12. Gladman DD, Ibanez D, Urowitz MB. Systemic lupus erythematosus disease activity index 2000. J Rheumatol 2002; 29(2):288-91.
13. The SELENA-SLEDAI index. In: Tsokos G, Gordon C, Smolen J, editors. Systemic Lupus Erythematosus: a Companion to Rheumatology. First ed. Elsevier Science and Technology Journals; 2007. Appendix B:525.