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 S 2 (1).

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

Section of guideline	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	**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
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	**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+
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	**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++
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	**D** Evidence level 3 or 4 or Extrapolated evidence from studies rated as 2+
3 Non-analytic studies, e.g. case reports, case series	
4 Expert opinion	

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	**Worral**	**Pons Estel**	**Font**	**Cervera**	**Lim**	**Isenberg**
Year	**1990**	**2004**	**2004**	**2009**	**2014**	**2010**
Reference	**(2)**	**(3)**	**(4)**	**(5)**	**(6)**	**(7)**
Number of patients in cohort studied	(n=100)	(n=1214)	(n=600)	(n=1000)	(n=1156)	(n=500)
Constitutional						
Fever	-	57	42	17	-	-
Weight Loss	-	27	-	-	-	-
Lymphadenopathy	-	15	1	-	-	-
Cutaneous						
Alopecia	27	58	18	-	-	-
Oral/nasal ulcers	36	42	30	13	22	26
Photosensitivity	48	56	41	23	26	35
Malar rash	90[a]	61	54	31	32	62[a]
Discoid rash	90[a]	12	6	8	23	62[a]
Subacute cutaneous	-	3	8	-	-	-
Raynaud's phenomenon	-	28	22	16	-	-
Musculoskeletal						
Arthralgia/Arthritis	94	93	83	48[b]	67[b]	94
Myalgia/myositis	-	18	7	4	-	-
Cardiorespiratory						
Pericarditis	57 [a]	17	28[a]	16[a]	43[a]	43[a]
Pleurisy	57 [a]	22	28[a]	16[a]	43[a]	43[a]
Pneumonitis	-	2	4	-	-	-
Myocarditis	-	3	2	-	-	-
Endocarditis	-	3	8	-	-	-
Neurological						
Seizures	45[c]	8	12[a]	19[a]	14[a]	21[c]
Psychosis	45[c]	4	12[a]	19[a]	14[a]	21[c]
Chorea	45[c]	0.4	0.5	-	-	21[c]
Transverse myelitis	45[c]	0.6	-	-	-	-
Organic brain syndrome	45[c]	2	-	-	-	-
Renal						
Proteinuria/sediment	29	46	34	28	34	31
Nephrotic Syndrome	-	7	-	-	-	-
ESRD	-	2	-	-	7	-

Gastrointestinal						
Ascites	-	1	-	-	-	-
Liver	-	-	0.3	-	-	-
Haematological						
Haemolytic anaemia	-	12	8	5	-	-
Leucopenia	57	42	66	-	75[a]	-
Lymphopenia	81	59	82	-	75[a]	-
Thrombocytopenia	21	19	31	13	-	-
Thrombosis	-	6	7	9	-	-
Serological						
ANA	99	98	99	-	82[d]	95
Anti-dsDNA	55	71	90	-	64[e]	64
Anti-Smith	-	48	13	10	64[e]	13
Anticardiolipin IgG/IgM	34	51/39	-	-	64[e]	21/9
Lupus anticoagulant	19	30	15	-	-	14
Anti-Ro	-	49	23	25	-	37
Anti-RNP	-	51	-	13	-	27
Rheumatoid factor	27	-	12	18	-	25
Low C3	-	49	31	-	-	44
Low C4	-	54	38	-	-	-

[a] combined incidence for items with same value. [b]confirmed arthritis only (usually non-erosive). [c]all neurological features associated with lupus combined. [d]possible failure of ascertainment but patients met ≥4 ACR criteria. [e]combined as met ACR criteria for immunological involvement. - not reported.

***BILAG-2004 Index data collection form*. Date: No:**

- **Only record manifestations/items due to SLE Disease Activity**
- **Assessment refers to manifestations occurring in the last 4 weeks (compared with the previous 4 weeks)**
- **TO BE USED WITH THE GLOSSARY**

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):**

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 m^2 ( ) **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 10^9/l) value ( ) **Y/N***
92. Neutrophils (x 10^9/l) value ( ) **Y/N***
93. Lymphocytes (x 10^9/l) value ( ) **Y/N***
94. Platelets (x 10^9/l) value ( ) **Y/N***
95. TTP ( )
96. Evidence of active haemolysis Yes/No ( )
97. Coombs' test positive (isolated) Yes/No ( )

From Yee et al. Numerical scoring for the BILAG-2004 index Rheumatology (2010) 49 (9): 1665-1669 [8].

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

From Yee et al. Numerical scoring for the BILAG-2004 index Rheumatology (2010) 49 (9): 1665-1669 [8].

(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

From Yee et al. Numerical scoring for the BILAG-2004 index Rheumatology (2010) 49 (9): 1665-1669 [8].

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

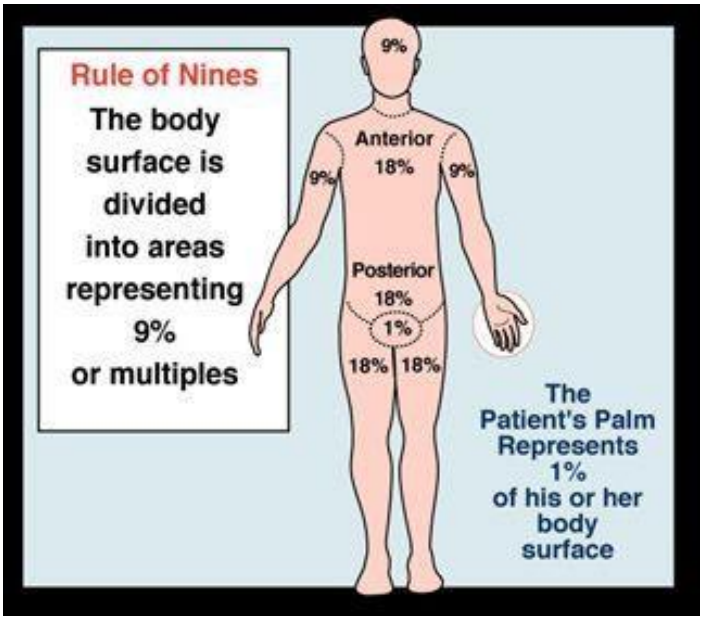
MUCOCUTANEOUS

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



From Yee et al. Numerical scoring for the BILAG-2004 index Rheumatology (2010) 49 (9): 1665-1669 [8].

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)

From Yee et al. Numerical scoring for the BILAG-2004 index Rheumatology (2010) 49 (9): 1665-1669 [8].

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

From Yee et al. Numerical scoring for the BILAG-2004 index Rheumatology (2010) 49 (9): 1665-1669 [8].

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

From Yee et al. Numerical scoring for the BILAG-2004 index Rheumatology (2010) 49 (9): 1665-1669 [8].

	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

From Yee et al. Numerical scoring for the BILAG-2004 index Rheumatology (2010) 49 (9): 1665-1669 [8].

	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

From Yee et al. Numerical scoring for the BILAG-2004 index Rheumatology (2010) 49 (9): 1665-1669 [8].

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

From Yee et al. Numerical scoring for the BILAG-2004 index Rheumatology (2010) 49 (9): 1665-1669 [8].

	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

From Yee et al. Numerical scoring for the BILAG-2004 index Rheumatology (2010) 49 (9): 1665-1669 [8].

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

From Yee et al. Numerical scoring for the BILAG-2004 index Rheumatology (2010) 49 (9): 1665-1669 [8].

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

From Yee et al. Numerical scoring for the BILAG-2004 index Rheumatology (2010) 49 (9): 1665-1669 [8].

	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 m^2 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/mm^3 (μl)) OR haematuria (> 5 RBC/hpf or > 10 RBC/mm^3 (μl)) OR red cell casts

From Yee et al. Numerical scoring for the BILAG-2004 index Rheumatology (2010) 49 (9): 1665-1669 [8].

	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

From Yee et al. Numerical scoring for the BILAG-2004 index Rheumatology (2010) 49 (9): 1665-1669 [8].

These items are required mainly for calculation of GFR

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

From Yee et al. Numerical scoring for the BILAG-2004 index Rheumatology (2010) 49 (9): 1665-1669 [8].

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**

From Yee et al. Numerical scoring for the BILAG-2004 index Rheumatology (2010) 49 (9): 1665-1669 [8].

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

From Yee et al. Numerical scoring for the BILAG-2004 index Rheumatology (2010) 49 (9): 1665-1669 [8].

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

From Yee et al. Numerical scoring for the BILAG-2004 index Rheumatology (2010) 49 (9): 1665-1669 [8].

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

From Yee et al. Numerical scoring for the BILAG-2004 index Rheumatology (2010) 49 (9): 1665-1669 [8].

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

From Yee et al. Numerical scoring for the BILAG-2004 index Rheumatology (2010) 49 (9): 1665-1669 [8].

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

From Yee et al. Numerical scoring for the BILAG-2004 index Rheumatology (2010) 49 (9): 1665-1669 [8].

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

From Yee et al. Numerical scoring for the BILAG-2004 index Rheumatology (2010) 49 (9): 1665-1669 [8].

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

From Yee et al. Numerical scoring for the BILAG-2004 index Rheumatology (2010) 49 (9): 1665-1669 [8].

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 m^2 and having fallen to < 67% of previous value; **or**
(c) GFR < 50 ml/min per 1.73 m^2, and last time was > 50 ml/min per 1.73 m^2 or was not measured.

4. Active urinary sediment
5. Histological evidence of active nephritis within last 3 months
6. Nephrotic syndrome

Category B
One of the following:

1. One of the Category A feature

2. Proteinuria (that has not fulfilled Category A criteria)
(a) urine dipstick which has risen by 1 level to at least 2+ (used only if other methods of urine protein estimation not available); **or**

(b) 24 hour urine protein ≥ 0.5 g that has not decreased (improved) by ≥ 25%; **or**
(c) urine protein-creatinine ratio ≥ 50 mg/mmol that has not decreased (improved) by ≥ 25%; **or**
(d) urine albumin-creatinine ratio ≥ 50 mg/mmol that has not decreased (improved) by ≥ 25%

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

Category C
One of the following:

From Yee et al. Numerical scoring for the BILAG-2004 index Rheumatology (2010) 49 (9): 1665-1669 [8].

1. Mild/Stable proteinuria defined as

(a) urine dipstick ≥ 1+ but has not fulfilled criteria for Category A & B (used only if other methods
of urine protein estimation not available); **or**
(b) 24 hour urine protein > 0.25 g but has not fulfilled criteria for Category A & B ; **or**
(c) urine protein-creatinine ratio > 25 mg/mmol but has not fulfilled criteria for Category A & B; **or**
(d) urine albumin-creatinine ratio > 25 mg/mmol but has not fulfilled criteria for Category A & B

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

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

From Yee et al. Numerical scoring for the BILAG-2004 index Rheumatology (2010) 49 (9): 1665-1669 [8].

HAEMATOLOGICAL

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

Any of the following:

Evidence of haemolysis and Haemoglobin < 8 g/dl
Platelet count < 25 x 10^9/l

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

Any of the following:

Evidence of haemolysis and Haemoglobin 8 - 9.9 g/dl
Haemoglobin < 8 g/dl (without haemolysis)
White cell count < 1.0 x 10^9/l
Neutrophil count < 0.5 x 10^9/l
Platelet count 25 - 49 x 10^9/l

Category C
Any of the following:

Evidence of haemolysis and Haemoglobin ≥ 10g/dl
Haemoglobin 8 - 10.9 g/dl (without haemolysis)
White cell count 1 - 3.9 x 10^9/l
Neutrophil count 0.5 - 1.9 x 10^9/l
Lymphocyte count < 1.0 x 10^9/L
Platelet count 50 - 149 x 10^9/l
Isolated Coombs' test positive

Category D
Previous involvement

Category E
No previous involvement

From Yee et al. Numerical scoring for the BILAG-2004 index Rheumatology (2010) 49 (9): 1665-1669 [8].

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**)

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	$\geq$ 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	> 38^{o}C. Exclude infectious cause
1	Thrombocytopenia	< 100 x 10^9 platelets/L, exclude drug causes
1	Leukopenia	< 3 x 10^9 WBC/L, exclude drug causes

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)

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 10^9 platelets/L, exclude drug causes
24	**1**	**Leukopenia**	< 3 x 10^9 WBC/L, exclude drug causes

_____ Total SCORE

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 Nephrol 2004; 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.