

Appendix 1: BILAG assessment form (version 3)

(All events refer to the previous month unless noted otherwise)

Patient:

Hospital number:

Date of assessment:

Treatment:

Maximum dose in last month or since last visit

GENERAL

Answer: 1) Improving 2) Same 3) Worse 4) New

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

MUCOCUTANEOUS

Answer: 1) Improving 2) Same 3) Worse 4) New

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

NEUROLOGICAL

Answer: 1) Improving 2) Same 3) Worse 4) New

24. Deteriorating level of consciousness ()
25. Acute psychosis or delirium or confusional state ()
26. Seizures ()
27. Stroke or stroke syndrome ()
28. Aseptic meningitis ()
29. Mononeuritis multiplex ()
30. Ascending or transverse myelitis ()
31. Peripheral or cranial neuropathy ()
32. Disc swelling/cyloid bodies ()
33. Chorea ()
34. Cerebellar ataxia ()
35. Headaches – severe unremitting ()
36. Organic depressive illness ()
37. Organic brain syndrome inc. pseudotumour cerebri ()
38. Episodic migrainous headaches ()

MUSCULOSKELETAL

Answer: 1) Improving 2) Same 3) Worse 4) New

39. Definite myositis (Bohan and Peter) ()
40. Severe polyarthritis – with loss of function ()
41. Arthritis ()
42. Tenosynovitis ()
43. Mild chronic myositis ()
44. Arthralgia ()

45. Myalgia ()
46. Tendon contractures and fixed deformity ()
47. Aseptic necrosis ()

CARDIOVASCULAR AND RESPIRATORY

Answer: 1) Improving 2) Same 3) Worse 4) New

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

VASCULITIS

Answer: 1) Improving 2) Same 3) Worse 4) New

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

RENAL

Answer with number (value) or Y/N

68. Systolic BP mmHg ()
69. Diastolic BP (5th phase) ()
70. Accelerated hypertension Y/N
71. Dipstick (– = 1, ++ = 2, +++ = 3) ()
72. 24 h urine protein (g) ()
73. Newly documented proteinuria of >1 g/24 h ()
74. Nephrotic syndrome Y/N
75. Creatinine (plasma/serum) ()
76. Creatinine clearance/CFR (ml/min) ()
77. Active urinary sediment Y/N
78. Histological evidence of active nephritis (within 3 months) ()

HAEMATOLOGY

Answer with number (value) or Y/N

79. Haemoglobin (g/dl) ()
80. Total white cell count $\times 10^9/l$ ()
81. Neutrophils $\times 10^9/l$ ()
82. Lymphocytes $\times 10^9/l$ ()
83. Platelets $\times 10^9/l$ ()
84. Evidence of active haemolysis ()
85. Coombs test positive ()
86. Evidence of circulating anticoagulant ()

SCORE

Gen	Muc	NS	Msk	Car	Vas	Ren	Hae
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Scoring system for the BILAG index (version 3)

It is implicit in this scoring system that all features scored are thought to be due to active lupus. The questionnaire asks whether features are improving, the same, worse or new. If a new feature has developed in the last month (or since the

SLEDAI: DATA COLLECTION SHEET

Chart no.: _____

Date of Visit: _____

M.D.: _____

Patient's Name: _____

(Enter weight in SLEDAI Score column if descriptor present at the time of the visit or in the preceding 10 days.)

Weight	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, disorganized, or catatonic behavior. Exclude uremia 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. Include clouding of consciousness with reduced capacity to focus, and 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 exudate 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 nonresponsive to narcotic analgesia.
8	_____	CVA	New onset of 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	More than 2 joints with pain and signs of inflammation (i.e., tenderness, swelling, or effusion).
4	_____	Myositis	Proximal muscle aching/weakness, associated with elevated creatine phosphokinase/aldolase or electromyogram changes or a biopsy showing myositis.
4	_____	Urinary casts	Heme-granular or red blood cell casts.
4	_____	Hematuria	>5 red blood cells/high power field. Exclude stone, infection, or other cause.
4	_____	Proteinuria	>0.5 gm/24 hours. New onset or recent increase of more than 0.5 gm/24 hours.
4	_____	Pyuria	>5 white blood cells/high power field. Exclude infection.
2	_____	New rash	New onset or recurrence of inflammatory type rash.
2	_____	Alopecia	New onset or recurrence of abnormal, patchy or diffuse loss of hair.
2	_____	Mucosal ulcers	New onset or recurrence of 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 electrocardiogram or echocardiogram confirmation.
2	_____	Low complement	Decrease in CH50, C3, or C4 below the lower limit of normal for testing laboratory.
2	_____	Increased DNA binding	>25% binding by Farr assay or above normal range for testing laboratory.
1	_____	Fever	>38°C. Exclude infectious cause.
1	_____	Thrombocytopenia	<100,000 platelets/mm ³ .
1	_____	Leukopenia	<3,000 white blood cells/mm ³ . Exclude drug causes.
TOTAL SLEDAI SCORE		_____	

Figure 6. Systemic Lupus Erythematosus Disease Activity Index (SLEDAI) form.

cess of making a judgment. We used the former method.

Many of the existing indices of disease activity

have been derived using ad hoc strategies (5), with no particular theoretical base. Two systematic approaches have been used in the literature concerned

SLE ACTIVITY MEASURE (OVER LAST MONTH)		NAME	DATE	RATER	
CONSTITUTIONAL					
	ABSENT or NORMAL	MILD - MODERATE	SEVERE	NOT RECORDED	
1. Weight loss	<input type="checkbox"/> 0	<input type="checkbox"/> 1 <10% body weight	<input type="checkbox"/> 3 >10%	<input type="checkbox"/>	
2. Fatigue	<input type="checkbox"/> 0	<input type="checkbox"/> 1 No limits on activity	<input type="checkbox"/> 3 Functional limitation	<input type="checkbox"/>	
3. Fever	<input type="checkbox"/> 0	<input type="checkbox"/> 1 37.5-38.5°C	<input type="checkbox"/> 3 >38.5°C	<input type="checkbox"/>	
INTEGUMENT					
		MILD	MODERATE	SEVERE	NOT RECORDED
4. Oral/nasal ulcers, or periungual erythema, or malar rash, or photosensitive rash or nail fold infarct	<input type="checkbox"/> 0	<input type="checkbox"/> 1 Present			<input type="checkbox"/>
5. Alopecia	<input type="checkbox"/> 0	<input type="checkbox"/> 1 Hair loss with trauma	<input type="checkbox"/> 2 Spontaneous hair loss		<input type="checkbox"/>
6. Erythematous, maculopapular rash, or discoid lupus, or lupus profundus, or bullous lesions	<input type="checkbox"/> 0	<input type="checkbox"/> 1 <20% Total Body Surface (TBA)	<input type="checkbox"/> 2 20-50% TBA	<input type="checkbox"/> 3 >50% TBA	<input type="checkbox"/>
7. Vasculitis (leucocytoclastic vasculitis, urticaria, palpable purpura, livedo reticularis, ulcer or panniculitis)	<input type="checkbox"/> 0	<input type="checkbox"/> 1 <20% TBA	<input type="checkbox"/> 2 20-50% TBA	<input type="checkbox"/> 3 >50% TBA or necrosis	<input type="checkbox"/>
EYE					
8. Cytoid bodies	<input type="checkbox"/> 0	<input type="checkbox"/> 1 Present	<input type="checkbox"/> 3 visual acuity < 20/200	<input type="checkbox"/>	
9. Hemorrhages (retinal or choroidal) or episcleritis	<input type="checkbox"/> 0	<input type="checkbox"/> 1 Present	<input type="checkbox"/> 3 visual acuity < 20/200	<input type="checkbox"/>	
10. Papillitis or pseudotumor cerebri	<input type="checkbox"/> 0	<input type="checkbox"/> 1 Present	<input type="checkbox"/> 3 visual acuity < 20/200 or field cut	<input type="checkbox"/>	
RETICULOENDOTHELIAL					
	ABSENT or NORMAL	MILD	MODERATE	SEVERE	NOT RECORDED
11. Diffuse lymphadenopathy (cervical, axillary, epitrochlear)	<input type="checkbox"/> 0	<input type="checkbox"/> 1 Shotty	<input type="checkbox"/> 2 >1cm x 1.5cm		<input type="checkbox"/>
12. Hepato- or splenomegaly	<input type="checkbox"/> 0	<input type="checkbox"/> 1 Palpable only with inspiration	<input type="checkbox"/> 2 Palpable without inspiration		<input type="checkbox"/>

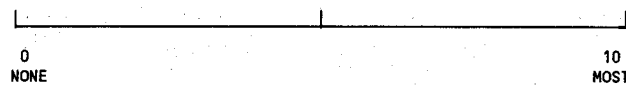
Figure 1. The Systemic Lupus Activity Measure (SLAM). SLE = systemic lupus erythematosus; Diast. = diastolic blood pressure; EKG = electrocardiogram; sx = symptoms; Δ = change; WBC = white blood cells; T = thousand; ESR = erythrocyte sedimentation rate; CrCl = creatinine clearance; RBC = red blood cells; hpf = high power field; 24° = 24 hours.

	ABSENT or NORMAL	MILD	MODERATE	SEVERE	NOT RECORDED
PULMONARY					
13. Pleural effusion/pleurisy	<input type="checkbox"/> 0	<input type="checkbox"/> 1 Shortness of breath or pain only with prompting. Exam normal or near normal.	<input type="checkbox"/> 2 Shortness of breath or pain with exercise, decreased breath sounds and dull lower lobe(s).	<input type="checkbox"/> 3 Shortness of breath or pain at rest, decreased breath sounds and dull middle and lower lobes.	<input type="checkbox"/>
14. Pneumonitis	<input type="checkbox"/> 0	<input type="checkbox"/> 1 X-ray infiltrates only	<input type="checkbox"/> 2 Shortness of breath with exercise	<input type="checkbox"/> 3 Shortness of breath at rest	<input type="checkbox"/>
CARDIOVASCULAR					
15. Raynaud's	<input type="checkbox"/> 0	<input type="checkbox"/> 1 Present			<input type="checkbox"/>
16. Hypertension	<input type="checkbox"/> 0	<input type="checkbox"/> 1 Diast. 90-105	<input type="checkbox"/> 2 Diast. 105-115	<input type="checkbox"/> 3 Diast. >115	<input type="checkbox"/>
17. Carditis	<input type="checkbox"/> 0	<input type="checkbox"/> 1 Pericarditis by EKG &/or RUB &/or effusion by echo; no sx	<input type="checkbox"/> 2 Chest pain or arrhythmia	<input type="checkbox"/> 3 Myocarditis with hemodynamic compromise &/or arrhythmia	<input type="checkbox"/>
GASTROINTESTINAL					
18. Abdominal pain (Serositis, pancreatitis, ischemic bowel, etc.)	<input type="checkbox"/> 0	<input type="checkbox"/> 1 Complaint	<input type="checkbox"/> 2 Limiting Pain	<input type="checkbox"/> 3 Peritoneal signs/ascites	<input type="checkbox"/>
NEUROMOTOR					
19. Stroke syndrome (includes mononeuritis multiplex, transient ischemic attack(TIA),reversible ischemic neurologic deficit (RIND), cerebrovascular accident (CVA), retinal vascular thrombosis)	<input type="checkbox"/> 0	<input type="checkbox"/> 1 Single TIA	<input type="checkbox"/> 2 Multiple TIA/RIND, or mononeuritis multiplex or cranial neuropathy or chorea	<input type="checkbox"/> 3 CVA/myelitis, retinal vascular occlusion	<input type="checkbox"/>
20. Seizure	<input type="checkbox"/> 0	<input type="checkbox"/> 1 1-2/month	<input type="checkbox"/> 2 > 2/month	<input type="checkbox"/> 3 Status epilepticus	<input type="checkbox"/>
21. Cortical dysfunction	<input type="checkbox"/> 0	<input type="checkbox"/> 1 Mild depression/personality disorder or cognitive deficit	<input type="checkbox"/> 2 Δ in sensorium or severe depression or limiting cognitive impairment	<input type="checkbox"/> 3 Psychosis or dementia or coma	<input type="checkbox"/>
22. Headache (including migraine equivalents)	<input type="checkbox"/> 0	<input type="checkbox"/> 1 Symptoms or transient neuro deficit	<input type="checkbox"/> 2 Interferes somewhat with normal activities	<input type="checkbox"/> 3 Incapacitating/aseptic meningitis	<input type="checkbox"/>
23. Myalgia/myositis	<input type="checkbox"/> 0	<input type="checkbox"/> 1 Complaint	<input type="checkbox"/> 2 Limits some activity	<input type="checkbox"/> 3 Incapacitating	<input type="checkbox"/>

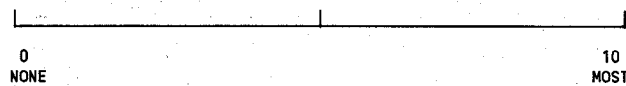
Figure 1. (cont'd)

JOINTS		ABSENT or NORMAL	MILD	MODERATE	SEVERE	NOT RECORDED
24. Joint pain from synovitis and/or tenosynovitis	<input type="checkbox"/> 0	<input type="checkbox"/> 1 Arthralgia only	<input type="checkbox"/> 2 Objective inflammation	<input type="checkbox"/> 3 Limited function	<input type="checkbox"/>	
<hr/>						
OTHER		<input type="checkbox"/> 0	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
25. (Write rules for ascertainment and ad hoc scale)						
<hr/>						
LABORATORY		NORMAL	MILD	MODERATE	SEVERE	UNKNOWN, NOT RECORDED
26. Hematocrit	<input type="checkbox"/> 0 >35	<input type="checkbox"/> 1 30-35	<input type="checkbox"/> 2 25-29.9	<input type="checkbox"/> 3 <25	<input type="checkbox"/>	
27. WBC	<input type="checkbox"/> 0 >3500	<input type="checkbox"/> 1 3500-2000	<input type="checkbox"/> 2 2000-1000	<input type="checkbox"/> 3 <1000	<input type="checkbox"/>	
28. Lymphocyte count	<input type="checkbox"/> 0 1500-4000	<input type="checkbox"/> 1 1499-1000	<input type="checkbox"/> 2 999-500	<input type="checkbox"/> 3 <499	<input type="checkbox"/>	
29. Platelet count	<input type="checkbox"/> 0 >150T	<input type="checkbox"/> 1 100-150T	<input type="checkbox"/> 2 99-50T	<input type="checkbox"/> 3 <50T	<input type="checkbox"/>	
30. ESR (Westergren)	<input type="checkbox"/> 0 <25	<input type="checkbox"/> 1 25-50	<input type="checkbox"/> 2 51-75	<input type="checkbox"/> 3 >75	<input type="checkbox"/>	
31. Serum creatinine or creatinine clearance	<input type="checkbox"/> 0 0.5-1.3mg/dl or 80-100%CrCl	<input type="checkbox"/> 1 1.4-2mg/dl or 79-60%CrCl	<input type="checkbox"/> 2 2.1-4mg/dl or 30-60%CrCl	<input type="checkbox"/> 3 >4mg/dl or <30%CrCl	<input type="checkbox"/>	
32. Urine sediment	<input type="checkbox"/> 0	<input type="checkbox"/> 1 >5 RBC &/or WBC/hpf &/or 0 to 1-3 granular &/or cellular casts/hpf &/or 1-2+ proteinuria &/or <500 mg/L 24 ^o urine protein	<input type="checkbox"/> 2 > 10 RBC &/or WBC/hpf or >3 granular &/or cellular casts/hpf &/or 3 or 4+ &/or 500 mg/L-3.5 g/L 24 ^o urine protein	<input type="checkbox"/> 3 >25 RBC or WBC/hpf &/or Red cell cast &/or >4+ proteinuria &/or >3.5 g/L > 3.5 g/L 24 ^o urine protein	<input type="checkbox"/>	

MD GLOBAL RATING OF SLE ACTIVITY *



PATIENT GLOBAL RATING OF THEIR DISEASE ACTIVITY



* Over last month. Use most active rating during the month.

Assume that "NONE" and "MOST ACTIVE" refer to what patient has experienced.

Figure 1. (cont'd)