<table>
<thead>
<tr>
<th>Term</th>
<th>Description</th>
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<tbody>
<tr>
<td>Non-infectious fever</td>
<td>The objective determination of an elevation of body temperature above the normal range (i.e. 37 ± 1 °C).</td>
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<td>Weight loss</td>
<td>Decrease in body weight that is not voluntary.</td>
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<td>Anorexia</td>
<td>Reduction or loss of appetite or desire for food.</td>
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<td>Butterfly (Malar) Rash</td>
<td>Diffuse or patchy erythema of the malar eminence(s). Lesions may be flat or raised, involving cheeks and/or the bridge of the nose but tending to spare the nasolabial folds; may be unilateral and may involve adjacent or other areas.</td>
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<tr>
<td>Photosensitivity</td>
<td>An unusual skin reaction from exposure to sunlight (typically UV-B). Examples would be persistent erythema, edema, urticaria or vesicular-bullous lesions, located in sun-exposed areas.</td>
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<td>Discoid LE (Discoid rash)</td>
<td>Rash occurring predominantly (but not exclusively) in sun exposed areas and characterized by erythematous, raised patches with adherent keratotic scaling and follicular plugging; atrophic scarring, telangiectasias, hyperpigmentation (peripheral) and hypopigmentation (central) may be present in older lesions.</td>
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<td>Subacute LE</td>
<td>Widespread photosensitive, nonscarring eruptions, either papulosquamos (psoriasiform) or annular.</td>
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<tr>
<td>Alopecia</td>
<td>An abnormal patchy or diffuse loss of hair, particularly scalp hair, non-scarring.</td>
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<td>Purpura</td>
<td>Intracutaneous or subcutaneous hemorrhage as evidenced by red to dark purple areas in the skin.</td>
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<td>Cutaneous vasculitis</td>
<td>Confirmed by skin biopsy or convincing clinical presentation when present on acral sites where biopsy is not feasible.</td>
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<td>Genital ulcers</td>
<td>A break in the skin or mucous membrane found on the penis, scrotum, labia, vestibule or vagina. Lesions may be painful or painless, single or multiple, recurrent or persistent.</td>
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<tr>
<td>Chronic urticaria</td>
<td>A disorder of the superficial skin consisting of well circumscribed discrete wheals with erythematous raised serpiginous borders and blanched centers. It is usually intensely pruritic, and may be localized or generalized.</td>
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<tr>
<td>Teleangiectasias</td>
<td>Visible macular dilatation of superficial cutaneous blood vessels. These blood vessels collaps upon pressure and fill slowly when pressure is released.</td>
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<td>Rheumatoid nodules</td>
<td>Firm, usually painless lumps of variable size found in patients with rheumatoid arthritis. Rheumatoid nodules are commonly foun over areas subject to mechanical trauma (e.g., elbows, heels, walls of olecranon bursa), and occasionally in various internal organs such as lungs and heart.</td>
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<tr>
<td>Panniculitis</td>
<td>Nodular, subcutaneous angitiis with fat-cell necrosis or clinically erythema nodosum.</td>
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<td><strong>Bullae</strong></td>
<td>Vesicular elevation of the cuticle containing transparent watery fluid.</td>
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<td><strong>Periorbital oedema or cyanosis</strong></td>
<td>Violaceous periorbital erythema often with upper eyelid swelling or periorbital oedema.</td>
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<td><strong>Gottron’s sign</strong></td>
<td>Erythematous patches. Scaly hyperemic patches present over the extensor surface of the knuckles (DIP, PIP and MCP). The eruption may have atrophic features as well.</td>
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<td><strong>Livedo reticularis</strong></td>
<td>Reddish/cyanotic reticular discoloration of the skin. Appears on legs, arms, and torso.</td>
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<td><strong>Pitting scars</strong></td>
<td>Digital scarring with loss of substance after acral ulcers.</td>
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<td><strong>Proximal scleroderma</strong></td>
<td>Thickening, tightening, nonpitting induration of the skin of both extremities proximal to the MCP (or MTP) joints and the trunk (anterior chest, abdomen, upper or lower back or flanks).</td>
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<td><strong>Psoriasis</strong></td>
<td>A chronic hyperkeratotic recurrent skin disorder most often characterized by somewhat raised, sharply margined papules or plaques which are scaling and distributed predominantly on the scalp, elbows, knees, chest, umbilicus, back and buttocks. Frequent involvement of the fingernails and toenails is present.</td>
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<td><strong>Morning stiffness</strong></td>
<td>The subjective complaint of localized or generalized lack of easy mobility of the joints upon arising.</td>
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<td><strong>Puffy fingers</strong></td>
<td>A diffuse, usually nonpitting increase in soft tissue mass of the digits extending beyond the normal confines of the joint capsule.</td>
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<td><strong>Polyarthritis</strong></td>
<td>Symmetric involvement of more than three joints with clinical signs of synovitis.</td>
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<td><strong>Oligoarthritis</strong></td>
<td>Clinical signs of synovitis in three or less joints, often asymmetric.</td>
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<td><strong>Axial arthritis</strong></td>
<td>Radiographic signs of sacroiliitis or inflammatory spondylarthropathy.</td>
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<td><strong>Arthralgia</strong></td>
<td>Subjective reporting of pain in the joints.</td>
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<td><strong>Erosions on x-ray</strong></td>
<td>An erosion is a localized area of bone destruction at or near the joint surface.</td>
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<td><strong>Tendinitis</strong></td>
<td>Tenosynovitis determined clinically or by ultrasound.</td>
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<td><strong>Fibromyalgia</strong></td>
<td>Widespread pain and tender points as defined by ACR.</td>
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<td><strong>Myositis</strong></td>
<td>Muscle weakness accompanied by elevated plasma levels of muscle enzymes. The myopathy is further confirmed by muscle biopsy and/or electromyography.</td>
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<td><strong>Keratoconjunctivitis sicca</strong></td>
<td>Confirmed by ophthalmological evaluation using Schirmer test, break up time or Rose Bengal dye.</td>
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<tr>
<td><strong>Scleritis or Episcleritis</strong></td>
<td>Clinical signs of inflammation of the sclera and episclera.</td>
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<tr>
<td><strong>Anterior uveitis</strong></td>
<td>Inflammation of the iris (iritis) or of the iris and the ciliary body (iridocyclitis) is referred to as anterior uveitis and results in photophobia, some decrease in visual acuity, and a variable degree of ocular pain. In contrast to the acute anterior uveitis, the chronic anterior uveitis associated with juvenile rheumatoid arthritis is frequently asymptomatic.</td>
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Posterior uveitis

Inflammation of the choroid usually involves the retina and the term posterior uveitis and chorioretinitis are often used interchangeably.

Retinal vasculitis

Observed by ophthalmoscopy or by a fluorescein angiogram.

Raynaud’s phenomenon

Sudden, reversible "dead white" pallor of an acral structure (e.g., fingers, whole hand, toes, tip of nose, earlobe or tongue), precipitated by cold exposure or emotion.

Venous or arterial thrombosis

Thrombosis should only be recognized when clinical suspicion is confirmed by relevant paraclinical method.

Cerebral infarction

Confirmed by relevant neuroimaging technique (CT, MRI)

Transient ischemic attack

Clinical picture of cerebral infarction with full remission of symptoms within 24 hours.

Avascular bone necrosis

Confirmed by conventional radiography, CT or MRI.

Claudicatio intermittens

Muscle pain (ache, cramp, numbness or sense of fatigue), classically in the calf muscle, which occurs during exercise and is relieved by a short period of rest.

Arterial hypertension

Blood pressure > 140/90 and/or commencement of antihypertensive treatment.

>1 spontaneous abortion

Note time for 2nd spontaneous abortion.

Endocarditis

Non-infectious endocarditis verified by ultrasonography.

Myocarditis

Myocarditis may cause arrhythmias and/or cardiac failure and confirmed by myocardial biopsy.

Pericarditis

Pericardial pain with at least 1 of the following: rub, effusion, or electrocardiogram or echocardiogram confirmation.

Angina pectoris

Severe chest pain due cardiac ischemia without signs of myocardial infarction.

Myocardial infarction

Confirmed by elevated cardiac enzyme levels and electrocardiogram.

Arrythmia

Atrial or ventricular arrhythmias, conduction disturbances documented by electrocardiogram.

Cardiac failure

Includes both right and left ventricular failure.

Pleuritis

Pleuritic chest pain with pleural rub or effusion, or pleural thickening.

Alveolitis / fibrosis

Active inflammatory alveolitis and/or pulmonary fibrosis confirmed by bronchoalveolar lavage, high resolution CT or conventional radiography.

Pulmonary hypertension

Mean pulmonary artery pressure exceeding 15 mmHg calculated by means of echocardiography or measured by cardiac catheterization.

Asthma or COLD

Confirmed by spirometry indicating intermittent or chronic obstructive ventilatory pattern, FEV1/FVC<70% of expected.

Proteinuria

>0.5 gram/24hours.

Hematuria

>5 red blood cells/high power field.

Sterile pyuria

>5 white blood cells/high power field.

Cellular casts

Heme-granular or red blood cell casts.

Glomerulonephritis

verified by renal biopsy.

WHO-class

1: normal/minimal changes
2: mesangioproliferative GN
3: focal, segmental proliferative GN
4: diffuse proliferative GN
5: membraneous GN
6: end-stage GN

Oral or nasal ulcers
Erosions, superficial or deep, of the buccal, labial, lingual, palatal, pharyngeal, or nasal mucosa. They may be painful or painless.

Xerostomia
Oral dryness based on salivary gland destruction documented by sialometry, salivary scintigraphy or salivary gland biopsy.

Sterile peritonitis or ascites
Documented by imaging or puncture.

Intestinal vasculitis
Confirmed by abdominal angiography or histologically.

Autoimmune hepatitis
Exclusion of viral etiology and confirmed by liver biopsy.

Primary biliary cirrhosis
Elevated serum levels of alkaline phosphatase, often anti-mitochondrial antibodies. Confirmed by liver biopsy.

Celiac disease
1) evidence of malabsorption, 2) abnormal jejunal biopsy showing characteristic changes of the villi, and 3) clinical, and serological improvement after institution of a gluten-free diet.

Non-hemolytic anemia
Blood level of hemoglobin below lower normal range without signs of hemolysis. Evaluation of hemolysis may include reticulocyte count, serum levels of LDH, free hemoglobin and haptoglobin.

Immunohemolytic anemia
Blood level of hemoglobin below lower normal range and positive direct antiglobulin (Coombs') test for autoantibodies directed against the rbc membrane antigens.

Leucocytopenia
<3.000 white blood cells x 10^9 / L.

Lymphocytopenia
Below local lower normal range.

Thrombocytopenia
<100.000 platelets x 10^9 / L.

Lymphadenopathy
An enlargement of lymph nodes greater than normal for the particular region examined.

Headache
Includes: Migraine, Tension headache, Cluster headaches, Pseudotumor cerebri (benign intracranial hypertension) and Intractable non-specific headache.

Lupus headache: Severe persistent headache; may be migrainous, but must be non-responsive to narcotic analgesia.

Cognitive dysfunction
The types of cognitiv deficits patients manifest include complex attention, aspects of memory (e.g. learning and recall), visual-spatial processing, language (e.g. verbal), psychomotor speed. Cognitive dysfunction can range from mild impairment to severe dementia. It represents a decline from a previously higher level of functioning and may impede social, educational or occupational functioning. Subjective complaints of cognitive dysfunction are common, although not always objectively verifiable.
Neuropsychological testing is the diagnostic procedure of choice for suspected cognitive dysfunction.

Aseptic meningitis
A clinical syndrome of fever, headache and meningeal irritation with CSF pleocytosis and negative cultures.

Seizures or chorea
Seizures: Abnormal paroxysmal neuronal discharge in the brain causing abnormal function. Seizures may occur with or without the loss of consciousness. Seizures are divided in two groups, partial and generalized. Partial seizures have clinical and electroencephalographic evidence of a focal onset: the abnormal discharge usually arises in a portion of a hemisphere and may spread to the rest of the brain during a seizure. Primary generalized seizures have no interictal evidence on EEG of focal onset. A generalized seizure can be primary or secondary.

Chorea
Chorea consists of irregular, involuntary and jerky movements, that may involve any portion of the body in random sequence. Each movement is brief and unpredictable.

Peripheral neuropathy
Mononeuropathy single/multiplex: Disturbance of the function of one or more peripheral nerve(s). Weakness and paralysis can be due to either conduction block in the motor nerve fibers or to axonal loss. Conduction block is related to demyelinisation with preservation of axon continuity. Remyelinisation can be rapid and complete. If axonal interruption takes place, axonal degeneration occurs below the site of interruption and recovery is often slow and incomplete. Sensory symptoms and sensory loss may affect all modalities or be restricted to certain forms of sensation.

Plexopathy
A disorder of the brachial or lumbosacral plexus producing muscle weakness, sensory deficit and/or reflex change that do not correspond to the territory of a single root or nerve.

Polyneuropathy
Acute or chronic disorder of sensory and motor peripheral nerves with variable tempo characterized by symmetry of symptoms and physical findings in a distal distribution.

Autonomic neuropathy
A disorder of the autonomous nervous system which gives rise to orthostatic hypotension, sphincteric erectile/ejaculatory dysfunction, anhidrosis, heat intolerance, constipation.

Cranial nerve affection
A clinical syndrome affecting the specific sensory and/or motor function of the cranial nerve(s).

Transverse myelopathy
Disorder of the spinal cord characterized by rapidly evolving paraparesis and/or sensory loss, with a demonstrable motor and/or sensory cord level and/or sphincter involvement.

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 two days of the following:
perceptual disturbance, incoherent speech, insomnia or daytime drowsiness, or increased or decreased psychomotor activity.

**Psychosis**
Severe disturbance in the perception of reality characterized by delusions and/or hallucinations.

**Affective disorder**
Prominent or persistent disturbance in mood characterized by either: - Depressed mood or markedly diminished interest or pleasure in almost all activities or – Elevated, expansive or irritable mood.

**Thyreoditis**
Diagnosis supported by thyroid scintigraphy, thyroid hormone status depends on phase of thyreoiditis.

**Type 1 diabetes mellitus**
Immune-mediated diabetes mellitus characterized by young age of onset, normal to wasted body habitus, low to absent plasma insulin, and high suppressible plasma glucagon.

**Amenorrea**
Failure of menarche by age 16 or absence of menstruation for 6 months in a woman with previous periodic menses.

**>2 major infections**
Note time of 3rd infection requiring hospitalization.