## ScantiDNA-study nomenclature

Non-infectious fever The objective determination of an elevation of body

temperature above the normal range (i.e.  $37 \pm 1$  °C).

Weight loss Decrease in body weight that is not voluntary.

Anorexia Reduction or loss of appetite or desire for food.

Butterfly (Malar) Rash 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.

Photosensitivity An unusaul 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.

Discoid LE (Discoid rash)

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.

Subacute LE Widespread photosensitive, nonscarring eruptions, either

papulosquamos (psoriasiform) or annular.

Alopecia An abnormal patchy or diffuse loss of hair, particularly

scalp hair, non-scarring.

Purpura Intracutaneous or subcutaneous hemorrhage as

evidenced by red to dark purple areas in the skin.

Cutaneous vasculitis Confirmed by skin biopsy or convincing clinical

presentation when present on acral sites where biopsy is

not feasible.

Genital ulcers 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.

Chronic urticaria 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. Visible macular dilatation of superficial cutaneous blood

Teleangiectasias Visible macular dilatation of superficial cutaneous blood vessels. These blood vessels collaps upon pressure and

fill slowly when pressure is released.

Rheumatoid nodules 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.

Panniculitis Nodular, subcutaneous angiitis with fat-cell necrosis or

clinically erythema nodosum.

Bullae Vesicular elevation of the cuticle containing transparent

watery fluid.

Periorbital oedema or cyanosis Violaceous periorbital erythema often with upper eye lid

swelling or periorbital oedema.

Gottron's sign 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.

Livedo reticularis Reddish/cyanotic reticular discoloration of the skin.

Appears on legs, arms, adn torso.

Pitting scars

Digital scarring with loss of substance after acral ulcers.

Proximal scleroderma

Thickening, tightening, nonpitting induration of the skin of

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).

Psoriasis A chronic hyperkeratotic recurrent skin disorder most often

characterized by somewhat raised, sharply marginated 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.

Morning stiffness The subjective complaint of localized or generalized lack

of easy mobility of the joints upon arising.

Puffy fingers A diffuse, usually nonpitting increase in soft tissue mass of

the digits extending beyond the normal confines of the

joint capsule.

Polyarthritis Symmetric involvement of more than three joints with

clinical signs of synovitis.

Oligoarthritis Clinical signs of synovitis in three or less joints, often

asymmetric.

Axial arthritis Radiographic signs of sacroiliitis or inflammatory

spondylarthropathy.

Arthralgia Subjective reporting of pain in the joints.

Erosions on x-ray An erosion is a localized area of bone destruction at or

near the joint surface.

Tendinitis

Tenosynovitis determined clinically or by ultrasound.

Fibromyalgia

Widespread pain and tender points as defined by ACR.

Myositis

Muscle weakness accompanied by elevated plasma levels

of muscle enzymes. The myopathy is further confirmed by

muscle biopsy and/or electromyography.

Keratoconjunctivitis sicca Confirmed by ophthalmological evaluation using Schirmer

test, break up time or Rose Bengal dye.

Scleritis or Episcleritis

Anterior uveitis

Clinical signs of inflammation of the sclera and episclera. 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.

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.

Thrombosis should only be recognized when clinical

Venous or arterial thrombosis Thrombosis should only be recognized when clinical

suspicion is confirmed by relevant paraclinical method.

Cerebral infarction

Cerebral infarction

Confirmed by relevant paraclinical metriod.

Cerebral infarction

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.

Transient ischemic attack

Endocarditis Non-infectious endocarditis verified by ultrasonography.
Myocarditis may cause arrythmias 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 arrythmias, 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 Intestinal vasculitis

Documented by imaging or puncture.

Autoimmune hepatitis Primary biliary cirrhosis Confirmed by abdominal angiography or histologically. Exclusion of viral etiology and confirmed by liver biopsy. Elevated serum levels of alkaline phosphatase, often anti-

mitochondrial antibodies. Confirmed by liver biopsy.

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

Celiac disease

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 Lymphocytopenia Thrombocytopenia Lymphadenopathy

<3.000 white blood cells x 10<sup>9</sup>/L. Below local lower normal range. <100.000 platelets x 10<sup>9</sup>/L.

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 consciuosness. 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 consists of irregular, involuntary and jerky

movements, that may involve any portion of the body in random sequence. Each movement is brief and

unpredictable.

Chorea

Peripheral neuropathy Mononeuropathy singel/multiplex: Disturbance of the

function of one or more peripheral nerve(s). Weakness and paralysis can be due to either conduciton block in the motor nerve fibers or to axonal loss. Conduciton block is related to dymyelinisation with preservation of axon continuity. Remyelinisation can be rapid and complete. If axonal interrupeion takes place, axonal degeneration occurs below the site of interruption and recovery is ofetn 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. A disorder of the autonomous nervous system which gives

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 acitivity.

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.

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.