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Classification/Diagnostic Criteria for Common Rheumatologic Conditions

(Excluding vasculitis)

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1. Rheumatoid Arthritis

The 2010 American College of Rheumatology/European League Against Rheumatism classification criteria for Rheumatoid Arthritis (*for newly presenting patients*)

Score

Target population (Who should be tested?): Patients who
1) have at least 1 joint with definite clinical synovitis (swelling)[1]
2) with the synovitis not better explained by another disease[2]

Classification criteria for RA (score-based algorithm: add score of categories A-D; a score of ≥6/10 is needed for classification of a patient as having definite RA)[3]

A. Joint involvement[4]	
1 large joint[5]	0
2−10 large joints	1
1−3 small joints (with or without involvement of large joints)[6]	2
4−10 small joints (with or without involvement of large joints)	3
>10 joints (at least 1 small joint)[7]	5
B. Serology (at least 1 test result is needed for classification)[8]	
Negative RF *and* negative ACPA	0
Low-positive RF *or* low-positive ACPA	2
High-positive RF *or* high-positive ACPA	3
C. Acute-phase reactants (at least 1 test result is needed for classification)[9]	
Normal CRP *and* normal ESR	0
Abnormal CRP *or* normal ESR	1
D. Duration of symptoms[10]	
<6 weeks 0	0
≥6 weeks 1	1

1. The criteria are aimed at classification of newly presenting patients. In addition, patients with erosive disease typical of rheumatoid arthritis (RA) with a history compatible with prior fulfillment of the 2010 criteria should be classified as having RA.
   Patients with long-standing disease, including those whose disease is inactive (with or without treatment) who, based on retrospectively available data, have previously fulfilled the 2010 criteria should be classified as having RA.
2. Differential diagnoses differ in patients with different presentations, but may include conditions such as systemic lupus erythematosus, psoriatic arthritis and gout. If it is unclear about the relevant differential diagnoses to consider, an expert rheumatologist should be consulted.
3. Although patients with a score of less than 6/10 are not classifiable as having RA, their status can be reassessed and the criteria might be fulfilled cumulatively over time.
4. Joint involvement refers to any swollen or tender joint on examination, which may be confirmed by imaging evidence of synovitis. Distal interphalangeal joints, first carpometacarpal joints and first metatarsophalangeal joints are excluded from assessment. Categories of joint distribution are classified according to the location and number of involved joints, with placement into the highest category possible based on the pattern of joint involvement.
5. 'Large joints' refers to shoulders, elbows, hips, knees and ankles.
6. 'Small joints' refers to the metacarpophalangeal joints, proximal interphalangeal joints, second to fifth metatarsophalangeal joints, thumb interphalangeal joints and wrists.
7. In this category, at least one of the involved joints must be a small joint; the other joints can include any combination of large and additional small joints, as well as other joints not specifically listed elsewhere (eg, temporomandibular, acromioclavicular, sternoclavicular, etc.).

8. Negative refers to international unit (IU) values that are less than or equal to the upper limit of normal (ULN) for the laboratory and assay; low-positive refers to IU values that are higher than the ULN but three of less times the ULN for the laboratory and assay; high-positive refers to IU values that are more than three times the ULN for the laboratory and assay. When rheumatoid factor (RF) information is only available as positive or negative, a positive result should be scored as low-positive for RF.
9. Normal/abnormal is determined by local laboratory standards.
10. Duration of symptoms refers to patient self-report of the duration of signs or symptoms of synovitis (eg, pain, swelling, tenderness) of joints that are clinically involved at the time of assessment, regardless of treatment status.

(ACPA: anti-citrullinated protein antibody. CRP: C-reactive protein. ESR: erythrocyte sedimentation rate.)

Daniel Aletaha et al, 2010 Rheumatoid arthritis classification criteria: an American College of Rheumatology/European League Against Rheumatism collaborative initiative. Ann Rheum Dis 2010; 69:1580–1588.

1987 revised criteria for the classification of rheumatoid arthritis (ARA)*

Criterion	Definition
1. Morning stiffness	Morning stiffness in and around the joints, lasting at least 1 hour before maximal improvement
2. Arthritis of 3 or more joint areas	At least 3 joint areas simultaneously have had soft tissue swelling or fluid (not bony overgrowth alone) observed by a physician. The 14 possible areas are right or left PIP, MCP, wrist, elbow, knee, ankle, and MTP joints
3. Arthritis of hand joints	At least 1 area swollen (as defined above) in a wrist, MCP, or PIP Joint
4. Symmetric arthritis	Simultaneous involvement of the same joint areas (as defined in 2) on both sides fo the body (bilateral involvement of PIPs, MCPs, or MTPs is acceptable without absolute symmetry)
5. Rheumatoid nodules	Subcutaneous nodules, over bony prominences, or extensor surfaces, or in juxtaarticular regions, observed by a physician
6. Serum rheumatoid factor	Demonstration of abnormal amounts of serum rheumatoid factor by any method for which the result has been positive in <5% of normal control subjects
7. Radiographic changes	Radiographic changes typical of rheumatoid arthritis on posteroanterior hand and wrist radiographs, which must include erosions or unequivocal bony decalcification localized in or most marked adjacent to the involved joints (osteoarthritis changes alone do not qualify)

* For classification purposes, a patient shall be said to have rheumatoid arthritis if he/she has satisfied at least *4 or these 7* criteria. Criteria 1 through 4 must have been present for *at least 6 weeks*. Patients with 2 clinical diagnoses are not excluded. Designation as classic, definite, or probable rheumatoid arthritis is *not* to be made.

Arnett FC et al. The American Rheumatism Association 1987 revised criteria for the classification of rheumatoid arthritis. Arthritis & Rheumatism 1988; 31:315-24.

2. Spondyloarthritis

European Spondyloarthropathy Study Group (ESSG) criteria

Inflammatory spinal pain ***OR*** **Synovitis: asymmetrical or predominantly in the lower limbs**
AND **One or more of the following:** 1. Positive Family history 2. Psoriasis 3. Inflammatory Bowel Disease 4. Urethritis, cervicitis, or acute diarrhea within one month before arthritis 5. Buttock pain alternating between right and left gluteal areas 6. Enthesopathy 7. Sacroiliitis

Specifications

Variable	Definition
Inflammatory spinal pain*	History or present symptoms of spinal pain in back, dorsal, or cervical region, with at least four of the following: (a) onset before age 45, (b) insidious onset, (c) improved by exercise, (d) associated with morning stiffness, (e) at least 3 months duration
Synovitis	Past or present asymmetric arthritis or arthritis predominantly in the lower limbs
Family history	Presence in first-degree or second-degree relatives of any of the following: (a) ankylosing spondylitis, (b) psoriasis, (c) acute uveitis, (d) reactive arthritis, (e) inflammatory bowel disease
Psoriasis	Past or present psoriasis diagnosed by a doctor
Inflammatory bowel disease	Past or present Crohn disease or ulcerative colitis diagnosed by a doctor and confirmed by radiographic examination or endoscopy
Alternating buttock pain	Past or present pain alternating between the right and left gluteal regions
Enthesopathy	Past or present spontaneous pain or tenderness at examination at the site of the insertion of the Achilles tendon or plantar fascia
Acute diarrhea	Episode of diarrhoea occurring within 1 month before arthritis
Urethritis/cervicitis	Non-gonococcal urethritis or cervicitis occurring within 1 month before arthritis
Sacroiliitis	Bilateral grade 2–4 or unilateral grade 3–4, according to the following radiographic grading system: 0=normal, 1=possible, 2=minimal, 3=moderate and 4=ankylosis

ASAS criteria for classification of axial spondyloarthritis

(To be applied in patients with chronic back pain and age at onset of back pain less than 45 years)

Sacroiliitis on imaging *plus* one or more SpA feature[1] ***OR*** **HLA-B27 *plus* 2 or more other SpA features[2]**	
[1]SpA Features	**[2]Sacroiliitis on Imaging**
1. Inflammatory back pain 2. Arthritis 3. Enthesitis (heel) 4. Uveitis 5. Dactylitis 6. Psoriasis 7. Crohn's/colitis 8. Good response to NSAIDs 9. Family history for SpA 10. HLA-B27 11. Elevated CRP	1. Active (acute) inflammation on MRI highly suggestive of sacroiliitis associated with SpA 2. Definite radiographic sacroiliitis according to modified NY criteria

Specifications

Clinical criterion	Definition
IBP	IBP according to experts: four out of five of the following parameters present: (1) age at onset , 40 years, (2) insidious onset, (3) improvement with exercise, (4) no improvement with rest, (5) pain at night (with improvement upon getting up)
Arthritis	Past or present active synovitis diagnosed by a doctor
Family history	Presence in first-degree or second-degree relatives of any of the following: (a) ankylosing spondylitis, (b) psoriasis, (c) uveitis, (d) reactive arthritis, (e) inflammatory bowel disease
Psoriasis	Past or present psoriasis diagnosed by a doctor
Inflammatory bowel disease	Past or present Crohn disease or ulcerative colitis diagnosed by a doctor
Dactylitis	Past or present dactylitis diagnosed by a doctor
Enthesitis	Heel enthesitis: past or present spontaneous pain or tenderness at examination at the site of the insertion of the Achilles tendon or plantar fascia at the calcaneus
Uveitis anterior	Past or present uveitis anterior, confirmed by an ophthalmologist
Good response to NSAIDs	At 24–48 h after a full dose of NSAID the back pain is not present anymore or much better
HLA-B27	Positive testing according to standard laboratory Techniques
Elevated CRP	CRP above upper normal limit in the presence of back pain, after exclusion of other causes for elevated CRP concentration
Sacroiliitis by X-rays	Bilateral grade 2–4 or unilateral grade 3–4, according to the

	modified New York criteria
Sacroiliitis by MRI	Active inflammatory lesions of sacroiliac joints with definite bone marrow oedema/osteitis suggestive of sacroiliitis associated with spondyloarthritis

J Sieper et al The Assessment of SpondyloArthritis International Society (ASAS) handbook: a guide to assess spondyloarthritis Ann Rheum Dis 2009; 68(Suppl II):ii1–ii44.

Classification criteria for peripheral spondyloarthritis (ASAS) 2010

Arthritis or Enthesitis or Dactylitis
plus 1 or more of: 1. Psoriasis 2. Inflammatory bowel disease 3. Preceding infection 4. HLA-B27 5. Uveitis 6. Sacroiliitis on imaging (radiographs or MRI)
OR
plus 2 or more of the remaining: 1. Arthritis 2. Enthesitis 3. Dactylitis 4. IBP in the past 5. Positive family history for SpA

The criteria are applicable to patients with peripheral arthritis (usually predominantly of the lower limbs and/or asymmetric arthritis), and/or enthesitis, and/or dactylitis.

M Rudwaleit et al The Assessment of SpondyloArthritis International Society classification criteria for peripheral spondyloarthritis and for spondyloarthritis in general Ann Rheum Dis 2011; 70: 25-31

3. Psoriatic Arthritis

*Cl**AS**s*ification *C*riteria for *P*soriatic *AR*thritis (CASPAR) - 2006

To meet the CASPAR criteria, a patient must have inflammatory articular disease (joint, spine, or entheseal) with 3 or more points from the following 5 categories:

1.	Evidence of Psoriasis (one of the three)	Current psoriasis	psoriatic skin or scalp disease present today as judged by a rheumatologist or dermatologist
		Personal history of psoriasis	A history of psoriasis that may be obtained from a patient, family physician, dermatologist, rheumatologist, or other qualified health care provider
		Family history of psoriasis	History of psoriasis in a first- or second-degree relative according to patient report
2.	Psoriatic nail dystrophy	Includes onycholysis, pitting, and hyperkeratosis observed on current physical examination.	
3.	Negative test for rheumatoid factor	By any method except latex but preferably by enzyme-linked immunosorbent assay or nephelometry, according to the local laboratory reference range	
4.	Dactylitis (one of the two)	Current	Swelling of an entire digit
		History	Recorded by a rheumatologist
5.	Radiography	Evidence of juxtaarticular new bone formation, appearing as ill-defined ossification near joint margins (but excluding osteophyte formation) on plain radiographs of the hand or foot.	

William Taylor et al. Classification Criteria for Psoriatic Arthritis Development of New Criteria From a Large International Study Arthritis & Rheumatism 2006; 54(8): 2665–2673

4. Gout

American College of Rheumatology preliminary criteria for the classification of the acute arthritis of primary gout

Gout may be diagnosed if one of the following criteria is present:

1) Monosodium urate crystals in synovial fluid, *or*
2) Tophi confirmed with crystal examination, *or*
3) At least six of the following findings:
    1. Asymmetric swelling within a joint on a radiograph
    2. First metatarsophalangeal joint is tender or swollen (i.e., podagra)
    3. Hyperuricemia
    4. Maximal inflammation developed within one day
    5. Monoarthritis attack
    6. More than one acute arthritis attack
    7. Redness observed over joints
    8. Subcortical cysts without erosions on a radiograph
    9. Suspected tophi
    10. Synovial fluid culture negative for organisms during an acute attack
    11. Unilateral first metatarsophalangeal joint attack
    12. Unilateral tarsal joint attack

Wallace SL et al. Preliminary criteria for the classification of the acute arthritis of primary gout. Arthritis Rheum 1977; 20:896.

5. Osteoarthritis

Radiologic and clinical criteria for hand and knee osteoarthritis (ACR)

Hand Osteoarthritis – Clinical Criteria

Hand pain, aching, or stiffness and 3 or 4 of the following features: 1. Hard tissue enlargement of 2 or more of 10 selected joints 2. Hard tissue enlargement of 2 or more DIP joints 3. Fewer than 3 swollen MCP joints 4. Deformity of at least 1 of 10 selected joints
* Ten selected joints include bilateral second and third distal interphalangeal joints, second and third prximal interphalangeal joints, and first carpometacarpal joint.

Altman R et al. The American College of Rheumatology criteria for the classification and reporting of osteoarthritis of the hand. Arthritis Rheum1990; 33:1601-10.

Knee Osteoarthritis – Criteria

Clinical and laboratory	Clinical and radiographic	Clinical
Knee Pain *Plus at least 5 out of 9:* 1. Age > 50 years 2. Stiffness < 30 minutes 3. Crepitus 4. Bony Tenderness 5. Bony enlargement 6. No palpable warmth 7. ESR <40 mm/hour 8. RF <1:40 9. SF = OA*	**Knee Pain** *Plus at least 1 out of 3:* 1. Age > 50 years 2. Stiffness < 30 minutes 3. Crepitus *Plus* Osteophytes	**Knee Pain** *Plus at least 3out of 6:* 1. Age > 50 years 2. Stiffness < 30 minutes 3. Crepitus 4. Bony Tenderness 5. Bony enlargement 6. No palpable warmth

*SF OA = synovial fluid signs of OA (clear, viscous, or white blood cell count <2,000/mm3)

R. Altman et al. The American College of Rheumatology criteria for the classification and reporting of osteoarthritis of the knee. Arthritis Rheum 1986;29:1039--1049.

6. Systemic Lupus Erythematosus

Clinical and immunologic criteria for Systemic Lupus Erythematosus

The SLICC classification system 2012*

Classify a patient as having SLE if he or she satisfies *4 of the clinical and immunologic criteria* used in the SLICC classification criteria, *including at least one* clinical criterion and one immunologic criterion,

OR

if he or she has *biopsy-proven nephritis* compatible with SLE *in the presence of ANAs or anti-dsDNA*. antibodies.

Clinical criteria

1	Acute cutaneous lupus, including:	Lupus malar rash (do not count if malar discoid) Bullous lupus Toxic epidermal necrolysis variant of SLE Maculopapular lupus rash Photosensitive lupus rash *in the absence of dermatomyositis* *OR* subacute cutaneous lupus (nonindurated psoriaform and/or annular polycyclic lesions that resolve without scarring, although occasionally with postinflammatory dyspigmentation or telangiectasias)
2	Chronic cutaneous lupus, including:	Classic discoid rash • Localized (above the neck) • Generalized (above and below the neck) Hypertrophic (verrucous) lupus Lupus panniculitis (profundus) Mucosal lupus Lupus erythematosus tumidus Chillblains lupus Discoid lupus/lichen planus overlap
3	Oral ulcers	Palate (Buccal, Tongue) OR nasal ulcers *in the absence of other causes, such as vasculitis, Behcet's disease, infection (herpes virus), inflammatory bowel disease, reactive arthritis, and acidic foods*
4	Nonscarring alopecia	diffuse thinning or hair fragility with visible broken hairs *in the absence of other causes such as alopecia areata, drugs, iron deficiency, and androgenic alopecia*
5	Synovitis involving 2 or more joints	characterized by swelling or effusion *OR* tenderness in 2 or more joints and at least 30 minutes of morning stiffness
6	Serositis	Typical pleurisy for more than 1 day *OR* pleural effusions *OR* pleural rub

		Typical pericardial pain (pain with recumbency improved by sitting forward) for more than 1 day *OR* pericardial effusion *OR* pericardial rub *OR* pericarditis by electrocardiography *in the absence of other causes, such as infection, uremia, and Dressler's pericarditis*
7	Renal	Urine protein–to-creatinine ratio (or 24-hour urine protein) representing 500 mg protein/24 hours *OR* red blood cell casts
8	Neurologic	Seizures Psychosis Mononeuritis multiplex *in the absence of other known causes such as primary vasculitis* Myelitis Peripheral or cranial neuropathy *in the absence of other known causes such as primary vasculitis, infection, and diabetes mellitus* Acute confusional state *in the absence of other causes, including toxic/metabolic, uremia, drugs*
9	Hemolytic anemia	
10	Leukopenia OR Lymphopenia	Leukopenia (<4,000/mm^3 at least once) *in the absence of other known causes such as Felty's syndrome, drugs, and portal hypertension* OR Lymphopenia (<1,000/mm^3 at least once) *in the absence of other known causes such as corticosteroids, drugs, and infection*
11	Thrombocytopenia (<100,000/mm^3)	at least once *in the absence of other known causes such as drugs, portal hypertension, and thrombotic thrombocytopenic purpura*

Immunologic criteria

1	ANA level above laboratory reference range
2	Anti-dsDNA antibody level above laboratory reference range (or > 2-fold the reference range if tested by ELISA)
3	Anti-Sm: presence of antibody to Sm nuclear antigen
4	Antiphospholipid antibody positivity as determined by any of the following: Positive test result for lupus anticoagulant False-positive test result for rapid plasma reagin Medium- or high-titer anticardiolipin antibody level (IgA, IgG, or IgM) Positive test result for anti–2-glycoprotein I (IgA, IgG, or IgM)
5	Low complement : Low C3, Low C4, Low CH50
6	Direct Coombs' test *in the absence of hemolytic anemia*

Criteria are cumulative and need not be present concurrently.

SLICC = Systemic Lupus International Collaborating Clinics

Michelle Petri et al. Derivation and Validation of the Systemic Lupus International Collaborating Clinics Classification Criteria for Systemic Lupus Erythematosus. Arthritis & Rheumatism 2012; 64(8):2677–2686

7. Antiphospholipid Antibody Syndrome

Revised classification criteria for the antiphospholipid syndrome

International consensus 2006

Antiphospholipid antibody syndrome (APS) is present if at least *one of the clinical criteria and one of the laboratory criteria* that follow are met. Classification of APS should be avoided if less than 12 weeks or more than 5 years separate the positive aPL test and the clinical manifestation.

Clinical Criteria		
1	Vascular thrombosis[1]	One or more clinical episodes[2] of arterial, venous, or small vessel thrombosis§, in any tissue or organ. Thrombosis must be confirmed by objective validated criteria (i.e. unequivocal findings of appropriate imaging studies or histopathology). For histopathologic confirmation, thrombosis should be present without significant evidence of inflammation in the vessel wall.
2	Pregnancy morbidity	(a) One or more unexplained deaths of a morphologically normal fetus at or beyond the 10th week of gestation, with normal fetal morphology documented by ultrasound or by direct examination of the fetus, *OR* (b) One or more premature births of a morphologically normal neonate before the 34th week of gestation because of: (i) eclampsia or severe preeclampsia defined according to standard definitions, *or* (ii) recognized features of placental insufficiency[3], *OR* (c) Three or more unexplained consecutive spontaneous abortions before the 10th week of gestation, with maternal anatomic or hormonal abnormalities and paternal and maternal chromosomal causes excluded. In studies of populations of patients who have more than one type of pregnancy morbidity, investigators are strongly encouraged to stratify groups of subjects according to a, b, or c above.
Laboratory criteria**		
1	*Lupus anticoagulant* (LA) present in plasma, on two or more occasions at least 12 weeks apart, detected according to the guidelines of the International Society on Thrombosis and Haemostasis (Scientific Subcommittee on LAs/phospholipid-dependent antibodies).	
2	*Anticardiolipin (aCL) antibody* of IgG and/or IgM isotype in serum or plasma, present in medium or high titer (i.e. >40 GPL or MPL, or >the 99th percentile), on two or more occasions, at least 12 weeks apart, measured by a standardized ELISA.	
3	*Anti-b2 glycoprotein-I antibody* of IgG and/or IgM isotype in serum or plasma (in titer >the 99th percentile), present on two or more occasions, at least 12 weeks apart, measured by a standardized ELISA, according to recommended procedures.	

1. Coexisting inherited or acquired factors for thrombosis are not reasons for excluding patients from APS trials. However, two subgroups of APS patients should be recognized, according to: (a) the presence, and (b) the absence of additional risk factors for thrombosis.

Indicative (but not exhaustive) such cases include: age (>55 in men, and >65 in women), and the presence of any of the established risk factors for cardiovascular disease (hypertension, diabetes mellitus, elevated LDL or low HDL cholesterol, cigarette smoking, family history of premature cardiovascular disease, body mass index equal to or more than 30 kg m^2, microalbuminuria, estimated GFR <60 mL min^{-1}, inherited thrombophilia, oral contraceptives, nephrotic syndrome, malignancy, immobilization, and surgery. Thus, patients who fulfil criteria should be stratified according to contributing causes of thrombosis.

2. A thrombotic episode in the past could be considered as a clinical criterion, provided that thrombosis is proved by appropriate diagnostic means and that no alternative diagnosis or cause of thrombosis is found. §Superficial venous thrombosis is not included in the clinical criteria.

3. Generally accepted features of placental insufficiency include: (i) abnormal or non-reassuring fetal surveillance test(s), e.g., a non-reactive non-stress test, suggestive of fetal hypoxemia, (ii) abnormal Doppler flow velocimetry waveform analysis suggestive of fetal hypoxemia, e.g., absent end-diastolic flow in the umbilical artery, (iii) oligohydramnios, e.g., an amniotic fluid index of 5 cm or less, or (iv) a postnatal birth weight less than the 10th percentile for the gestational age.

**Investigators are strongly advised to classify APS patients in studies into one of the following categories: I, more than one laboratory criteria present (any combination); IIa, LA present alone; IIb, aCL antibody present alone; IIc, anti-b2 glycoprotein-I antibody present alone.

Miyakis S et al. International consensus statement on an update of the classification criteria for definite antiphospholipid syndrome (APS). J Thromb Haemost 2006; 4: 295–306.

8. Sjogren's Syndrome

Classification criteria for Sjogren's syndrome (SjS) - 2012

The classification of SjS, which applies to individuals with signs/symptoms that may be suggestive of SjS, will be met in patients who have *at least 2 of the following 3 objective features*:

1. Positive serum anti-SSA/Ro and/or anti-SSB/La or (positive rheumatoid factor and ANA titer 1:320)
2. Labial salivary gland biopsy exhibiting focal lymphocytic sialadenitis with a focus score 1 focus/4 mm^2 (1)
3. Keratoconjunctivitis sicca with ocular staining score 3 (assuming that individual is not currently using daily eye drops for glaucoma and has not had corneal surgery or cosmetic eyelid surgery in the last 5 years) (2)

Prior diagnosis of any of the *following conditions would exclude participation* in SjS studies or therapeutic trials because of overlapping clinical features or interference with criteria tests:

a. History of head and neck radiation treatment
b. Hepatitis C infection
c. Acquired immunodeficiency syndrome
d. Sarcoidosis
e. Amyloidosis
f. Graft versus host disease
g. IgG4-related disease

(1) Using histopathologic definitions and focus score assessment methods as previously described.
(2) Using ocular staining score as previously described.

S. C. Shiboski et al, American College of Rheumatology Classification Criteria for Sjogren's Syndrome: A Data-Driven, Expert Consensus Approach in the Sjogren's International Collaborative Clinical Alliance Cohort. Arthritis Care & Research 2012; 64(4): 475–487

9. Systemic Sclerosis

Criteria for the classification of Systemic Sclerosis (SSc) – ACR/EULAR 2013*

Item	Sub-item	Weight
Bilateral skin thickening of fingers extending proximal to MCP joints		**9**
Skin thickening of the fingers[1]	Puffy fingers	**2**
	Whole Finger, distal to MCP	**4**
Finger tip lesions[1]	Digital Tip Ulcers	**2**
	Pitting Scars	**3**
Telangiectasia		**2**
Abnormal nailfold capillaries		**2**
Pulmonary arterial hypertension and/or interstitial lung disease[2]		**2**
Raynaud's phenomenon		**3**
Scleroderma related antibodies[3]		**3**

* These criteria are applicable to any patient considered for inclusion in an SSc study. The criteria are not applicable to patients with skin thickening sparing the fingers or to patients who have a scleroderma-like disorder that better explains their manifestations (e.g., nephrogenic sclerosing fibrosis, generalized morphea, eosinophilic fasciitis, scleredema diabeticorum, scleromyxedema, erythromyalgia, porphyria, lichen sclerosis, graft-versus-host disease, diabetic cheiroarthropathy).

[1] Count the higher score only. [2]Maximum score is 2. 3Maximum score is 3.

The total score is determined by adding the maximum weight (score) in each category. Patients with a total *score of 9 or more* are classified as having definite SSc.

Definitions of items/subitems

Skin thickening	Skin thickening or hardening not due to scarring after injury, trauma, etc.
Puffy fingers	Swollen digits—a diffuse, usually nonpitting increase in soft tissue mass of the digits extending beyond the normal confines of the joint capsule. Normal digits are tapered distally with the tissues following the contours of the digital bone and joint structures. Swelling of the digits obliterates these contours. Not due to other causes such as inflammatory dactylitis.
Fingertip ulcers or pitting scars	Ulcers or scars distal to or at the proximal interphalangeal joint not thought to be due to trauma. Digital pitting scars are depressed areas at digital tips as a result of ischemia, rather than trauma or exogenous causes.
Telangiectasia	Telangiectasiae are visible macular dilated superficial blood vessels, which collapse upon pressure and fill slowly when pressure is released. Telangiectasiae in a scleroderma-like pattern are round and well demarcated and found on hands, lips, inside of the mouth, and/or are large mat-like telangiectasiae. Distinguishable from rapidly filling spider angiomas with central arteriole and from dilated superficial vessels.
Abnormal nailfold capillary pattern consistent with SSc	Enlarged capillaries and/or capillary loss with or without pericapillary hemorrhages at the nailfold. May also be seen on the cuticle.
Pulmonary arterial hypertension	Pulmonary arterial hypertension diagnosed by right-sided heart catheterization according to standard definitions.
Interstitial lung disease	Pulmonary fibrosis seen on high-resolution computed tomography or chest radiography, most pronounced in the basilar portions of the lungs, or occurrence of “Velcro” crackles on auscultation, not due to another cause such as congestive heart failure.
Raynaud’s phenomenon	Self-reported or reported by a physician, with at least a 2-phase color change in finger(s) and often toe(s) consisting of pallor, cyanosis, and/or reactive hyperemia in response to cold exposure or emotion; usually one phase is pallor.
SSc-related autoantibodies	Anticentromere antibody or centromere pattern seen on antinuclear antibody testing, anti–topoisomerase I antibody (also known as anti–Scl-70 antibody), or anti–RNA polymerase III antibody. Positive according to local laboratory standards.

Frank van den Hoogen, Dinesh Khanna, et al 2013 Classification Criteria for Systemic Sclerosis

An American College of Rheumatology/European League Against Rheumatism Collaborative Initiative.

Arthritis & Rheumatism 2013; xx:xx (to be published in November 2013)

10. Polymyositis and Dermatomyositis

Diagnostic Criteria for Polymyositis and Dermatomyositis (Bohan & Peter)

1	Symmetric proximal muscle weakness with or without dysphagia and respiratory muscle weakness developing over weeks or months
2	Elevation of serum skeletal muscle enzymes (creatine kinase and aldolase), transaminases and lactate dehydrogenase.
3	The electromyographic triad of a. short, small, polyphasic motor unit potentials; b. fibrillations, positive sharp waves, and insertional irritability; and c. bizarre, high frequency repetitive discharges.
4	Muscle biopsy specimen abnormalities of degeneration, regeneration, necrosis, phagocytosis, and interstitial mononuclear infiltrate.
5	Typical skin rash of dermatomyositis, including a heliotrope rash, Gottron's sign, and Gottron's papules.

Interpretation

Diagnosis	**Polymyositis**	**Dermatomyositis**
Definite	4 Criteria	3 – 4 Criteria plus rash
Probable	3 Criteria	2 Criteria plus rash
Pssible	2 Criteria	1 Criterion plus rash

Bohan A, Peter JB. Polymyositis and dermatomyositis. N Engl J Med 1975; 292: 344–47, 403–07.

11. Mixed Connective Tissue Disease

Criteria for Diagnosis of Mixed Connective Tissue Disease (MCTD)

(Alarcon-Segovia)

Clinical Criteria	
1	Swollen hands
2	Acrosclerosis with or without proximal Systemic Sclerosis (SSc)
3	Raynaud's phenomenon
4	Myositis: based on laboratory and/or histological findings
5	Synovitis
Serologic Criterion	
1	Anti-RNP-antibodies with a titer of > 1 : 1600 at the hem-agglutinin assay

Diagnosis of MCTD is based on serologic criterion *plus* 3 out of 5 criteria.

Criteria 4 and 5 are also required to distinguish MCTD from SSc.

Alarcon-Segovia D, Mixed connective tissue disease and overlap syndromes. Clin Dermatol 1994; 12(2):309-16

12. Fibromyalgia

Fibromyalgia Diagnostic Criteria (ACR 2010)

Criteria
A patient satisfies diagnostic criteria for fibromyalgia if the following 3 conditions are met: 1) *Widespread pain index (WPI)*>/=7 and *symptom severity (SS)* scale score >/=5 *or* WPI 3–6 and SS scale score >/=9. 2) Symptoms have been present at a similar level for at least 3 months. 3) The patient does not have a disorder that would otherwise explain the pain.
Ascertainment
1. **WPI**: note the number areas in which the patient has had pain over the last week. In how many areas has the patient had pain? Score will be between 0 and 19. Shoulder girdle (left and right), Upper arm (left and right), Lower arm (left and right), Hip (buttock, trochanter) – left and right, Upper leg (left and right), Lower leg (left and right), Jaw (left and right), Chest, Abdomen, Upper back, Lower back, Neck 2. **SS scale score**: 1. Fatigue 2. Waking unrefreshed 3. Cognitive symptoms. For the each of the 3 symptoms above, indicate the level of severity over the past week using the following scale: 0 = no problem 1 = slight or mild problems, generally mild or intermittent 2 = moderate, considerable problems, often present and/or at a moderate level 3 = severe: pervasive, continuous, life-disturbing problems Considering somatic symptoms in general, indicate whether the patient has: * 0 = no symptoms 1 = few symptoms 2 = a moderate number of symptoms 3 = a great deal of symptoms The SS scale score is the sum of the severity of the 3 symptoms (fatigue, waking unrefreshed, cognitive symptoms) *plus* the extent (severity) of somatic symptoms in general. The final score is between 0 and 12.
* Somatic symptoms that might be considered: muscle pain, irritable bowel syndrome, atigue/tiredness, thinking or remembering problem, muscle weakness, headache, pain/cramps in the abdomen, numbness/tingling, dizziness, insomnia, depression, constipation, pain in the upper abdomen, nausea, nervousness, chest pain, blurred vision, fever, diarrhea, dry mouth, itching, wheezing, Raynaud's phenomenon, hives/welts, ringing in ears, vomiting, heartburn, oral ulcers, loss of/change in taste, seizures, dry eyes, shortness of breath, loss of appetite, rash, sun sensitivity, hearing difficulties, easy bruising, hair loss, frequent urination, painful urination, and bladder spasms.

Frederick Wolfe et al, The American College of Rheumatology Preliminary Diagnostic Criteria for Fibromyalgia and Measurement of Symptom Severity. Arthritis Care & Research 2010; 62(5): 600–610

13. Joint Hypermobility

Beighton score for joint hypermobility syndrome

One point is gained for each side of the body for the first four maneuvers listed below, such that the hypermobility score is a maximum of 9 if all are positive.

1	Passive dorsiflexion of the fifth metacarpophalangeal joint to 90° or more
2	Opposition of the thumb to the volar aspect of the ipsilateral forearm
3	Hyperextension of the elbow to 10° or more
4	Hyperextension of the knee to 10° or more
5	Placing of hands flat on the floor without bending the knees (1 point only)

1998 Brighton criteria for classification of joint hypermobility syndrome

Joint hypermobility syndrome is diagnosed in the presence of:

1. 2 major criteria;
2. 1 major criterion *plus* 2 minor criteria; *or*
3. 4 minor criteria.

Two minor criteria will suffice where there is an unequivocally affected first degree relative.

The syndrome is excluded by the presence of Marfan's or Ehlers-Danlos syndromes

Major criteria	
1	Beighton score of ≥4 (either currently or previously)
2	Arthralgia for longer than three months in four or more joints
Minor criteria	
1	Beighton score of 1, 2, or 3 (0, 1, 2, or 3 if aged >50 years)
2	Arthralgia in 1-3 joints or back pain or spondylosis, spondylolysis and/or spondylolisthesis
3	Dislocation in more than 1 joint or in one joint on more than one occasion
4	Three or more soft tissue lesions (e.g., epicondylitis, tenosynovitis, bursitis)
5	Marfanoid habitus (tall, slim, ratio of span to height greater than 1.03 and/or ratio of upper segment to lower segment less than 0.89, arachnodactyly)
6	Abnormal skin: striae, hyperextensibility, thin skin, papyraceous scarring
7	Eye signs: drooping eyelids, myopia, or antimongoloid slant
8	Varicose veins, hernia, or uterine or rectal prolapse
9	Mitral valve prolapse

Beighton P et al. Articular mobility in an African population. Ann Rheum Dis 1973; 32:413-8.

Grahame R, Bird HA, Child A. The revised (Brighton 1998) criteria for the diagnosis of benign joint hypermobility syndrome (BJHS). J Rheumatol 2000;27:1777-9.