

Systemic Sclerosis Classification Criteria

One sufficient criterion plus a 7-item weighted scoring system for classification of systemic sclerosis, validated against the 1980 ARA and 2001 LeRoy/Medsger criteria.

2013 ACR-EULAR Criteria
Classification Criteria for Systemic Sclerosis
van den Hoogen et al. • Arthritis Rheum
2013;65:2737–2747

NOTE

Classification (not diagnostic) criteria – applicable to any patient considered for a SSc study; not applicable to skin thickening sparing the fingers or SSc-like disorders (see below).

1 Applicability & Sufficient Criterion

Not applicable to patients with skin thickening sparing the fingers, or with an SSc-like disorder better explaining findings: nephrogenic sclerosing fibrosis, generalized morphea, eosinophilic fasciitis, scleredema diabeticorum, scleromyxedema, erythromyalgia, porphyria, lichen sclerosus, graft-versus-host disease, diabetic cheiropathy.

Sufficient criterion – classify as SSc without further scoring if: **skin thickening of the fingers of both hands extending proximal to the metacarpophalangeal (MCP) joints** is present.

2 Weighted Items (if Sufficient Criterion Not Met)

ITEM	WEIGHT
Skin thickening of the fingers – puffy fingers (2) or sclerodactyly distal to MCP but proximal to PIP (4) – highest score only	2–4 pts
Fingertip lesions – digital tip ulcers (2) or fingertip pitting scars (3) – highest score only	2–3 pts
Telangiectasia	2 pts
Abnormal nailfold capillaries	2 pts
Pulmonary arterial hypertension and/or interstitial lung disease (max score 2 regardless of one or both present)	2 pts
Raynaud's phenomenon	3 pts
SSc-related autoantibodies – anti-centromere, anti-topoisomerase I (anti-Scl-70), or anti-RNA polymerase III (max score 3 regardless of number positive)	3 pts

3 Scoring, Threshold & Test Performance

Scoring: add the maximum weight in each item category (only the highest sub-item score counts per category) to calculate the total score. A total score ≥ 9 classifies the patient as having **definite systemic sclerosis**.

Performance (validation sample, N=405): sensitivity 91% (95% CI 87–94%), specificity 92% (95% CI 86–96%) – outperforming the 1980 ARA criteria (75%/72%) and 2001 LeRoy/Medsger criteria (75%/78%) in the same cohort.

Definitions: PAH = proven by right heart catheterization. ILD = pulmonary fibrosis on HRCT/chest radiograph, most pronounced at lung bases, or 'velcro' crackles on auscultation not due to another cause (e.g. CHF).