New Classification Criteria for Systemic Sclerosis (Scleroderma)

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KEYWORDS

• Systemic sclerosis • Scleroderma • Classification criteria • Diagnosis • Validation • Performance • Sensitivity • Specificity

KEY POINTS

- The American College of Rheumatology/European League Against Rheumatism classification criteria for systemic sclerosis (SSc) are more sensitive and specific than the previous 1980 American Rheumatism Association criteria for SSc.
- Classification criteria are not diagnostic criteria and were developed for clinical studies, but most patients diagnosed with SSc should meet classification criteria.
- There is no gold standard for SSc diagnosis and the criteria should only be applied if SSc diagnosis is suspected.

INTRODUCTION

Systemic sclerosis (SSc; also called scleroderma) refers to an autoimmune connective tissue disease with autoantibodies, vasculopathy, and fibrosis.[1] Most patients have sclerodactyly (thickened skin of the fingers) and may or may not have more extensive

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skin fibrosis. There are large differences in prevalence of internal organ involvement and other features, although nearly all have Raynaud phenomenon (RP), many have esophageal dysmotility, and 8% to 50% have pulmonary arterial hypertension (PAH), cardiac involvement, interstitial lung disease, inflammatory arthritis, and digital ulcers.[2] There is no laboratory test to diagnose SSc. With heterogeneous features and no diagnostic gold standard, it is important to classify patients with SSc along the disease spectrum as accurately as possible. Because SSc is rare, if a significant proportion of patients are not included in the classification then they will be missed when studying the disease.

Classification criteria are not diagnostic criteria. However, classification criteria should be similar to diagnostic criteria (with a large overlap of patients included in both criteria).[3] This article discusses the evolution of SSc classification criteria, strengths and weaknesses, and how to use the new American College of Rheumatology (ACR)/European League Against Rheumatism (EULAR) SSc classification criteria.

PREVIOUS SYSTEMIC SCLEROSIS CLASSIFICATION CRITERIA

Classification criteria have been published in SSc.[4–7] Classification criteria are used to include patients with a similar clinical entity for research studies,[3] but are used in a clinical setting to help to classify patients. The American Rheumatism Association (ARA) 1980 preliminary scleroderma (SSc) criteria were the most commonly cited criteria for SSc.[4,5,8] The 1980 preliminary scleroderma criteria were an important step in SSc classification.[4,5] There was 1 sufficient (major) criterion and 3 minor criteria. The major (sufficient) criterion was proximal cutaneous sclerosis/skin thickening (nonpitting) of the fingers that also extended proximal to the metacarpophalangeal joints (MCPs). If this criterion was not met, then at least 2 of 3 items had to be present: (1) sclerodactyly, (2) digital pitting scars of fingertips or loss of substance of the distal finger pad (digital tuft resorption), and (3) bibasilar pulmonary fibrosis.[4,5]

LIMITATIONS OF PREVIOUS CRITERIA

There are currently more patients with SSc who are at the mild end of the spectrum and in the limited cutaneous SSc subset (lcSSc) compared with patients in whom the 1980 criteria were tested, in whom a larger proportion of patients with diffuse cutaneous SSc (dcSSc) were identified; possibly because of the evolution of SSc over time, more recognition caused by available commercial autoantibodies, and/or earlier diagnosis. Many patients whom experts would now classify as SSc were not classified in the 1980 criteria. For example, a patient with sclerodactyly, telangiectasia, calcinosis, RP, proven PAH, and a positive anticentromere antibody test would not be classified as having SSc in the 1980 scheme. Patients with sclerodactyly, RP, anticentromere antibodies, dysphagia, dilated nail fold capillaries, and calcinosis would also not meet the previous criteria for SSc classification.

Often 20% of patients with lcSSc did not meet the criteria and were excluded from clinical studies.[1,9–11] This percentage means that 1 in 5 patients with lcSSc would not be classified with SSc even though they had a diagnosis of SSc.

OTHER PREVIOUSLY PROPOSED SYSTEMIC SCLEROSIS CLASSIFICATION CRITERIA

LeRoy and colleagues[12] proposed criteria that included clinical features, autoantibodies, and capillaroscopy. In 2001, LeRoy and Medsger[7] suggested revising the classification criteria to include early cases of SSc, using nail fold capillary changes