Updated systemic sclerosis criteria improve disease classification

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New classification criteria for systemic sclerosis have just been published and are more sensitive than the 1980 criteria, enabling earlier identification and treatment of this disabling autoimmune disease. The 2013 criteria, developed by a joint committee commissioned by the American College of Rheumatology (ACR) and European League Against Rheumatism (EULAR), are published in the ACR journal, *Arthritis & Rheumatism*.

Systemic sclerosis, also known as scleroderma, is a connective tissue disease that is characterized by sclerodermatous skin changes–a hardening of tissue due to increased collagen deposits; Raynaud's phenomenon–spasms of small blood vessels in response to cold or stress that cause color changes in fingers or toes, to obliteration of blood vessels (vasculopathy) leading to tissue death; and internal organ fibrosis–formation of excess tissue that scars organs. The ACR estimates that systemic sclerosis affects 49,000 U.S. adults.

The joint ACR-EULAR committee was led by Dr. Janet Pope from Western University, St. Joseph's Health Care London in Ontario, Canada, and Dr. Frank van den Hoogen from St. Maartenskliniek in The Netherlands. The committee's intent was to improve the classification of systemic sclerosis by clustering items and simplifying the weighting of the different criteria. The new criteria set was tested for specificity and sensitivity by comparing scleroderma cases with controls (patients with disorders similar to scleroderma), and validated by experts viewing cases with and without the disease.

"There is a need for improved classification criteria for systemic sclerosis," explains Dr. van den Hoogen. "The 1980 ACR criteria were not sensitive enough to identify patients with early disease or limited cutaneous system sclerosis. Our efforts with the joint committee addressed this sensitivity issue, resulting in the 2013 classification criteria for systemic sclerosis."

Based on the new criteria, a patient with thickening of the skin in the middle part of the fingers (from proximal to the metacarpophalangeal joints) would be classified as having systemic sclerosis, regardless of other features. If this criterion was not met, however, then seven items with varying weights would need to be assessed in order to obtain a scleroderma classification: skin thickening of the fingers, fingertip lesions, telangiectasia, abnormal nailfold capillaries, pulmonary arterial hypertension and/or interstitial lung disease, Raynaud's phenomenon, and SSc-related antibodies.

The results of the validation testing show that sensitivity and specificity were both greater than 90% for the 2013 systemic sclerosis classification criteria compared to 75% for the 1980 ACR criteria. "The new systemic sclerosis classification criteria should correctly classify more patients with the disease," concludes Dr. Pope. "Criteria that are more specific will allow for earlier identification and better treatment for those with systemic sclerosis."

More information: "2013 Classification Criteria for Systemic Sclerosis. An American College of Rheumatology and European League Against Rheumatism Collaborative Initiative" Frank van den Hoogen, Dinesh Khanna, Jaap Fransen, Sindhu R. Johnson, Murray Baron, Alan Tyndall, Marco Matucci-Cerinic, Raymond P. Naden, Thomas A. Medsger Jr., Patricia E. Carreira, Gabriela Riemekasten, Philip J. Clements, Christopher P. Denton, Oliver Distler, Yannick Allanore, Daniel E. Furst, Armando Gabrielli, Maureen D. Mayes, Jacob M. van Laar, James R. Seibold, Laszlo Czirjak, Virginia D. Steen, Murat Inanc, Otylia Kowal-Bielecka, Ulf Mu¨ller-Ladner, Gabriele Valentini, Douglas J. Veale, Madelon C. Vonk, Ulrich A. Walker, Lorinda Chung, David H. Collier, Mary Ellen Csuka, Barry J. Fessler, Serena Guiducci, Ariane Herrick, Vivien M. Hsu, Sergio

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