Letter

The Challenge of Very Early Systemic Sclerosis

To the Editor:

We read with great interest the article by Blaja, *et al*¹ on the challenge of very early systemic sclerosis (SSc). We have been addressing this topic for some years. Actually, VEDOSS (very early diagnosis of systemic sclerosis)², subsequently renamed very early SSc³, was proposed before the development of the 2013 American College of Rheumatology/European League Against Rheumatism (ACR/EULAR) classification criteria⁴. The careful analysis of these criteria prompted us to emphasize the need to clearly define the boundaries of the condition⁵. In addition, the subsequent detection of an evolution into SSc satisfying criteria^{6,9} in only 50% of strictly defined very early/early SSc led to the proposed term *undifferentiated connective tissue disease at risk for SSc* (UCTD-risk-SSc)⁶.

Defining which of the patients satisfying VEDOSS² entry criteria already fulfill SSc classification criteria requires a careful assessment of each patient by history, physical examination, and physiologic and imaging investigations^{5,6}.

Predicting the evolution into definite SSc by those strictly labeled UCTD-risk-SSc has long been considered an accomplished task⁷. In that regard, based on the disease course of 102 patients, we have developed a weighted score (Table 1)⁸. By receiver-operation characteristic curve analysis, patients with a score at admission ≥ 2.75 evolved into SSc, with a sensitivity of 91.3% and a 73.2% specificity.

The analysis of disease evolution in the VEDOSS cohort patients who at enrollment did not fulfill ACR/EULAR criteria for SSc and did not present any manifestation consistent with SSc sine scleroderma⁹ might validate or disprove the use of the criteria.

Antonella Riccardi¹ , MD
Antonella Marcocchia² , MD
Gabriele Valentini¹ , MD

¹Rheumatology Unit, Department of Precision Medicine,
University of Campania Luigi Vanvitelli, Naples;

²Centro di Riferimento Interdisciplinare, Interdipartimentale
per la diagnosi precoce della Sclerodermia, Sandro Pertini Hospital,
Rome, Italy.

Address correspondence to Dr. A. Riccardi, Rheumatology Unit,
Department of Precision Medicine, University of Campania Luigi
Vanvitelli, Via Pansini 5, 80131 Naples, Italy.
Email: antonellariccardi84@gmail.com.

Table 1. Variables independently predictive of development of SSc based on multivariate regression. Relative weight in a 10-point score.

Variable	HR	95% CI	p	Weight
Anti-Scl-70	11.57	3.12–42.88	0.0003	3.00
ANA $\geq 1:320$	6.93	1.81–26.51	0.0049	2.75
Avascular areas	5.03	1.36–18.59	0.0158	2.25
ACA	3.92	1.13–13.59	0.0319	2.00

From Riccardi, *et al*.⁸ SSc: systemic sclerosis; anti-Scl-70: anti-DNA topoisomerase I; ANA: antinuclear antibody; ACA: anticentromere antibodies.

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