## 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 SSc[3], was proposed before the development of the 2013 American College of Rheumatology/European League Against Rheumatism (ACR/EULAR) classification criteria[4]. The careful analysis of these criteria prompted us to emphasize the need to clearly define the boundaries of the condition[5]. In addition, the subsequent detection of an evolution into SSc satisfying criteria[4,9] 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 VEDOSS[2] 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 task[7]. 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 scleroderma[9] might validate or disprove the use of the criteria.

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*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 ≥ 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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