Table 1. Independent predictive variables in multivariate regression analysis and the resulting weighted prediction model *

Variable	β	HR	95% CI	P	Weight
Anti-Sc170	2.9553	19.21	4.87-75.76	<0.001	3.25
Cs $\leq$ 5/mm	1.9093	6.75	2.07-22.00	0.001	2
ANA $\geq$ 1:320	1.7402	5.70	1.42-22.85	0.01	2
ACA	1.6740	5.33	1.51-1.90	0.01	1.75
GC $>$ 5	1.0049	2.73	1.44-5.17	0.002	1

* β : regression coefficients; HR: hazard ratio; 95% CI: 95% confidence interval; Cs: Capillaries; ANA: anti-nuclear antibodies; ACA: anti-centromere antibodies; GC: giant capillaries

Conclusion: Assessing the mean number of capillaries/mm and the total number of giant capillaries instead of avascular areas at videocapillaroscopy, resulted in improving the sensitivity and specificity of the score recently developed to predict the evolution of UCTD-risk-SSc into definite SSc.

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Figure 1. ROC analysis identify the score with the highest sensitivity and specificity. A score 3.25 predicted evolution to SSc with 93.5% sensitivity and 75% specificity.

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SAT0341

PREDICTORS TO PROGRESSION TO SYSTEMIC SCLEROSIS IN A GROUP OF SECONDARY RAYNAUD PHENOMENON OBSERVED IN A LARGE SINGLE BRAZILIAN COHORT

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Background: The 2013 ACR/EULAR classification criteria for systemic sclerosis (SSc) allowed the inclusion of a subset of patients without skin involvement, emphasizing the need of early diagnosis of SSc.

Objectives: In this setting, the objective of this study was to classify, according to ACR/EULAR SSc criteria, patients with Raynaud phenomenon (RP) and at least one SSc manifestation, except for skin involvement, and analyze predictors for development of SSc.

Methods: This is a cross-sectional single-center analysis of consecutive patients presenting RP and a characteristic SSc manifestation (SSc autoanti-body, SSc visceral involvement or SD pattern at nailfold capillaroscopy - NFC), without skin involvement, who attended a scleroderma outpatient clinic between 2010 and 2019. Patients were classified as SSc (according to ACR/EULAR SSc criteria) or undifferentiated connective tissue disease (UCTD) in their last medical visit and compared. Additionally, a longitudinal retrospective analysis of both groups was performed to find predictors for development of SSc. Data were obtained from an electronic register database. Statistical significance was set up as $p < 0.05$.

Results: Among 217 patients, 153 (70.5%) were classified as SSc, at the last medical visit, including 65 (30%) after the first investigation. During a com-
parable median follow-up [4.9 $\pm$ 3.8 vs. 4.6 $\pm$ 3.0 years; $p=0.90$], patients with SSc presented more frequently puffy fingers (PF: 73% vs. 6.3%; $p < 0.01$), pitting scars (87.4% vs. 4.7%; $p < 0.01$), digital ulcers (25.2% vs. 1.6%; $p < 0.01$), telangiectasias (52.3% vs. 7.8%; $p < 0.01$), interstitial lung disease (ILD: 66.7% vs. 34.4%; $p < 0.01$), pulmonary hypertension (PH: 18.3% vs. 7.9%; $p=0.049$) and esophageal dysmotility (ED: 58.9% vs. 43.8%; $p=0.02$) than secondary RP patients without SSc. Moreover, SSc patients had more SD pattern at NFC (83.4% vs. 57.8%; $p < 0.01$), anti-Sc170 antibody (16.6% vs. 3.1%; $p < 0.01$), and nucleolar ANA pattern (22.5% vs. 10.9%; $p=0.048$). Furthermore, SSc patients were more frequently treated with immunosuppressive therapy (80.1% vs. 37.5%; $p < 0.01$) and presented an increased death rate (9.9% vs. 1.6%; $p < 0.01$ and Figure 1). For the analysis of the predictors to SSc, patients who had already fulfilled SSc criteria at base-line were excluded and 152 patients were analysed: 88 progressed to SSc and 64 remained as UCTD. After multivariate analyses, puffy fingers (OR=4.48; 95%CI 1.17-17.17; $p=0.029$) and a higher SSc score, both at baseline, (OR: 1.97; 95%CI 1.33-2.92; $p < 0.001$) were predictors to SSc, while esophageal symp-toms (OR=0.08; 95%CI 0.05-0.67; $p=0.01$) and absence of clinical symptoms (OR=0.08; 95%CI 0.01-0.40; $p=0.002$) were negative predictive factors. When analyzing the combination of SSc features, RP + PF + positive ANA (OR=15.5; 95%CI 4.5-53.1; $p < 0.001$), RP + PF + positive ANA + SD-NFC (OR=12.3; 95%CI 2.8-54.2; $p < 0.001$), RP + PF + SD-NFC (OR=12; 95%CI 2.79-54.1; $p < 0.01$), and RP + lung disease (ILD/HAP) + SD-NFC (OR=3.0; 95%CI 1.1-10.5; $p < 0.04$) were associated with classification as SSc, while combinations without non-RP clinical symptoms, as RP + SD-NFC (OR=0.027; 95%CI 0.003-0.210; $p < 0.001$) and RP + anticentromere + SD-NFC (OR=0.050; 95%CI 0.007-0.440; $p < 0.001$) were not associated with SSc.

Conclusion: Among patients with RP and SSc features, but without skin involve-
ment, those with SSc had a more severe disease with worse prognosis than UCTD with RP. Puffy fingers and a higher baseline SSc score, especially in com-binations including PF or lung disease were predictors of SSc, reinforcing the strength of the ACR/EULAR classification criteria and the presence of specific non-RP symptoms.

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SAT0342

INNATE LYMPHOID CELLS ARE PREDICTORS OF DISEASE PROGRESSION IN SYSTEMIC SCLEROSIS: A 3-YEARS FOLLOW-UP STUDY

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Background: Activation of the immune system is a characteristic feature of SSc. Numerous studies have suggested that type 2 are key drivers of progressive fibrosis. Recently, innate lymphoid cells (ILC) are emerging as an important cellular source of type 2 cytokines triggering fibrotic tissue remodeling independently of the adaptive immune system. Increased levels of ILC2 were found in patients with SSc. However, the contributive role of ILC2 in pathogenesis of SSc is not completely understood.