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SAT0273 PREDICTIVE VALUE OF THE REVISED EUROPEAN SCLERODERMA TRIALS AND RESEARCH GROUP ACTIVITY INDEX (EUSTAR-AI)

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Background: Disease activity as measured by the European Scleroderma Study Group Activity Index (EScSG-AI) (1) has been recently found to predict the development of damage over time in an early systemic sclerosis (SSc) cohort (2). The European Scleroderma Trials and Research Group Task Force for the Development of Revised Activity Criteria for SSc recently succeeded in identifying a preliminarily revised activity index (EUSTAR-AI) that had a greater construct validity than the EScSG-AI i.e. performing better in identifying patients with active disease (3).

Objectives: To assess in patients with SSc the predictive value of the EUSTAR-AI for disease severity accrual.

Methods: SSc patients from the EUSTAR database with a disease duration from the onset of the first non-Raynaud sign/symptom ≤ 5 years were first extracted. Patients were considered for the study if they presented the following features: a) availability of items included in the EUSTAR-AI, in the EScSG-AI and in the Medsger severity scale at baseline and yearly for 2 consecutive years; b) availability of vital status at the last observation; c) availability of other disease features known to predict disease progression (male sex, clinical and serological subset).

To capture the disease activity variations over time, we calculated the EUSTAR-AI and EScSG-AI adjusted mean (area under the curve over time divided by time interval). Disease progression was based on the Medsger severity scale and included accrual of a $\Delta \geq 1$ summed severity score and of a $\Delta \geq 1$ severity in each organ systems at the 2 year follow-up visit compared with the initial visit. To explore specific determinants of disease progression, logistic regression analysis was carried out.

Results: A total of 549 patients satisfied the entry criteria. At univariate logistic regression analysis (among sex, age, clinical and serological subset, EScSG-AI and EUSTAR-AI adjusted mean and baseline severity score), EScSG-AI adjusted mean (OR 1.41 95%CI 1.20-1.67), antiScl70 antibody positivity (OR 1.72 95%CI 1.20-2.47), diffuse subset (OR:1.46 95%CI 1.01-2.10) and EUSTAR-AI adjusted mean (OR 1.41 95%CI 1.23-1.61) predicted disease severity accrual. Multivariate analysis revealed that the EUSTAR-AI adjusted mean was the best predictor of disease progression (Table1). Moreover, at multivariate analysis the EUSTAR-AI adjusted mean also predicted severity accrual of lung (OR 1.32), heart (OR 1.40), skin (OR 1.48) and peripheral vascular disease (OR 1.45).

Conclusion: The adjusted mean EUSTAR-AI has a distinct predictive value for disease progression and development of severe organ involvement in SSc and works better than EScSG-AI.

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Table 1.. Predictive features of severity progression by multivariate regression analysis

Predictive factors	OR (IC 95%)	severity deterioration	p- value
Adjusted mean EUSTAR-AI	1.41 (1.23-1.61)	Δ summed severity score ≥ 1	<0,0001
Adjusted mean EUSTAR-AI	1.32 (1.14-1.54)	Δ lung severity score ≥ 1	0,0002
Adjusted mean EUSTAR - AI age	1.40 (1.15-1.70)	Δ heart severity score ≥ 1	0,0005 0,0223
	1.03 (1.01-1.05)		
Adjusted mean EUSTAR-AI age	1.48 (1.21-1.82)	Δ Skin severity score ≥ 1	0,0002 0,0359
	0.97 (0.95-0.99)		
Adjusted mean EUSTAR-AI	1.45 (1.24-1.70)	Δ Peripheral Vascular severity score ≥ 1	<0,0001

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SAT0274 THE EFFECT OF RITUXIMAB ON LUNG FUNCTION AND SKIN SCORE IN SYSTEMIC SCLEROSIS-ASSOCIATED INTERSTITIAL LUNG DISEASE. LONG-TERM OBSERVATION

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Background: There is a large clinical experience about the efficiency of rituximab (RTX) for the treatment of systemic sclerosis (SSc). There are several studies showing the decrease in skin induration and interstitial fibrosis in the lungs as the effect of therapy. However, there are not many long-term observations.

Objectives: To describe the efficacy of RTX on lung function and skin score in patients with systemic sclerosis-associated interstitial lung disease, in long-term follow-up.

Methods: This study included 71 patients (pts) with SSc. Data were collected prospectively. The mean follow-up period was 42 mo (12-72). Mean age was 46 years (17-66), female-59 (83%), diffuse cutaneous subset of the disease had 42 (59%), Scl-70 positivity-73% of pts. Duration of the disease was 5,6±4,4 yrs. All pts received concomitant treatment with low dose prednisolone and 45% - with immunosuppressants. The following indicators were evaluated: forced vital capacity,% predicted (FVC), diffusing capacity for carbon monoxide,% predicted (DLCO) and Rodnan skin score (mRss) over a periods of 12-18 months (point 1), 24-30 months (point 2), 36-42 months (point 3), 48-54 months (point 4) and 60-72 months (point 5) after the start of therapy. The results are presented in the form of mean values, delta, median, upper and lower quartile.