Results: Antinuclear antibodies were detected in 2129 out of 2809 systemic sclerosis patients enrolled in the multi-center cohort and 4.2% of them were negative. There was significant difference between patients with negative and positive antibodies based on gender (29/80 vs 29/1746, p<0.001). The presence of Raynaud’s phenomenon is less common (71.8% vs 99.8%, p<0.001) in the ANA-negative patients. In addition, compared with ANA-positive patients, the incidence of certain critical organ involvements, including gastrointestinal reflux (5.6% vs 18.5%, p=0.002), interstitial lung disease (65.2% vs 77.9%, p=0.015) and pulmonary arterial hypertension (11.5% vs 29.0%, p=0.006) were significantly lower in ANA-negative patients than in the positive group. The proportion of IgG elevation, an indicator of disease activity and severity of inflammation, was significantly lower in the ANA-negative patients than that in the positive group (14.3% vs 41.2%, p<0.001), while no significant differences were found in other inflammatory indicators and skin scores.

Conclusion: This study describes the clinical features of SSc patients with negative ANAs, which have been rarely mentioned or focused in existing studies. Antinuclear antibody is proved to be strongly associated with the clinical manifestations of systemic sclerosis patients and ANA-negative SSc patients tend to be in relatively milder conditions, including a less common involvement of critical organs and a more temperate inflammatory severity.

References:

Disclosure of Interests: None declared.

DOI: 10.1136/annrheumdis-2020-eular.3168

SAT0328 OUTCOME OF INTERSTITIAL LUNG DISEASE (ILD) IN ANTI-PM/SCL PATIENTS WITH SYSTEMIC SCLEROSIS: RESULTS FROM AN EUSTAR CASE-CONTROL STUDY.

M. G. Lazzaroni1, C. Campochiaro3,3, E. Marasco4, J. De Vries-Bouwstra5, F. Franceschini1, F. Del Galdo4, C. Denton2, L. Cavagna4, O. Distler7, Y. Alainore1, P. Airo1 on behalf of EUSTAR Co-Authors.

Background: The main clinical associations of anti-PM/Scl in Systemic Sclerosis (SSc) so far reported include calcinosis, myositis and interstitial lung disease (ILD). Nevertheless, data regarding the long-term outcome of ILD in these patients are lacking. A single centre Spanish cohort reported a better functional outcome in 14 SSc-ILD patients anti-Pm/Scl+ as compared to 49 anti-topo I after a mean follow-up of 7 years (1).

Objectives: To analyze the long-term outcome of ILD in a large multicentre EUSTAR study dedicated to anti-Pm/Scl SSc patients.

Methods: A case-control study within the EUSTAR cohort collected 165 anti-PM/Scl-SSc cases and 257 anti-PM/Scl-SSc controls, matched for sex, cutaneous subset, disease duration, and age at onset. Data for ILD at HRCT were available for 162/165 cases and 249/257 controls. Data for pulmonary function tests (PFT) at the baseline (T0), 1 year after diagnosis (T1) and at the last visit (LV) were analyzed.

Results: A significantly higher frequency of ILD was reported in anti-PM/Scl+ cases vs anti-PM/Scl- controls (62.3% vs 39.4%, p<0.0001, OR 95%, CI 2.55, 1.70-3.83). Complete PFTs data were available for 81/101 ILD anti-PM/Scl+ cases and 78/98 anti-PM/Scl- ILD controls, with similar age at onset and female/male ratio and disease duration at LV (112±81 months vs. 115±64 months, p=0.77). Diffuse cutaneous involvement was less frequent in cases than in controls (22% vs. 44.9%, p=0.03).

In ILD cases, %pFVC tended to improve from T0 (85.1±18.3) to T1 (89.5±16.5, p=0.045) and to LV (87.9±16.9, p=0.057), while in ILD controls remained stable from T0 (90.4±18.5) to T1 (91.1±16.5, p=0.38) and significantly declined to LV (85.0±18.0, p=0.0002). %pDLCO remained stable from T0 (50.5±16.8) to T1 (50.1±17.6, p=0.87) and to LV (50.4±16.9, p=0.77) in ILD cases, while significantly declined from T0 (60.7±18.9) to T1 (62.7±18.2, p=0.0016) and to LV (59.6±18.4, p=0.0001) in the control group. Mean %pFVC and %pDLCO at the 3 time points were not significantly different between the two groups.

Delta %pFVC (LV-T0) was 2.85±11.3 for the anti-PM/Scl+ group vs -5.42±13.4 in the control group (p=0.0004) with a significant smaller proportion of patients with FVC loss ≥10% from T0 to LV in the anti-PM/Scl+ group (12.3% vs. 39.7%, p=0.001). Delta %pDLCO (LV-T0) was -0.13±10.8 for the anti-PM/Scl+ group vs -7.36±18.6 in the control group (p=0.0015), with a significant smaller proportion of patients with DLCO loss ≥10% from T0 to LV in the anti-PM/Scl+ group (13.6% vs. 42.3%, p=0.0001).

Conclusion: In this multicenter real-life study, the long-term pulmonary functional outcome in SSc-ILD patients with anti-Pm/Scl positivity seems to be more favorable than in patients with anti-Pm/Scl antibodies.
Discourse of Interests: A. Lescoatal1, S. Jouneaua2, B. Crestanab3, G. Riemekastenc3, Y. Kondohs, V. Smitht1, N. Patelt1, J. Huggins2, C. Stock,3 M. Gahlemann3,4, M. Alves1, C. Denton1,5, CHU South Hospital, Internal Medicine, Rennes, France; 2Department of Respiratory Medicine, Competence Centre for Rare Pulmonary Diseases, CHU Rennes, Univ Rennes, Rennes, France; 3Hospital Bichat, Pneumology, Paris, France; 4University Hospital Charité, Rheumatology and Clinical Immunology, Berlin, and University Hospital Schleswig-Holstein, Rheumatology, Lübeck, Germany; 5Taisei General Hospital, Department of Respiratory Medicine and Allergy, Seto, Japan; 6Department of Rheumatology, Ghent University Hospital, Ghent, Belgium; 7Department of Internal Medicine, Ghent University, Ghent, Belgium; 8Columbia University College of Physicians and Surgeons/New York-Presbyterian Hospital, Division of Pulmonary, Allergy, and Critical Care Medicine, New York, New York, United States of America; 9Medical University of South Carolina, Charleston, South Carolina, United States of America; 10Boehringer Ingelheim Pharma GmbH & Co KG, Ingelheim, Germany; 11Boehringer Ingelheim (Schweiz) GmbH, Basel, Switzerland; 12Boehringer Ingelheim International GmbH, Ingelheim am Rhein, Germany; 13Centre for Respiratory and Connective Tissue Diseases, University College London Division of Medicine, London, United Kingdom.

Background: In the SENSCIS trial, nintedanib reduced the progression of SSC-ILD vs placebo, as shown by a lower rate of decline in forced vital capacity (FVC). The adverse event (AE) profile of nintedanib was characterised mainly by gastrointestinal (GI) events, including weight loss.

Objectives: Assess FVC decline and AEs in subgroups by weight loss ≤5% vs >5% over 52 weeks in the SENSCIS trial.

Methods: Patients with SSC-ILD with first non-Raynaud symptom <7 years before screening and ≥10% fibrosis of the lungs on an HRCT scan were randomised to nintedanib or placebo. In a non-randomised comparison, we analysed the rate of decline in FVC (mL/year) and AEs over 52 weeks in subgroups by weight loss (≤5% vs >5%) over 52 weeks.

Results: In the nintedanib (n=288) and placebo (n=288) groups, respectively, 112 (38.9%) and 43 (14.9%) patients had weight loss >5% over 52 weeks. At baseline, patients with weight loss >5% over 52 weeks had a higher mean age (57.0 vs 52.9 years), greater proportion of females (81.3% vs 72.9%), and similar mean BMI (26.5 vs 25.7 kg/m2, respectively) and FVC % predicted (71.0% vs 73.1%, respectively) vs patients with weight loss ≤5%. In the placebo group, the mean (SE) annual rate of decline in FVC was similar between patients who had weight loss ≤5% and >5% over 52 weeks (-92.7 [14.7] mL/year and -96.4 [34.9] mL/year, respectively). The estimated annual rate of decline in FVC was lower in patients treated with nintedanib than placebo, with between-group differences in patients who had weight loss ≤5% and >5% of 49.8 mL/year [95% CI 4.2, 95.6] and 30.2 mL/year [95% CI -50.5, 110.9], respectively, with no evidence of heterogeneity between subgroups by weight loss (p=0.68 for interaction). Standardised differences in baseline values of potential confounders were <0.2 (indicating negligible differences). The most frequent AEs in patients treated with nintedanib were diarrhoea (74.4% and 77.7% of patients with weight loss ≤5% and >5%, respectively), nausea (30.1% and 33.9%, respectively) and vomiting (19.3% and 33.3%, respectively). In the nintedanib and placebo groups, respectively, AEs leading to discontinuation of study drug occurred in 170% and 8.6% of patients with weight loss ≤5% and ≤5% and >5% over 52 weeks. AEs leading to discontinuation of nintedanib were not more frequent in patients with weight loss >5% vs ≤5%.

Conclusion: In the SENSCIS trial in patients with SSC-ILD, a greater proportion of patients treated with nintedanib than placebo had weight loss >5% over 52 weeks. The rate of decline in FVC was numerically lower in the nintedanib group than in the placebo group both in patients with weight loss ≤5% and >5% over 52 weeks. AEs leading to discontinuation of nintedanib were not more frequent in patients with weight loss >5% vs ≤5%.