classification criteria, in annual follow-up (for a total of 165 patients/year) with Pulmonary Function Tests (PFTs), Health Assessment Questionnaire - Disability Index (HAQ-DI), Scleroderma Health Assessment Questionnaire (sHAQ) and Cochin Hand Function Score (CHFS). Hand disability index was assessed by CHFS and global disability index was assessed by HAQ and sHAQ. Patient reported arthritis activity was assessed by Visual Analogical Scale for Arthritis Activity (VAS3). Based on the median of VAS3, patients were classified in two groups and the evaluation of global and hand disability index was performed for each group. Furthermore, we assessed the correlation between the change of VAS3 and the modification of disability scores (Δ HAQ, Δ sHAQ, Δ CHFS) over 12 months of follow-up. Following analysis of distribution, Spearman or Pearson Test were used to determine correlation coefficients, as appropriate (Prism 7).

Results: The median disease duration was 5 years (IQR 3-10). The median of VAS3 was 35 (IQR 2 - 66). In patients with VAS3 <35 and VAS3 $\geq$ 35 the HAQ-DI medians were 0.625 (IQR 0.25 - 1.5) and 1.75 (IQR 1.125-2.25) respectively, ($p < 0.0001$); the sHAQ medians were 0.628 (IQR 0.255 - 1.114) and 1.701 (IQR 1.234-2.059), respectively, ($p < 0.0001$); and the CHFS medians were 4 (IQR 0 - 19) and 28 (IQR 10 - 46) respectively, ($p < 0.0001$). A significant correlation was observed between VAS3 and HAQ ($r = 0.463$, $p < 0.0001$), sHAQ ($r = 0.651$, $p < 0.0001$), CHFS ($r = 0.497$, $p < 0.0001$); between Δ VAS3 and Δ sHAQ ($r = 0.493$, $p < 0.0001$).

Conclusion: This analysis of a monocentric non-selected population supports the key role of joint involvement in determining global patient reported functional and hand disability in SSc. Severity of musculoskeletal involvement should be carefully considered when interpreting PROs in patients with SSc.

References:

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FRI0242

IMPACT OF PULMONARY ARTERIAL HYPERTENSION WITH OR WITHOUT INTERSTITIAL LUNG DISEASE ON SCLERODERMA: A RETROSPECTIVE COHORT STUDY FROM THE NATIONWIDE SPANISH SCLERODERMA (RESCLE) AND PULMONARY ARTERIAL HYPERTENSION (REHAP) REGISTRIES

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Background: Pulmonary arterial hypertension (PAH) and interstitial lung disease (ILD) are the major life-threatening complications in systemic sclerosis (SSc). Data on the impact of PAH and/or ILD in SSc patients (pts) are limited by their low prevalence.

Objectives: To assess differences in demographic/clinical characteristics of SSc pts according to presence of PAH and how these are affected by ILD. The impact on characteristics and survival of PAH + ILD was also assessed.

Methods: We compared data on SSc pts without PAH from the Spanish registry of patients with SSc (RESCLE) (SSc pts) and SSc pts with PAH from the Spanish registry of pts with PAH (REHAP) (SSc-PAH pts). Only data common in both registries were analyzed. Sub analyses were performed according to the presence/absence of ILD. Transplant-free survival from diagnosis of PAH was estimated using the Kaplan-Meier method.

Results: 1,579 pts with SSc (RESCLE) and 364 pts with SSc-PAH (REHAP) were analyzed. Compared to SSc pts, SSc-PAH pts had worse functional status (NYHA FC III/IV: 70.6% vs. 8.2% in SSc pts) and pulmonary function (lower

mean forced vital capacity [FVC, 81.2 $\pm$ 20.6% vs. 93.6 $\pm$ 20] and diffusing capacity for carbon monoxide [DLCO, 45.3 $\pm$ 17.7% vs. 79.0 $\pm$ 36.6%]). More patients had FVC/DLCO $\geq$ 1.4 (77.8% vs. 34.8%), tricuspid regurgitation (91.4% vs. 46.1%) or pericardial effusion (30.0% vs. 5.1%). Mean systolic pulmonary artery pressure (sPAP) was higher (70.0 $\pm$ 21.3 vs. 27.5 $\pm$ 9.1 mmHg) (all respectively; $P < 0.001$ for all). Prevalence of ILD on high-resolution computerized tomography was similar (44.9% [n=92] vs. 41.8% [n=422] in SSc pts; $P = 0.408$). These differences were also found when splitting both cohorts according to the presence / absence of ILD. Compared to SSc-PAH pts without ILD (n=128), pts with PAH + ILD (n=92) had worse functional status (NYHA FC III: 69.6% vs. 55.5% in SSc-PAH pts without ILD; $P = 0.036$), lower mean FVC (70.9 $\pm$ 21.9 vs. 86.2 $\pm$ 18.6) and lower DLCO (39.4 $\pm$ 17.0 vs. 49.1 $\pm$ 17.9) both $P < 0.001$. Five-year survival rate was 35% in SSc-PAH pts with ILD vs. 45% in SSc-PAH without ILD ($P = 0.444$ [figure 1]).

Conclusion: PAH has a profound impact on functional status, pulmonary function and right ventricle function of SSc patients, independently of presence of ILD. Despite the deleterious effect of functional status and pulmonary function, in pts with SSc and PAH, presence of concomitant ILD has no impact on 5-year survival.

Figure 1. Survival rate estimates in SSc-PAH pts with and without ILD

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FRI0243

SARCOPENIA IS ASSOCIATED WITH MALNUTRITION IN PATIENTS WITH SYSTEMIC SCLEROSIS

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Background: Sarcopenia is one of the major health problems in older patients and is defined as a progressive decrease in muscle mass and function¹. Sarcopenia has only rarely been studied in systemic sclerosis (SSc) and its impact in clinical characteristics of SSc is poorly investigated.

Objectives: To evaluate the associations between sarcopenia and clinical features in SSc patients.

Methods: Cross-sectional study, including 82 patients who met the ACR/EULAR 2013 classification criteria for SSc. Dual-energy X-ray absorptiometry, handgrip strength, and short physical performance battery were used to assess sarcopenia according the European Working Group on Sarcopenia in Older People's (EWG-SOP) diagnostic criteria updated in 2019¹. Malnutrition was evaluated according to the European Society of Clinical Nutrition and Metabolism (ESPEN)², using the Malnutrition Universal Screening Tool (MUST) to screen risk for malnutrition.