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.

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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.

Results: The mean age was 60.4 years and 91.5% were women (table 1). Sarcopenia was identified in 15 (18.3%) SSc patients. Malnutrition was diagnosed in 12 (14.6%) and was more common in patients with sarcopenia ($P=0.038$). Also, there were significant differences between patients with and without sarcopenia regarding Body Mass Index ($P=0.001$), Fat Free Mass Index ($P<0.001$), knee extension strength ($P=0.049$), and Timed Up and Go test ($P=0.041$). There were no differences regarding age, disease duration, Rodnan skin score (mRSS), FVC and DLCO.

Table 1. Clinical characteristics of SSc patients with and without sarcopenia. Data presented as number (%) of patients, except when indicated otherwise.

Patients features	Whole (n=82)	SSc patients without sarcopenia (n=67)	SSc patients with sarcopenia (n=15)	P
Females	75 (91.5)	62 (92.5)	13 (86.6)	0.606
Caucasian	68 (82.9)	53 (79.1)	15 (100)	0.258
Age (years) ^b	60.4 (10.6)	60.2 (10.3)	61.5 (12.3)	0.678
Diffuse skin involvement	16 (19.5)	16 (23.8)	0 (0)	0.082
Disease duration (years) ^a	12.8 (7.5, 19.2)	12.7 (7.1, 19.2)	13.4 (8.9, 19.8)	0.324
Rodnan skin score ^a	4.0 (2.0, 10.0)	5.0 (2.0, 10.0)	2.0 (2.0, 10.0)	0.076
Interstitial lung disease on HRCT	32 (39.5)	25 (37.3)	7 (10.4)	0.737
FVC (% predicted) ^b	88.4 (16.6)	88.4 (16.9)	88.3 (15.3)	0.991
DLCO (% predicted) ^b	63.7 (11.9)	63.6 (12.2)	64.2 (11.2)	0.855
BMI (kg/m ²) ^b	25.6 (4.6)	26.5 (4.4)	21.8 (4.1)	0.001
FFMI (kg/m ²) ^b	15.6 (1.7)	16.1 (1.6)	13.8 (1.2)	<0.001
FMI (kg/m ²) ^b	9.5 (3.7)	9.9 (3.6)	7.5 (3.5)	0.023
Malnutrition	12 (14.6)	7 (10.4)	5 (33.3)	0.038
MUST - Low risk	58 (70.7)	51 (76.1)	7 (46.7)	0.046
Moderate and high risk	24 (29.3)	16 (23.9)	8 (53.3)	
HAQ ^b	0.778 (0.591)	0.797 (0.575)	0.833 (0.576)	0.825
Timed Up and Go test (s) ^a	8.41 (7.41, 10.59)	8.39 (7.5, 9.4)	10.23 (7, 14)	0.041
Knee extension strength (kgf) ^a	21.40 (15.65, 27.52)	21.70 (18.05, 28.35)	14.82 (9.5, 24)	0.049

^a Median (25, 75th percentiles)

^b Mean (standard deviation)

Abbreviations: BMI: body mass index; DLCO: diffusion capacity of carbon monoxide; FFMI: fat free mass index; FMI: fat mass index; FVC: forced vital capacity; HAQ: Health Assessment Questionnaire; HRCT: high-resolution computed tomography; MUST: Malnutrition Universal Screening Tool; SSc: systemic sclerosis.

Conclusion: Sarcopenia is relatively common and is associated with malnutrition in patients with SSc. In our population, sarcopenia was not associated with other features related to a more severe disease. The role of sarcopenia in the prognosis of SSc needs to be better understood in longitudinal studies.

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FRI0244

EOSINOPHILIC FASCIITIS-LIKE SECONDARY TO CHRONIC GRAFT-VERSUS-HOST DISEASE: CLINICAL DESCRIPTION OF 28 PATIENTS

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Background: Eosinophilic fasciitis (EF) is an uncommon chronic inflammatory disease characterized by myalgia, soft tissue hardening, peripheral eosinophilia and increased acute phase reactants, often triggered after strenuous physical exercise. Its appearance has been described as a rare complication after allogeneic transplantation of hematopoietic progenitors in the context of chronic

graft-versus-host disease (cGVHD). Its etiopathogenesis is not well known and usually the treatment with systemic corticosteroids is effective

Objectives: Describe the clinical and transplant-related characteristics of a cohort of patients with eosinophilic fasciitis-like in the context of cGVHD

Methods: Observational, retrospective and descriptive study of the clinical characteristics of 28 patients affected by EF-like followed in a multidisciplinary consultation of cGVHD, started in March, 2014. Regular demographic variables, clinical characteristics related to the transplant and with the cGVHD, laboratory parameters, rescue therapies and their response were collected. The statistical analysis was done with Microsoft Excel 2007.

Results: Seventeen (60.7%) patients were male and 11 (39.35%) were women with a mean age of 48.75 years (range from 10 to 74). Acute myeloid leukemia was the most frequent cause of the transplant in 11 patients (39.3%). Transplant related characteristics are reflected in Table 1 and the clinical manifestations, therapies received and their response in Table 2. Four (14.2%) patients died during their follow-up, being the cause of death in 2 cases due to sepsis, and in 1 case attributable to GVHD.

Table 1. Baseline and transplant related characteristics (N = 28).

Variables	N (%)
Donor Type (related / not related)	13(46.4%)/15(53.6%)
Type of conditioning (reduced intensity / myeloablative)	18(64.25%)/10(35.7%)
Source of cells (Peripheral blood / bone marrow)	27(96.4%)/1(3.6%)
cGVHD type quiescent / de novo / progressive	11(39.3%)/13(46.4%)/4(14.3%)
Other affected organs (cGVHD score)	
-Mouth	6(21.4%)
-Eyes	10(35.7%)
-Lung	2(7.1%)
-Liver	3(10.7%)
-Gastrointestinal tract	0(0%)
-Genital	2(7.1%)
-Cutaneous	16(57.14%)
Global Score	4(14.2%)/14(50%)/10(35.7%)
NIH ¹ (mild / moderate / severe)	

¹ NIH: National Institute of Health.

Table 2. Clinical manifestations and therapies (N = 28).

Variables	N (%) / Median (range)
Prodromic symptoms: yes / no	20(71%)/8(29%)
- Stiffness	2(7.1%)
- Artromyalgia	17(60.7%)
- Edema	3(10.7%)
Time until first visit	31.3 months (range 9-73)
Contracture Yes / No	18(64.3%)/10(35.7%)
Mobility limitation (mild / moderate)	13(46.4%)/9(32.1%)
ECOG ¹ affected	11(39.2%)
Eosinophilia	17(60.7%)
Positive autoantibodies	8(28.5%)
First line therapies (corticosteroids)	28(100%)
Extracorporeal photopheresis	19(67.9%)
Therapies of 2nd line/ 3rd or more	6(21.4%)/12 (42.8%)
Physiotherapy	14 (50%)
Response: complete / sequels	10(35.7%)/18(64.2%)

¹ECOG: Eastern Cooperative Oncology Group scale to assess the quality of life

Conclusion: Nonspecific joint symptoms such as stiffness, edema or arthromyalgia in patients undergoing allogeneic transplantation of hematopoietic progenitors may be factors that predict the development of sclerotic GVHD type eosinophilic fasciitis-like and should be closely monitored in order to be able to perform early stage diagnoses of the disease. It is necessary to deepen the pathogenesis of this entity and the multidisciplinary approach to improve the prognosis of patients with GVHD

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FRI0245

PULMONARY ARTERIAL HYPERTENSION IN SYSTEMIC SCLEROSIS IS NEARLY ALWAYS ACCOMPANIED BY A LOW DIFFUSING CAPACITY

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Background: Scleroderma (systemic sclerosis; SSc) has high morbidity and mortality. Pulmonary hypertension (PH) and pulmonary arterial hypertension