the associations with joint and muscle involvement, lung fibrosis, and intestinal symptoms were confirmed (table 1).

Table 1 Results of the univariable and multivariable analysis adjusted on sex, age at disease onset and disease duration (n=8142 patients). Results are presented as number/number available data (%) unless stated otherwise.

Characteristics	Univariable analysis			Multivariable analysis		
	Anti-PM/Scl -	Anti-PM/Scl +	p Value	N available data (%)	OR (95% CI)	p Value
Age at disease onset (years, mean (SD) (n available))	46.7 (14.5) (6885)	44.7 (15.1) (140)	0.081	7025 (86.3)	0.98 (0.90 to 0.99)	0.0015
Disease duration (months, mean (SD) (n available))	152.2 (107.6) (6874)	129.5 (99.6) (139)	0.052	7013 (86.1)	0.997 (0.995 to 0.999)	0.0136
Male sex	1140/7992 (14.2)	31/150 (20.7)	0.036	8142 (100.0)	1.201 (0.706 to 2.043)	0.4995
Caucasian ethnicity	6462/7992 (80.9)	126/140 (90.0)	0.332	8142 (100.0)		
Raynaud's phenomenon	6904/7549 (91.5)	146/150 (97.3)	0.016	7699 (94.6)		
Oesophageal symptoms	4317/766 (56.4)	93/150 (62.0)	0.190	7811 (95.9)		
Stomach symptoms	1553/7337 (21.2)	49/150 (32.7)	0.008	7487 (92.0)		
Intestinal symptoms	1803/7587 (23.8)	63/150 (42.0)	<0.0001	7737 (95.0)	2.894 (1.890 to 4.430)	<0.0001
Scleroderma renal crisis	123/7605 (1.6)	8/150 (5.3)	0.0015	7755 (95.2)	1.489 (0.525 to 4.219)	0.4540
Diffuse cutaneous subtype	1990/7412 (54.7)	55/148 (45.3)	0.022	7560 (92.9)	0.825 (0.525 to 1.296)	0.4033
Active digital ulcers ever	2597/4973 (52.2)	66/126 (52.3)	0.97	5099 (62.6)		
Joint synovitis	734/7411 (9.9)	34/149 (22.8)	<0.0001	7560 (92.9)	2.963 (1.199 to 3.215)	0.0073
Joint contractures	2191/7152 (29.9)	64/149 (43.0)	<0.0001	7301 (89.7)		
Tendon friction rubs	404/7087 (5.8)	14/149 (9.4)	<0.0001	7236 (88.9)		
Muscle weakness	1177/7512 (15.7)	59/148 (39.9)	<0.0001	7660 (94.1)		
Muscle atrophy	668/7334 (9.1)	37/148 (25.0)	<0.0001	7482 (91.9)		
CX elevation	380/6259 (6.1)	47/140 (33.6)	<0.0001	6399 (78.6)	6.695 (4.181 to 10.720)	<0.0001
Conduction blocks	880/5333 (16.5)	35/131 (26.7)	0.029	5464 (67.1)	1.468 (0.905 to 2.382)	0.1199
Elevated sPAP (ECHO)	976/5592 (17.5)	19/137 (13.9)	0.33	5729 (70.4)		
Lung fibrosis on plain X-rays	64/117 (54.7)	1542/4527 (51.6)	<0.0001	4644 (57.0)	2.512 (1.629 to 3.872)	<0.0001

Conclusions: In the largest series of anti-PM-Scl positive patients so far reported, well-known clinical associations were confirmed. Moreover, scleroderma renal crisis was more frequent than in the antibody-negative patient controls (which included a majority of anticentromere-positive patients, and a relatively small number of anti-RNA polymerase III-positive patients). However, this association was probably explained by covariates, such as joint and muscle involvement, or lung fibrosis. A possible role of corticosteroid therapy might therefore be suspected.

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THU0416

VERTEBRAL FRACTURE PREVALENCE AND MEASUREMENT OF THE SCANOGRAPHIC BONE ATTENUATION COEFFICIENT ON CT SCAN IN 70 PATIENTS WITH SYSTEMIC SCLERODERMA

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Background: Osteoporosis screening is not systematic in sclerodermic patients but some studies demonstrated a similar risk between rheumatoid arthritis and systemic scleroderma.^{1,2,3} Thoracic and/or TAP (thoraco-abdomino-pelvic) CT (Computed Tomography) scans are classically performed in the follow-up of scleroderma, mainly to evaluate lung involvement.

Objectives: To study vertebral fracture (VF) prevalence and the scanographic bone attenuation coefficient of the first lumbar vertebra (SBAC-L1) on CT scans in systemic scleroderma patients. Secondary objectives are to study specific risk factors of SBAC-L1 ≤145 Hounsfield Units (HU) and to evaluate SBAC-L1 measurements reliability.

Methods: This monocentric retrospective study included patients followed from 2000 to 2014 and fulfilling ACR/EULAR 2013 criteria for systemic scleroderma and who underwent a thoracic or TAP CT scan. Osteoporotic risk factors, Dual Energy X-ray Absorptiometry (DXA) measurements and clinical characteristics were collected. For CT scan, the VFs were determined according to Genant's

classification on sagittal sections. The SBAC-L1 was measured in Hounsfield Units (HU) on axial section of L1 in a Region of Interest drawn in trabecular bone. Intra- and inter-reader reliabilities for SBAC-L1 were calculated. An SBAC-L1 ≤145 HU (fracture threshold) was used to define patients at risk of VF.⁴ Predictive factors for VF or SBAC-L1 ≤145 HU were studied.

Results: A total of 70 patients were included (mean age: 62.3 (±15.6) years, women 88.5%, diffuse scleroderma 22.9% (n=16)) in the study. Sixty patients (85.7%) presented with at least one clinical risk factor for osteoporosis. Eighteen patients (25.7%) received vitaminocalcic supplementation and 10 (14.3%) received antiresorptive therapy. DXA was only performed on 30 patients (42.8%), and 5 (16.7%) of them presented a T-score ≤ -2.5 DS. 3 VFs were detected in 3 patients (4.3%). The mean SBAC-L1 was 157.26 HU (±52.1), and 35 patients (50%) presented a SBAC-L1 ≤145 HU. SBAC-L1 measurements were highly reliable (Kappa >0.9 for both intra- and inter-reader reliability). For the univariate analysis, a SBAC-L1 ≤145 HU was significantly associated with age (OR=1.09, CI 95%: 1.04–1.13), calcinosis (OR=6.3, CI 95%: 1.61–24.75) and periarticular calcifications (OR=3.22, CI 95%: 1.06–9.77). For the multivariate analysis, age (especially patients older than 63 years), calcinosis and acro-osteolysis were independently associated with a SBAC-L1 ≤145 HU.

Conclusions: On a large sample of sclerodermic patients with clinical risks of osteoporosis, only 42.8% were screened for DXA and 16.7% of them were osteoporotic. The VF prevalence on CT scan was 4.3% and the SBAC-L1 measurement identified 50% of the population at the fracture threshold. The presence of calcinosis, periarticular calcifications or acro-osteolysis should lead to an osteoporosis screening, especially for patients under 63 years old.

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WHOLE BODY DISTRIBUTION AND CLINICAL ASSOCIATIONS OF TELANGIECTASIA IN SYSTEMIC SCLEROSIS: A CROSS-SECTIONAL STUDY

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Background: Telangiectasia (TA), one of the diagnostic criteria for systemic sclerosis (SSc), could be a clinical marker for the severity of vasculopathy, including pulmonary hypertension (PH).

Objectives: We designed a cross-sectional study: (i) to describe the whole-body distribution of TA, (ii) to assess the associations between the whole-body number of TA and the characteristics of patients, (iii) to determine whether the number of TA may be useful to discriminate SSc-PH patients.

Methods: Patients were included in the National Referral Centre for Rare Systemic And Autoimmune Diseases if they fulfilled the 2013 ACR/EULAR criteria for SSc. They were excluded if they had received laser treatment. The whole-body number and distribution of TA were recorded at inclusion. The associations were studied using univariate, adjusted and multiple linear regressions.

Results: 106 patients were enrolled, including 12 with PH. The median (interquartile range) number of TA was 30 (82.7). Their distribution was: 37.2% on the face, 33.2% on the upper limbs including 26.4% on the hands, 28.1% on the trunk including 17.1% for the upper part of the trunk, and 1.5% on the lower limbs. Using multivariate linear regression model, the whole-body telangiectasia number was independently associated with male gender (percentage change (95% CI)= +144.4% (7.5; 455.9), p=0.033), pulmonary hypertension (+162.8% (5.6; 553.8), p=0.038), history of pulmonary embolism (+336.4% (39.0; 1270.1), p=0.012), glomerular filtration rate (−1.6% (−3.2; −0.1) per 1 ml/min/1.73m² increase, p=0.038) and soluble endoglin (+28.2% (1.2; 62.5) per 1 ng/ml increase, p=0.039). The ROC analyses assessing the ability of telangiectasia to discriminate the presence of pulmonary hypertension revealed that the area under the curve was significant for the telangiectasia number on the whole body (0.77 (0.57; 0.88)), on the hands and face (0.81 (0.57; 0.91)) and on the hands (0.77 (0.57; 0.89)).

Conclusions: In SSc-patients, TA were predominantly located on the face, hands and the upper part of the trunk. They may reflect the vasculopathy of SSc and could represent a clinical biomarker for vascular disease, particularly for PH, one of the most severe vascular complications of the disease.

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THU0418

LONG-TERM EFFICACY AND SAFETY OF MONOTHERAPY VERSUS COMBINATION THERAPY IN SYSTEMIC SCLEROSIS-ASSOCIATED PULMONARY ARTERIAL HYPERTENSION (PAH): A RETROSPECTIVE COHORT STUDY FROM THE NATIONWIDE SPANISH SCLERODERMA REGISTRY (RESCLE)

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Background: Monotherapy with endotelin antagonist receptors (ERA) an phosphodiesterase 5 (PDE5) inhibitors is a first choice treatment for PAH in functional class (FC) II-III, with the same grade of evidence and recommendation than combination therapy. Recently, studies have proven superiority of combination therapy against monotherapy in combined morbi-mortality endpoints.

Objectives: To demonstrate superiority of combination therapy against monotherapy in a single mortality endpoint in SSc-associated PAH.

Methods: Retrospective cohort study including patients from the Spanish Scleroderma Registry (RESCLE) diagnosed with SSc-associated-PAH by right heart catheterization (RHC). Patients were divided in 3 groups: monotherapy vs. sequential combination therapy (>12 weeks between first and second treatment)

vs. upfront combination therapy (<12 weeks between treatments). Primary end-point was mortality from any cause.

Results: Seventy-six patients with PAH out of 1817 participants were included. Thirty-four (45%) were receiving monotherapy [with ERA (22 patients, 29%) or PDE5 inhibitors (12 patients, 16%)], 25 patients (33%) sequential combination therapy and 17 patients (22%) upfront combination therapy.

Baseline demographic, clinical and complementary tests were similar among groups. ILD (mainly moderate) was more frequent in both combination groups in 58% vs. 80% vs. 76.4%, without statistical significance. A worse FVC/DLCO in the sequential combination group was reported (2.9 ± 1.1 vs. 1.8 ± 0.4 vs. 2.3 ± 0.8 , global $p=0.085$ but $p=0.043$ comparing monotherapy with sequential combination) and also a worse mPAP in both sequential and upfront combination groups (37.2 ± 8.7 mmHg vs. 40.8 ± 8.8 vs. 46 ± 15.9 , $p=0.026$).

The treatment regimen prescribed ($p=0.017$) and FC at baseline ($p=0.007$) were found predictors of mortality. Sequential combination therapy was found a protective factor [HR=0.11 (95%CI 0.03–0.51), $p=0.004$] and the upfront combination therapy showed a tendency of protection [HR=0.68 (95%CI 0.23–1.97), $p=0.476$]. Survival rates from diagnosis of PAH among groups were: 78% vs. 95.8% vs. 94.1% at 1 year, 40.7% vs. 81.5% vs. 51.8% at 3 years and 31.6% vs. 56.5% vs. 34.5% at 5 years ($p=0.007$).

Side effects were not significantly different among groups.

Conclusions: Combined sequential therapy improves survival in SSc-PAH patients, even with moderate ILD.

Upfront combination therapy may improve survival, but did not reach statistical significance due to study limitations.

Treatment regimen and FC were found as prognostic factors for survival: sequential combination therapy was a protective factor and FC was a risk factor.

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THU0419

ASSOCIATION OF INFLAMMATORY MARKERS C-REACTIVE PROTEIN AND ERYTHROCYTE SEDIMENTATION RATE WITH PULMONARY FUNCTION TESTS AND EUROPEAN SCLERODERMA STUDY GROUP ACTIVITY INDEX (ESCSG-AI) IN SYSTEMIC SCLEROSIS – ASSOCIATED INTERSTITIAL LUNG DISEASE IN FOLLOW UP STUDY

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Background: inflammatory markers are very important to assess severity and activity of SSc-ILD, but it's role needs further investigation.

Objectives: To assess inflammatory markers of SSc such as hsCRP and ESR and compare with lung function test and ESCSG-AI in the long-term follow up study.

Methods: It was a longitudinal study involving 77 pts with SSc-ILD (mean age was 46.2 ± 13.4 ; 69% have limited subset of the disease; 93% were female). The mean duration of follow up was 58.9 ± 11.4 months. Pts. were investigated with HRCT twice (at first visit and at the end of the study) and according the CT-changes were divided into 3 groups: the 1st group (16 pts) with improvement; 2nd group (39 pts) without any changes and 3rd group (22 pts) with worsening of fibrosis. Other data collected including biological results (high-sensitivity C-reactive protein (hsCRP) and erythrocyte sedimentation rate (ESR)), PFT (forced vital capacity (FVC,% of predicted) and diffusing capacity of the lung for carbon monoxide (DLCO,% of predicted), composite score (ESCSG-AI).

Results: there were no significant differences between groups related to sex, frequency of diffuse form and duration disease. Mean levels of hsCRP and ESR didn't change significantly during the follow up. In all pts the mean levels of hsCRP and ESR correlated directly with each other at first visit and at the end of the study ($R=0.45$ and $R=0.4$ ($p<0.001$) accordingly). We compared the mean levels of hsCRP and ESR with mean dates of FVC, DLCO and ESCSG-AI score in first visit and the end of follow up. Mean levels of hsCRP inversely correlated with mean dates of DLCO at the first visit and at the end of the study ($R=-0.39$ and $R=-0.42$ ($p<0.05$) accordingly); in groups 2 and 3 ($R=-0.34$ and $R=-0.47$ ($p<0.05$) accordingly) at the end of the study; with mean dates of FVC in all pts and group 2 ($R=-0.42$ and $R=-0.47$ ($p<0.05$) accordingly) only at the end of the study; correlated directly with ESCSG-AI score in all pts and groups 2,3 ($R=0.58$ ($p<0.0001$), $R=0.46$ ($p<0.01$) and $R=0.77$ ($p<0.001$) accordingly) at the end of the study. While mean levels of ESR inversely correlated with mean dates of DLCO only in all pts and groups 1,2 ($R=-0.43$, $R=-0.66$ and $R=-0.39$ ($p<0.05$) accordingly) at first visit; correlated directly with ESCSG-AI score in all pts. ($R=0.309$ ($p<0.01$) at the end of the study. Mean levels of hsCRP inversely correlated with DLCO, FVC and directly correlated with ESCSG-AI and these correlations were more evident than with mean levels of ESR.

Conclusions: In our group of pts. the hsCRP has proven to be an accurate reflection of disease severity especially in pts with progression of ILD.