Diffuse scleroderma	6/11
Limited scleroderma	5/11
Pulmonary hypertension	4/11
History of digital ulcers	5/11
Time of endurance in minutes (ergospirometry)	5':32" (±65")
Diffusion capacity (%pred)	82±15
Pulmonary fibrosis	7/11
VE/VCO2 (mean values ± sd)	32±5
Patterns of capillaroscopy	1/11 normal 1/11 early 4/11 active 5/11 late

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[2] Castellví I, Simeón-Aznar CP, Sarmiento M, et al. Association Between Nailfold Capillaroscopy Findings and Pulmonary Function Tests in Patients with Systemic Sclerosis. J Rheumatol. 2015 Feb; 42(2): 222–7.
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AB0602 CAROTID ATHEROESCLEROTIC PLAQUES DETECTED BY ULTRASOUND IN SYSTEMIC SCLEROSIS PATIENTS UNDER 55 YEARS OLD

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Background: In Systemic sclerosis (SSc), as in other autoimmune diseases such as rheumatoid arthritis or systemic lupus erythematosus, cardiovascular events are one of the most frequent causes of mortality not attributed to the disease itself.
Objectives: The aim of this study was to assess the presence of subclinical atherosclerosis by carotid Doppler ultrasound in SSc patients under 55 years.
Methods: We conducted a cross-sectional study that included 78 SSc patients without cardiovascular events from H. Vall d'Hebron cohort (Barcelona). Carotid Doppler ultrasound was performed to measure the Carotid Intima Media Thickness (CIMT) of common carotid artery (CCA) and detection of cholesterol plaques in CCA, bulb and internal and external carotid arteries, according to Mannheim consensus criteria. The results were compared to a healthy cohort from Barcelona, adjusted to age and sex[1]. We used SCORE for populations with low risk and REGICOR as cardiovascular risk assessment charts.
Results: Risk factors and SSc related features are described in table 1. Twenty three patients (29.5%) had carotid plaques (CP) being the presence of CP statistically significant compared to the healthy cohort (29.5% vs 15.6%; $p<0.05$; IC 0.04–0.24). None of the patients were catalogued as high risk according to SCORE chart. According to REGICOR chart, 4 patients (5.1%) were catalogued as intermediate risk and none as high risk. The four intermediate risk patients had CP. Carotid Intima Media Thickness mean (CIMTm) and maximum (CIMTmax), were statistically significant increased compared to the healthy cohort (CIMTm 0,57 vs 0,53; $p<0,05$ and CIMTmax 0,74 vs 0,61; $p<0,05$).
We performed multivariate regression analysis. Age, CIMTmax, low High Density Lipid (HDL), the presence of pulmonary hypertension, and Diffuse cutaneous SSc patients were independent factors for the presence of CP.

Female	62 (79.5%)
Age	44 (20–55)
Disease duration	14 (0–38)
Limited cutaneous SSc	41 (57.7%)
Diffuse cutaneous SSc	17 (23.9%)
Sine Scleroderma SSc	8 (11.3%)
Initial Scleroderma	5 (7%)
Arterial Hypertension	18 (23.1%)
Diabetes mellitus	2 (2.8%)
Statin treatment	14 (17.9%)
Smokers	15 (19.2%)
Digital ulcers	30 (42.3%)
Pulmonary Hypertension*	4 (5.1%)
Interstitial lung disease	32 (41%)

*Mean pulmonary artery pressure >25 mm Hg at right heart catheterization.

Conclusions: SSc patients under 55 years old often have subclinical atheromatous disease which could be detected by carotid ultrasound. Risk charts do not correlate with the presence of subclinical atheromatous disease. The presence of carotid plaque is more frequent in SSc patients with pulmonary hypertension, diffuse cutaneous subtype and patients with low HDL. CIMT could be useful although it is currently not recommended in cardiovascular risk guidelines.
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AB0603 QUANTITATIVE VIDEOCAPILAROSCOPY STUDY IN SYSTEMIC SCLEROSIS

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Background: Systemic sclerosis (SSc) is an autoimmune disease characterized by immunological features, microvascular disturbances and fibrosis as a consequence of a massive production of extracellular matrix.
Objectives: The aim of this study was to analyse the alterations of the capillary nailfold bed and describe their correlation with clinical variables.
Methods: 134 patients were selected with SSc according to LeRoy and Medsger diagnostic criteria from the Hospital Universitari Vall d'Hebron cohort. Patients underwent an echocardiogram, pulmonary function test and nailfold videocapillaroscopy (NVC) within a 3 months period. NVC was performed from 2nd to 5th finger of both hands. NVC features were quantitatively measured. Informed consent was obtained by all the participants. SPSS 20.0 were used for statistical analysis. A P-value <0.05 was considered as significant.
Results: Female was the predominant gender (113, 84.3%). The most common cutaneous subtypes were limited SSc, diffused SSc (88, 65.7% and 28, 20.9%, respectively). Almost 80% of patients met the ACR/EULAR 2013 classification criteria. The age (± standard deviation) at NVC was 38.5 (± 15.7) years, and the interval from the first SSc symptom and NVC was 16 (± 12.6) years. Anticentromere antibodies (ACA) were the most frequent in 36.6% of patients, followed by anti-topoisomerase I in 23.1%. Regarding organ involvement, 62 (46.3%) patients suffered from digital ulcers, 58 (43.3%) interstitial lung disease (ILD), 11 (8.2%) pulmonary hypertension, 110 (82.1%) gastrointestinal disturbances and 103 (76.9%) some cardiac alterations. The forced vital capacity was 80.8 (± 20.1) % and diffusion capacity for carbon monoxide (DLCO) was 66.2 (± 23.7) %. Regarding echocardiography results, the tricuspid regurgitation velocity (TRV) was 2.8 (± 0.3) m/s and the right ventricular systolic pressure 30.7 (± 12.0) mmHg. NVC findings were described as follows: the median of number of capillaries was 5.4 /mm, enlarged capillaries 0.8/mm, giant capillaries 0.2 /mm, microhemorragies 0.1 /mm, ramified capillaries 0.3 /mm, tortuous capillaries 0.6 /mm and disorganized capillaries 0.0 /mm. SSc-ILD patients presented lower capillary density 4.8 /mm compared with 5.8 /mm in the other group (P=0.005), and also more frequent ramified capillaries 0.3 /mm compared with 0.2 /mm (P=0.013). With respect to the correlations, the number of capillaries was related with the DLCO (rho =0.26, P=0.003) but negatively with the RVSP (rho = - 0.21, P=0.03). Giant capillaries were correlated with LVEF (rho =0.27, P=0.001) and greater tricuspid annular plane systolic excursion (TAPSE) (rho =0.21, P=0.01). Microhemorraghies were associated with LVEF (rho =0.29, P=0.001) although negatively with age at NVC (rho = - 0.25, P=0.003) and time from first symptom (rho = - 0.25, P=0.003). Disorganized capillaries were related with age at NVC (rho =0.17, P=0.04) but negatively with DLCO (rho = - 0.18, P=0.035).
Conclusions: The existence of a correlation between NVC features and clinical variables suggests that microvascular alterations may play a role as a pathogenic link with the cardiopulmonary SSc manifestations.
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AB0604 RITUXIMAB EXPERIENCE IN PATIENTS WITH LONGSTANDING SYSTEMIC SCLEROSIS-ASSOCIATED INTERSTITIAL LUNG DISEASE: A SERIES OF 14 PATIENTS

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Background: Interstitial lung disease (ILD) is the leading cause of mortality in systemic sclerosis (SSc) patients. Treatment options are rather limited in SSc associated ILD (SSc-ILD).
Objectives: Objective of this study was to report the experience of RTX treatment in a series of patients with longstanding SSc-ILD in whom unsatisfactory response in lung functions was noted under conventional treatments.
Methods: We retrospectively reviewed charts of 197 SSc patients evaluated between April 2015 and November 2016. 14 patients who received rituximab (RTX) for SSc-ILD participated in this analysis. The severity of ILD based on PFTs was defined as follows; mild (FVC between 71% and 80% of predicted), moderate (FVC between 51% and 70% of predicted) and severe (FVC $\leq$50%). The extent of skin disease was clinically measured by using Modified Rodnan Skin Score (mRSS) tool. End of follow-up was considered as six months after the last RTX dose.
Results: Median (IQR, interquartile range) age was 53.2 (46.8–55.5) and median disease duration was 9.1 (5.1–13.6) years. Median FVC was 52.5 (41.5–64.0)

prior to RTX. At the end of follow-up, no significant change was revealed in FVC when compared with pre-RTX values [58.0 (44.7–58.7), p=0.065]. FVC was improved in four patients and stabilized remaining ten patients. All of the patients with improvement of PFTs had moderate or severe restrictive lung disease. High resolution thorax computed tomography (HRCT) findings remained stable in 7 and showed progression of ILD in 3 patients. In total, mRSS remained stable at the end of follow-up when compared with baseline [8.0 (5.2–12.2) vs. 6.0 (4.0–12.2),p=0,026].

Table 1. Demographic, clinical and laboratory data of patients

Age/ Sex	Disease duration, years	Cutaneous subset	Auto-antibodies	Previous immuno-supresive treatment	RTX cycles	Follow-up after RTX, months	FVC (predicted%)	
							Before RTX	After RTX
52/F	7.0	Diffuse	ANA, Scl-70	CYC, MMF	2	12	44	44
39/M	10.1	Diffuse	ANA, Scl-70	MMF	4	24	75	79
55/F	5.0	Diffuse	ANA, Scl-70	MMF	4	24	75	70
43/F	16.6	Limited	ANA, Scl-70	CYC, MMF	1	6	38	47
50/F	4.6	Limited	ANA, Scl-70	CYC, MMF	4	24	52	57
65/F	13,0	Diffuse	ANA, Scl-70	CYC, MMF	1	6	42	41
48/F	5.7	Limited	ANA, Scl-70	CYC	4	24	67	64
54/F	18.9	Diffuse	ANA, Scl-70	CYC, MMF	2	12	39	57
53/F	15.0	Limited	ANA, Scl-70	CYC, MMF	5	30	53	44
56/F	5,1	Limited	ANA, Scl-70	CYC	1	6	40	45
52/F	8.2	Limited	ANA	–	3	12	59	59
18/F	11.2	Diffuse	ANA, Scl-70	MMF	1	6	63	73
62/F	4.6	Limited	ANA, Scl-70	CYC, MMF	3	18	51	61
54/F	13.1	Limited	ANA, Scl-70	CYC, MMF	5	30	54	67

FVC, forced vital capacity; ANA, antinuclear antibody; Scl-70, antitopoisomerase-1 antibody; CYC, cyclophosphamide; MMF,mycophenolate mofetil; RTX, rituximab.

Conclusions: In this case series of SSc patients treated with RTX, improvement or stabilization of pulmonary functions was observed in most of SSc patients. RTX may be useful in SSc-ILD patients with longer disease duration and resistant to conventional immunsupresive therapies.
Disclosure of Interest: None declared
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AB0605 VITAMIN D SERUM CONCENTRATION IN EUROPEAN SYSTEMIC SCLEROSIS PATIENTS: CORRELATIONS WITH SEASONALITY, ORGAN INVOLVEMENT AND STANDARD ORAL SUPPLEMENTATION

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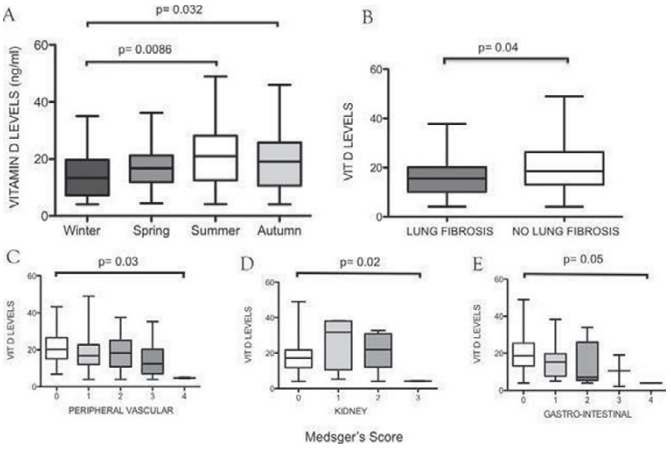
Background: Vitamin D deficiency is reported to interfere with immune responses and to correlate with course and outcome in several autoimmune diseases. In systemic sclerosis (SSc), low 25-hydroxyvitamin D (25(OH)D) serum concentration has been recognized.
Objectives: To investigate relations between 25(OH)D serum concentration and seasonality, clinical parameters as well as standard oral supplementation, in SSc patients.
Methods: 154 SSc patients (mean age 59±15 years, 24.7% diffuse form and 75.3% limited form) were evaluated, at any time of the year, in a retrospective survey. Serum 25(OH)D quantification was performed using the LIAISON 25-OH vitamin D assay (Diasorin, Italy). Pulmonary function test, chest x-ray, lung CT scan, electrocardiography, Doppler echocardiography, renal artery resistive index by eco color Doppler, Dual X-ray absorptiometry, were performed at the time of sample collection. Disease severity scale (DSS) was performed according to Medsger. Drug assumption (glucocorticoids, calcium channel blockers, cyclic intravenous iloprost, endothelin receptor antagonists) and supplementation with vitamin D analogues, were recorded. Non-parametric tests were used for statistical analysis.
Results: Average 25(OH)D serum concentration was found to be 18.7±9 ng/ml (<20 classified as deficiency). A significant difference was observed among seasonal 25(OH)D serum concentration (winter: 14.6±7.8 ng/ml, spring: 17.2±7.9 ng/ml, summer 21.43±10 ng/ml, autumn 20.2±10; p=*0.032*) (Figure 1). A significant correlation was found between 25(OH)D serum concentration and presence/absence of bi-basal fibrotic changes at lung computed tomography (CT) scan (average values: 16.1±8 ng/ml and 20±10 ng/ml, respectively, *p=0.04*). Peripheral vascular *(p=0.03*), kidney (*p=0.02*), gastrointestinal (*p=0.05*) Medsger's DSS parameters also were found to correlate with 25(OH)D serum concentration (Figure 1). Interestingly, no influence of treatment with vitamin D analogues (1,000 UI daily) was found regarding 25(OH)D serum concentration in treated (18.8±10 ng/ml) and in not treated (18.7±9 ng/ml) SSc patients (p=0.81).
Conclusions: In SSc is confirmed a serum 25(OH)D deficiency that we report to be associated with lung involvement, peripheral vascular, kidney and gastrointestinal Medsger's DSS parameters, as well as with seasonality. Supplementation with vitamin D analogues did not influence present results.



Therefore, for successful replacement, supra-physiological oral vitamin D3 doses or programmed UVB light exposure should be considered.
Disclosure of Interest: None declared
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AB0606 MORTALITY ASSOCIATED FACTORS TO IDIOPATHIC INFLAMMATORY MYOPATHIES (IIMS)

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Background: Idiopathic inflammatory myopathies (IIMs) include a group of muscular diseases characterized by the presence of muscle inflammation. The mortality of the IIMs has been estimated between 13 and 50%.
Objectives: To evaluate mortality rate and associated factors in patients with IIMs.
Methods: Retrospective, observational study, where patients with IIMs (Bohan & Peter 1975) were included. Data were obtained from medical records from patients with myopathy (Increase CK, muscle weakness, cutaneous involvement, interstitial lung involvement) evaluated in a reference rheumatology center of Argentina (1992–2016). Descriptive statistics were performed. Chi2 test, Student's test or Mann Whitney as appropriate multivariate logistic regression analysis.
Results: From 102 patients evaluated 89 enter the study, 73% were female. Mean age at diagnosis 48±14 years. Clinical Manifestations: Skin involvement 77% (erythema Heliotrope 51%, rash on the neck and V-sign 60%, back and shoulders 50%, photosensitivity 60%, Gottron's papules 50%, pruritus 33%, erythema peri nail 21%), pulmonary involvement 19% Raynaud 28%, muscle weakness 86%, muscle weakness of the neck 33%, respiratory muscles 13%, myalgias 60% and dysphagia 53%.
Muscle biopsy: performed in 36/89 with pathological findings in 83%, electromyogram performed in 35%. Intensive care unit admission 14/89 (16%). Laboratory: raised CPK 68% with an mean value 3527 IU/ml, raised Transaminase 60%, ANA positive 65%, SSA/RO 25%, Jo1 4.4%, RNP 7%, increased CRP 28% and ERA 59%.
Clinical Subtypes IIMs: Dermatomyositis (DM): 61%, Antisynthetic syndrome (AS): 6%, Myopathy associated with connective tissue disease: 19%, Associated with statins: 4, 4%, Polimyositis: 10%. Association with neoplasia was observed in 15%. Treatments: Corticoids pulses 21%, corticoids 97% (mean starting dose 45 mg meprednisone), methotrexate 77%, hydroxychloroquine 36%, azathioprine 30%, cyclophosphamide 16%, intravenous immunoglobulin 15%, biological 10% and cyclosporine 3%.

Univariate analysis

Variables	Mortality (Odds Ratio)	95% IC	
Male sex	3	1,03–8,4	P<0,039
Respiratory muscle weakness	5,47	IC: 1,4–20,59	p<0,007
ANA positive	6	1,27–27	P<0,01
Neoplasms	3,8	1,1–1,3p	P<0,026
Glucocorticoid pulses	5,7	1,81–17,8	P<0,001
Intravenous immunoglobulin	3,67	1,06–12,6	P<0,03
Serious infections	17	4,6–61,5	P<0, 0000012

Multivariate analysis of logistic regression

Variable	OR	IC (95,0%)		p
Malignant neoplasm	8,785	1,229	62,797	**0.03**
Serious infections	69,168	10,079	474,666	**0.0001**
Glucocorticoid pulses	3,745	0,635	22,092	0.145
Male sex	5,899	1,141	30,504	**0.034**
Intravenous immunoglobulin	0,906	0,100	8,217	0.93
Respiratory muscle weakness	0,524	0,050	5,527	0.59
ANA positive	4,247	0,638	28,259	0.135