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- supressive 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 immunosuppressive 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%, Antisynthet syndrome (AS): 6%, Myopathy associated with connective tissue disease: 19%, Associated with statins: 4, 4%, Polymyositis: 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	1,1–13,3	$P<0,026$
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	$P<0,000012$

Multivariate analysis of logistic regression

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

Mortality was 22.5% (20/89 patients), CI95 (14.3–32.5). Mean time from diagnosis to the event was 18 months. The primary cause of death was sepsis 14/20 (70%).
Conclusions: Mortality of patients with inflammatory myopathies was 22%, and the primary cause was infectious. In the analysis of multiple variables, male sex, presence of neoplasms and serious infectious complications were significantly factors associated with mortality.

References:

[1] J Clin Rheumatol 2016; 22: 51–56.

[2] Med Clin 1999; 112:521–6.

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AB0607 PULMONARY EMBOLISM IN SYSTEMIC SCLEROSIS – ONE YEAR FOLLOW UP

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Background: Risk of pulmonary embolism (PE) in systemic sclerosis (SSc) has been estimated between 2.51–3.47 fold higher when compared to non-SSc patients (pts). Proinflammatory state, vasculopathy and vascular injury may contribute to a prothrombotic state in SSc and increased risk for venous thromboembolism.

Objectives: Calculate frequency and identify possible risk factors for PE among SSc pts; analyse efficacy of longterm anticoagulation (ACO) in these pts.

Methods: We conducted a retrospective analysis of 110 pts with SSc followed in our Rheumatology department and selected those who performed lung ventilation/perfusion scintigraphy (V/Q scan) or CT pulmonary angiography due to worsening dyspnea/fatigue or isolated reduction of the carbon monoxide diffusing capacity (DLCO). We collected demographic features, comorbidities, age at SSc diagnosis, anti-nuclear antibody specificities, dyspnea according to New York Heart Association (NYHA) classes and results of cardiopulmonary exams. PE and related variables were assessed at baseline and after 12 months of ACO.

Results: PE was diagnosed in 12 out of 29 (41.4%) SSc pts that met inclusion criteria, with the majority presenting bilateral peripheral multisegmental defects. Most were females (91.7%), mean age of 59.4 (±12.7) years (yrs). Two thirds were diagnosed with limited SSc with mean disease duration of 13.3 (±12.9) yrs. Mean time between SSc and PE diagnosis was 8.5±8.2 yrs, although one third of the pts was diagnosed within the first year of SSc diagnosis. One patient was taking oral contraceptives and none had thrombophilia, previous surgery or cancer.

One third was classified as having NYHA class≥3, with a mean N-terminal pro-brain natriuretic peptide (NT-proBNP) of 1108pg/mL (19 to 8069). Six pts had concomitant interstitial lung disease (ILD) and 8 had an estimated pulmonary artery systolic pressure (PASP)≥35 mmHg (6 of them had concomitant ILD). From these only 2 had pulmonary hypertension confirmed by right heart catheterization and 1 died.

When comparing SSc pts with and without PE, Scl70 positivity was more common in pts with PE (p=0.041). No significant associations were found between PE and several cardiovascular risk factors.

From the 12 pts with PE, 10 were on longterm ACO:5 on rivaroxaban, 4 on warfarin and 1 on apixaban. Clinical reassessment after 12 months of ACO is shown in figure 1.

Patient	Anticoagulant	Symptoms*	NT-pro BNP	V/Q scan	PASP (mmHg)	DLCO
1	Warfarin	Discrete improvement	Reduction (>100 pg/mL)	Not available	Not available	Identic
2	Rivaroxaban	Identic	Identic	Identic	Identic	Identic
3	Non-pplicable [§]					
4	Rivaroxaban	anticoagulated for < 6 months				
5	Warfarin	Identic	Identic	Identic	Identic	Discrete improvement (29=36)
6	Apixaban	Discrete improvement	Discrete raise (±100 pg/mL)	Decrease in perfusion defects	Worsening (28=40) [¶]	Identic
7	Warfarin	Discrete improvement	Identic	Identic	Improvement (51=30)	Not available
8	Warfarin	Identic	Marked raise (>5000 pg/mL) [¶]	Not available	Not available	Not available
9	Non-pplicable [§]					
10	Rivaroxaban	Identic	Raise (±200 pg/mL)	Identic	Not available	Identic
11	Rivaroxaban	Discrete improvement	Identic	Identic	Not available	Not available
12	Rivaroxaban	anticoagulated for < 6 months				

*Symptoms evaluated were fatigue and dyspnea; § V/Q scan defects are too subtle and the risk is considered to outweigh the benefit of anticoagulation; ¶ Cardiac catheterization demonstrated important coronary disease; ¶ In relation with worsening of ILD

Figure 1 – Patients' reassessment after 12 months of ACO

Conclusions: Our results suggest that PE is frequent in SSc and must be considered in the differential diagnosis of worsening fatigue and dyspnea and/or reduction of DLCO/PASP increase. PE may occur more frequently in Scl70

positive pts and early in disease course, probably due to vasculopathy and vascular injury being more prominent in the early phase of the disease. Although there is no consensus regarding the optimal ACO, the disease's vasculopathy seems to be an important contributor, potentially preventing improvement in perfusion defects, regardless of the anticoagulant used.

Disclosure of Interest: None declared

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AB0608 PREDICTIVE FACTORS FOR LONG-TERM SURVIVAL AND DISEASE PROGRESSION OF SYSTEMIC SCLEROSIS – A LONGITUDINAL ANALYSIS

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Background: Systemic sclerosis (ScS) has unpredictable course and high mortality. Generalised Estimating Equations (GEE) is a technique useful for longitudinal data analysis, using data from all time points and adjusting for within-patient correlation, i.e. correlation between time points within the same patient. GEE does not require a normal distribution of dependent variables, making it attractive for analyzing ScS data.

Objectives: To identify predictive factors for death and unfavorable outcomes.

Methods: Data of ScS patients with ≥2 visits in our EUSTAR centre in 2004–2016 were analyzed. GEE investigated the relationship over time between outcomes (death, digital ulcers (DUs), forced vital capacity (FVC), modified Rodnan skin score (mRSS)) and potential predictors (age, gender, disease duration, cutaneous subset, mRSS at baseline, DUs history, DLCO, left ventricle ejection fraction (LVEF), proteinuria), separately for each predictor and in combined models.

Results: 89 patients (12.4% males, mean±SD age 49.2±12.2 years, disease duration 4.1±7.5 years) were included, with a follow-up of up to 13 years. There were 14 deaths, most due to lung involvement (7/14). In multivariable GEE analysis (Table 1), predictors of death were a shorter disease duration, DUs history, and a lower LVEF. Predictors for FVC decrease over time were diffuse cutaneous subset (dcSSc), younger age and lower DLCO. Younger age, shorter disease duration and higher baseline mRSS were the most important predictors for higher mRSS at follow-up. The only predictor for the development of new DUs was a history of DUs.

Table 1. Prediction factors for death and for evolution over time of parameters reflecting disease severity in ScS

Predictors	Death	DUs	FVC	mRSS
	OR (95% CI)	OR (95% CI)	B (95% CI)	B (95% CI)
Age	1.1 (0.9, 1.3)	0.9 (0.9, 1.0)	0.3 (0.1, 0.6)*	-0.1 (-0.2, -0.01)*
Disease duration	0.8 (0.7, 0.9)***	1.0 (0.9, 1.1)	-0.0 (-0.3, 0.3)	-0.1 (-0.3, -0.03)**
mRSS baseline	0.9 (0.9, 1.1)	1.1 (0.9, 1.1)	0.2 (-0.2, 0.7)	0.4 (0.3, 0.6)***
DLCO	0.9 (0.9, 1.0)	1.0 (0.9, 1.0)	0.5 (0.3, 0.6)***	0.1 (-0.0, 0.0)
LVEF	0.9 (0.90, 0.98)**	0.9 (0.9, 1.1)	0.2 (-0.2, 0.7)	0.0 (-0.2, 0.2)
Male gender	1.9 (0.3, 11.2)	0.5 (0.1, 1.9)	0.7 (-9.3, 10.7)	-1.2 (-3.9, 1.5)
dcSSc	6.8 (0.6, 81.7)	0.7 (0.2, 2.9)	-16.5 (-28.9, -4.9)**	1.7 (-0.1, 3.4)
DUs history	13.1 (3.1, 55.8)***	28.4 (2.3, 356.4)**	3.2 (-2.1, 8.4)	0.2 (-1.4, 1.7)
Proteinuria	1.1 (0.6, 1.8)	0.9 (0.4, 1.9)	2.2 (-2.5, 6.8)	1.7 (-0.1, 3.5)

*p<0.05, **p<0.01, ***p<0.001.

Conclusions: Patients with shorter disease duration, dcSSc, higher mRSS, lower DLCO and LVEF and a history of DUs had a more unfavorable course. GEE is a robust technique for longitudinal data analysis, excellent for identifying prediction factors in ScS.

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AB0609 CORRELATION OF PSYCHOLOGICAL PROFILE (MMPI-II A BDI TESTS) OF SCLERODERMA PATIENTS WITH ORGAN MANIFESTATION AND IMMUNOLOGICAL PROFILE

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Background: There is high prevalence of mood disorders in patients with systemic scleroderma. Psychical distress can influence some of clinical manifestations and can worsen the course of the disease and socioeconomical status of patients.

Objectives: The aim of study is detection of correlations of depression and psychological profile measures by MMPI-II (The Minnesota Multiphasic Personality Inventory) and BDI-II (The Beck Depression Inventory-II) and organ manifestation and immunological profile of patients with systemic sclerosis.