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.
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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 (NTproBNP) 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.
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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 SSc data.
Objectives: To identify predictive factors for death and unfavorable outcomes.
Methods: Data of SSc 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±12.2years, disease duration 4.1±7.5years) were included, with a follow-up of up to 13years. There were 14deaths, 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 difuse 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 SSc

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 SSc.
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AB0609 CORRELATION OF PSYCHOLOGICAL PROFILE (MMPI-II A BDII 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 influent some of clinical manifestations and can worse the course of the disease and socioeconomicall 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.