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 (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.

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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 sclerosis. Psychological 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.

Methods: From March 2015 to July 2017 we examined 42 patients with scleroderma, from which 37 filled the valorisation criteria of MMPI-II test. The data of MMPI-II scales and BDI-II was correlated with antropometric data, form of disease, disease duration, duration of RP, organ major manifestation, comorbidities, use of medication and immunological profile.

Results: In this study 37 patient were included (6 men, 31 women, mean of age = 58.8±10.6y., duration of disease 8.9±9.1y, duration of RP 13.9±13.4y.), 10 patient have diffuse, 27 patient limited form. Subjective perceived depression detected by BDI II test was present in 11 patients (35%, mild 6, moderate 3 and 2 severe). Total BDI-II score correlated with anticentromer Abs (p=0.05), total cholesterol (p=0.008), LDL (p=0.05) and gastrointestinal manifestation (p=0.002). Positivity in domain - Hypochondria, Depression, Hysteria, Psychastenia and Social introversion was highly prevalent (47–13%). This results of MMPI-II suggest a specific psychological profile of SSc patients. There was no correlation for age, subset of disease, cardiovascular manifestation, valvular damage and myositis. Smokers are much more introvert (p=0.012). Positivity of anti Scl70 Ab correlated with incredulity (p=0.038), schizophrenia, hypomania and family disharmony (p=0.046). Positivity of anticentromer antibodies was connected with incredulity (p=0.038), hypomania, alienation to family, egocentrism (p=0.042), psychotic thinking, psychomotoric retardation (p=0.037) and subscales of BDI-II test (Loss of pleasure, Self-Criticism, Self-Dislike). From organ manifestation, pulmonary hypertension correlated with himself degradation (p=0.037). Gastrointestinal manifestation has the same correlation and BDI-II subscale-Guilt Feelings (p=0.042). It is interesting, that absence of lung damage demonstrated with alienation self and family (p=0.049), posttraumatic stress disorder (p=0.025), problems in family (in more scales, p=0.028, 0.03, 0.029), schizophrenia (p=0.03). Patients without digital ulceration have much more neurologic symptoms (p=0.023). Using of antidepressant and anxiolytic medication correlated with a lot of scales in domain of depression, anxiety, social introversion, family distress and self devaluation. Disease duration and duration of RP correlated with psychomotoric retardation.

Conclusions: Patients with scleroderma may have special psychological profile. MMPI-II test shows specific psychologic abnormalities of scleroderma patients. Psychiatric and psychological intervention can be very useful in complex care of these patients.

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AB0610 NAILFOLD CAPILLARY MICROSCOPY AND LOW BODY MASS INDEX IN RAYNAUD'S PHENOMENON PATIENTS

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Background: Underweight patients develop Raynaud's phenomenon (RP) more frequently. [1] In a small study, skin temperature and perfusion in RP patients was positively associated with body mass index (BMI). [2] In clinical practice, we frequently observe an abnormal nailfold capillary microscopy (NCM) in underweight subjects with presumably primary RP, from which the implications are unknown.

Objectives: The aim is to study whether being underweight is associated with the presence of more abnormalities on NCM.

Methods: NCM, assessed by widefield videocapillaroscopy, was retrospectively assessed in consecutive patients with suspected RP. Patients were included when they had negative serology and did not develop any definite connective tissue disease or organ involvement after a maximum of 5 years follow-up. NCM pattern was classified as normal, non-specific, early, active or late, based on the Cutolo patterns. Weight and height were measured for clinical practice and patients were divided by BMI category: underweight (BMI < 18.5 kg/m²), normal weight (BMI 18.5–25 kg/m²), and overweight (BMI > 25 kg/m²). Dilated and giant capillaries were counted as well as the total number of capillaries.

Results: A total of 352 patients were included (median age 40.6 years (24.9–52.9), male/female: 111/241), of which 47 were underweight, 220 normal weight and 85 overweight. Patients who were underweight presented more abnormalities (dilated capillaries, capillary loss or giant capillaries) on NCM, as shown in figure 1. BMI was negatively associated with NCM pattern, number of dilated capillaries and number of giant capillaries (r=-0.247, r=-0.225 and r=-0.221 respectively, all p<0.001) and positively with the total number of capillaries (r=0.260, p<0.001).

Conclusions: In addition to well-known thermoregulatory abnormalities, clear NCM pattern abnormalities are more frequently observed in underweight patients with primary RP. BMI was negatively associated with the number of dilated capillaries and positively with capillary density. These data potentially suggest that damage to the microvasculature in underweight patients with primary RP may occur independently of the presence or development of a CTD and may be explained by other mechanisms.

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AB0611 MYCOPHENOLATE MOFETIL FOR THE TREATMENT OF INFLAMMATORY MYOPATHY RELATED INTERSTITIAL LUNG DISEASE: A SYSTEMATIC REVIEW

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Background: Inflammatory myopathies (IIM) are a heterogeneous group of rare autoimmune rheumatic diseases in which extramuscular manifestations, especially interstitial lung disease (ILD), are common and may occur in the absence of muscle symptoms. ILD has been identified as a marker of poorer prognosis, with sparse, mainly observational evidence, supporting the use of immunosuppressant therapies.

Despite the lack of clinical trial evidence, corticosteroids are considered first-line therapy. Cyclophosphamide is frequently used for severe disease and Azathioprine as maintenance therapy. Mycophenolate Mofetil (MMF) has emerged as a promising treatment, encouraged by data published from a randomised controlled trial comparing Cyclophosphamide with MMF in patients with systemic sclerosis and ILD, which showed similar efficacy and better tolerability in the MMF group (1).

Objectives: To examine the evidence supporting the use of MMF in patients with IIM associated ILD.

Methods: An electronic literature search was performed using the Ovid platform. Population: Adults diagnosed with IIM according to validated criteria (2, 3) or antisynthetase syndrome (defined by the presence of anti-synthetase antibodies), with ILD demonstrated by HRCT or lung biopsy and treated with MMF, with extractable outcomes regarding ILD. Outcomes were recorded as positive (improvement/stabilisation) and negative (deterioration) as defined by the American Thoracic Society consensus guidelines (4).

Results: 506 initial citations were identified, with 23 studies included in the review comprising a total of 82 patients. Median follow up was 12 months, median age 55 years. There was sufficient data to apply American Thoracic Society criteria in 37 patients: improvement/stabilisation n=29, deterioration n=8. Although the criteria could not be applied in 45 patients, improvement/stabilisation was reported in 44 with deterioration in one patient. A non-significant trend to better outcomes was observed when MMF was prescribed as first line therapy, in Jo-1 positive patients and non-Unusual Interstitial Pneumonia pattern. Mean Prednisolone dose reduced from 40 mg/day to 10.1 mg/day. No significant toxicity was reported.

Conclusions: Mycophenolate Mofetil is a promising drug with encouraging results, but randomised controlled clinical trials are needed to prove its efficacy.

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