(61.7% vs 24.9%), myopathy (21.3% vs 10.2%) and pulmonary hypertension (34.8% vs 22.3%). Significantly more SSc ILD patients were treated with cyclophosphamide, mycophenolate mofetil and azathioprine. Survival Kaplan-Meier curve patients demonstrate reduced survival in patients with ILD ($p < 0.01$). Five years survival rates between years 2000 and 2015 have not changed significant. Mortality risk assessed with Cox regression analysis in the whole group was significantly higher in males, Arabs, DcSSc, elder age, heart and muscle involvement, and treatment with CYC. In the ILD group, the mortality was significantly higher in Arabs (3.3 times), elder age (8.9 times), presence of PAH (3.1 times) and treatment with CYC (2.8 times) compared to patients without ILD.

Conclusion: In our SSc cohort, ILD affected about third of patients and had major impact on patients' outcome. Male gender, Arab nationality, elder age, DcSSc, topoisomerase antibodies, heart and muscle involvement were significantly associated with worst prognosis. Despite active approach to assessing and treatment, survival rates of patients with SSc and SSc-ILD have not improved in last decades. Enrichment of the cohort with severe patients to a tertiary center due to reference bias and low efficacy of existed immunomodulatory drugs in SSc and in SSc related ILD particularly, could explain our results.

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POS0872

CLINICAL FEATURES AND OUTCOME OF 1054 PATIENTS WITH SYSTEMIC SCLEROSIS: AN ANALYSIS OF THE PORTUGUESE REUMA.PT REGISTRY FOR SCLERODERMA (REUMA.PT/SSC)

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Background: Systemic sclerosis (SSc) may present distinctive manifestations and survival in different ethnic and geographic groups.

Objectives: To describe the clinical features, treatments, and survival of adult SSc patients registered in Reuma.pt/SSc.

Methods: Demographic features, SSc subsets, fulfilment of classification criteria, clinical and immunologic characteristics, comorbidities, medication and deaths were reviewed. Survival was calculated for patients included in the registry within the first 2 years of diagnosis.

Results: In total, 1054 patients were included, 87.5% female, mean age at diagnosis 52.7 ± 14.8 years. The most common subset was limited cutaneous (lc)SSc (56.3%), followed by diffuse cutaneous (dc)SSc (17.5%), preclinical SSc (13%), overlap syndrome (9.8%) and SSc sine scleroderma (3.3%). Raynaud's phenomenon (93.4%) and skin thickening (76.9%) were the most observed manifestations. Gastrointestinal (62.8% vs 47.8%), pulmonary (59.5% vs 23%) and cardiac (12.8% vs 6.9%) involvement were significantly more prevalent in dcSSc compared to lcSSc (Table 1). 52.5% of patients were ACA positive and 21% anti-topoisomerase positive, with significant differences between lcSSc and dcSSc. One third of patients was treated with immunomodulators, 53.6% with vasodilators, 23% received glucocorticoids and 2.3% biologics.

During the median follow-up 12.4 years, 83 deaths (7.9%) were verified. The overall 1, 2 and 5 years survival was 98.0%, 96.8% and 92.6% respectively, without significant differences between lcSSc and dcSSc (Figure 1).

Conclusion: Reuma.pt/SSc register is useful in routine patient monitoring and contributes to improve knowledge about this rare and complex disease. Clinical features of Portuguese SSc patients are similar to what has been described in other populations although the overall 5-year survival in recently diagnosed patients appears to be higher than previously reported.

Table 1. Cumulative clinical and immunologic characteristics of Portuguese SSc patients

Clinical and immunologic features	Total N=1054	Limited cutaneous SSc N= 576 (56.3%)	Diffuse cutaneous SSc N=180 (17.5%)	P value
Skin involvement – N(%) N=987	688 (90.6)	525 (90.7)	180 (100)	<0.01
Skin thickening + – N (%) N= 962	680 (76.9)	512 (88.9)	180 (100)	<0.01
Digital ulcers – N (%) N=970	325(33.5)	186 (34.7)	4 (51.5)	<0.01
Raynaud's Phenomenon – N (%) N=1010	943 (93.4)	539 (95.7)	157 (92.4)	0.06
Musculoskeletal involvement – N(%) N=972	346 (45.6)	247 (42.7)	99 (55)	<0.01
Cardiac involvement –N(%) – N=924	71 (7.7)	36 (6.9)	19 (12.8)	0.02
Renal involvement –N(%) – N= 917	17 (1.9)	8(1.5)	6 (4.1)	0.07
Gastrointestinal involvement - N(%) N=933	508 (48.2)	277 (47.8)	113 (62.8)	<0.01
Pulmonary involvement – N(%) N=915	261 (28.5)	119 (23)	88 (59.5)	<0.01
PAH – N(%) N= 871	14 (1.6)	10 (2)	1 (0.7)	0.23
Interstitial lung disease – N(%) N=765	218 (28.5)	100 (22.7)	75 (57.7)	<0.01
Antinuclear antibodies - N(%) N=1040	934 (89.8)	522 (90.2)	154 (88.5)	0.57
Anti-centromere – N(%) N= 1027	540 (52.6)	383 (67.1)	16 (9.5)	<0.01
Anti-Scl70 – N(%) N=1020	214 (21)	12 (3.3)	104 (60.1)	<0.01
Anti-RNA polymerase III – N(%) N=710	25 (3.5)	12 (3.3)	7 (5.6)	0.38
Comorbidities				
Hypertension – N(%) N=431	117 (27.1)	76 (29.7)	67 (20.7)	0.1
Hyperlipidemia – N(%) N=431	71 (13.4)	72 (12.2)	24 (15.9)	0.08
Neoplasia – N(%) N=1054	29 (2.8)	12 (2.1)	7 (3.9)	0.14

PDE-5 (phosphodiesterase-5); PPIs (proton pump inhibitors); PAH-Pulmonary arterial hypertension confirmed by right heart catheterization. Immunomodulators includes Methotrexate, Leflunomide, Hydroxychloroquine; Azathioprine, Mycophenolate Mofetil and Cyclophosphamide; * Does not include sclerodactyly.

Figure 1. Panel A - Survival in years from diagnosis of patients with SSc included in Reuma.pt in the first 2 years of disease (N=472). Panel B - survival according to SSc subset (lcSSc and dcSSc).

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POS0873

IMPROVEMENT IN MODIFIED RODNAN SKIN SCORE (MRSS) IN SYSTEMIC SCLEROSIS PATIENTS TREATED WITH RITUXIMAB: A SYSTEMATIC REVIEW AND META-ANALYSIS OF THE SCIENTIFIC LITERATURE

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Background: Systemic sclerosis (SSc) is a chronic autoimmune disease of the connective tissue characterized by vascular disease and fibrosis in different organs and systems such as lung and skin (1). Recently, several case reports and small series of patients reported on the efficacy of rituximab in SSc, showing a possible improvement in skin and lung affectations (2). However, registered clinical trials are lacking to determine factors associated with response, maintenance regimen, and long-term efficacy of rituximab in SSc.

Objectives: To analyze the efficacy of Rituximab in the treatment of skin fibrosis using the changes of the modified Rodnan Skin Score (mRSS) of patients diagnosed with systemic sclerosis from the data published in Registered Clinical Trials (RCTs) in the scientific literature.

Methods: We perform a systematic review and a meta-analysis using the main electronic databases to locate all the articles available so far: Medline, Embase, Cochrane Library and Web of science and ACR and EULAR abstracts congress were extracted to assess efficacy outcomes. That efficacy was measured based on the variation of mRSS at 12, 24 and 48 weeks for patients treated with Rituximab versus patients treated with another drug or placebo.

Results: 3 RCTs contained data regarding mRSS at week 12 of treatment with Rituximab. The estimated SMD was -1.071 (95% CI -1.608, -0.535 [$p < 0.001$]) with a non-significant P value in the Egger Test ($P = 0.703$) and non-significant heterogeneity through I² ($I^2 = 0.00\%$). 9 studies contained data regarding mRSS at week 24 of treatment with Rituximab. The estimated SMD was -1.743 (CI95% -2.622, -0.864 [$p < 0.001$], see image below) with a non-significant P value in the Egger Test ($P = 0.072$) and significant heterogeneity through I² ($I^2 = 86.6\%$). Meta-regression analysis could not be performed to assess such heterogeneity, due to the lack of comparable data.

8 RCTs contained data regarding mRSS at week 48 of Rituximab treatment. The estimated SMD was -1.327 (CI95% -2.018, -0.636 [$p < 0.001$]) with a significant P value in the Egger Test ($P = 0.018$), estimating that there may be publication bias in the studies analyzed and significant heterogeneity by I² ($I^2 = 85.2\%$). Meta-regression analysis could not be performed to assess such heterogeneity, due to the lack of comparable data.

Conclusion: Our meta-analysis shows that Rituximab treatment in patients affected with systemic sclerosis shows efficacy in the treatment of cutaneous fibrosis measured by the mRSS, turning this molecule into a potential drug to add to the therapeutic armamentarium of systemic sclerosis. However, more studies are necessary to try to elucidate whether this change is powerful enough to become the new gold standard for the treatment of systemic sclerosis skin involvement.

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POS0874

CLOT LYSIS TIME PREDICTS RECURRENT DIGITAL ULCERS IN SYSTEMIC SCLEROSIS AFTER ONE YEAR OF FOLLOW-UP: A NESTED CASE-CONTROL STUDY

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Background: Digital ulcers (DU) are a common visible manifestation of vasculopathy in systemic sclerosis (SSc), which could be recurrent and associated with disability and mortality (1). Although vasculopathy is connected with impaired coagulation/fibrinolysis system and aberrant expression of adhesion molecules, there are few data about their role in developing recurrent DU.

Objectives: To evaluate the possible role of Fibrin generation/Fibrinolysis parameters and adhesion molecules in the prediction of new ischaemic DU onset during a 1-year follow-up and their impact on the time to new DU onset (TD).

Methods: From 58 consecutive patients with SSc who fulfilled the 2013 ACR/EULAR SSc criteria and have never been treated with endothelin receptor antagonist, phosphodiesterase 5 inhibitors or prostanoids, a total of 38 patients with

ever had DU, either active at inclusion or in past, were enrolled in a prospective cohort study. Each patient was given a "DU diary". Demographic, clinical and serological data were recorded. The serum concentration of ICAM1 and E selectin were measured by ELISA. Haemostatic potential parameters: overall haemostasis (OHP), overall coagulation (OCP) and overall fibrinolysis (OFP) potential were assessed. Maximum absorbance (Cmax), reflects the fibrin clot density and clot lysis time (Lys50t0, time from initiation of clot formation to the time at which a 50% fall in absorbance from Cmax in the lysis assay), reflects fibrinolytic susceptibility, were calculated (2). Fibrin structure was visualised using scanning electron microscopy (SEM).

Results: Over the follow-up period, 18 patients (45.5%) developed new DU with the average TD of 7.4±2.9 months. There was no difference in ASA and CCB treatment among two groups ($p > 0.05$). The OFP value was significantly decreased ($p < 0.01$), Lys50t0 prolonged ($p < 0.05$), while OHP was increased ($p < 0.05$) in patients experienced new DU. Lys50t0 showed good validity in identifying patients with new DU onset (AUC 0.683 95% CI 0.5 - 0.9). By multivariate analysis including clinical data in model the Lys50t0 (HR 1.2, 95% CI 1.1-1.3, $p = 0.018$) and active DU at enrollment (HR 9.6, 95% CI 1.4-66.8, $p = 0.022$) were identified as independent risk factors for the occurrence of new DU. TD was inversely correlated with ICAM1 ($p < 0.001$), E selectin ($p < 0.05$), Cmax ($p < 0.05$) and Lys50t0 ($p < 0.001$). Model explaining 81.6% of the TD variability included Lys50t0 ($\beta = -0.55, p = 0.003$), E selectin ($\beta = -0.44, p = 0.014$) and fibrinogen ($\beta = -0.49, p = 0.017$). SEM revealed denser fibrin clots with thinner fibres in group experienced new DU compared to clots formed in plasma of patient without DU.

Conclusion: Our results provide evidence that impaired fibrinolysis has critical role in the progression of microvascular disease, identifying clot lysis time as a strong predictor in the onset of new digital ulcers in Systemic sclerosis. The higher level of E selectin and the longer lysis time are, the shorter is time until the onset of new digital ulcer. These results could be used in selection of patients at high risk of developing recurrent digital ulcer and therefore allow earlier therapeutic intervention.

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Figure 1. SEM images of fibrin network in 1 representative sample from a SSc patient with (A) and 1 from patient without (B) new DU onset

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POS0875

MYOSITIS-SPECIFIC ANTIBODIES IN A RETROSPECTIVE SINGLE-CENTER OBSERVATIONAL STUDY

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Background: Myositis-specific antibodies (MSA) are highly specific and useful to classify patients as having syndromes with distinct clinical features and prognosis. MSA are almost always mutually exclusive and quite specific, adding value as a useful biomarker for diagnosis. Although individual autoantibodies aren't sensitive enough to detect the full spectrum of idiopathic inflammatory myopathies (IIM), the sensitivity of a myositis panel is increasing as more autoantibodies are discovered, and as better assays become available.

Objectives: We aimed to analyze the usefulness of a myositis-specific immunoblot for the diagnosis of IIM in a hospital cohort from January 2019 to December 2020. We also seek to correlate immunological findings with the risk of associated interstitial lung disease (ILD), cancer, or death.