the cumulative responses revealed a relapsing and remitting course in 45.9%. Outcome predictors in univariate analysis were Jo-1 status, presence of arthritis, interstitial lung disease and pericardial effusion at baseline. On multivariate analysis, absence of pericardial effusion ($p=0.011$) and interstitial lung disease ($p=0.067$) at baseline were found to be predictors of complete response. Disease free survival probability estimated at 5 years and 10 years was 91.6% and 72.4% respectively. Estimating the probability gender wise, males achieved disease free status earlier than females.

Conclusion: A favorable clinical and functional outcome was seen in a significant proportion of these patients with IIM on long term follow up. Pericardial effusion and ILD were identified as predictors of poor clinical outcome.

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AB0597

PULMONARY FUNCTION IN PATIENTS DIAGNOSED OF EARLY SYSTEMIC SCLEROSIS: A NEW TOOL FOR SYSTEMIC SCLEROSIS CLASSIFICATION?

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Background: Interstitial lung disease (ILD) is a frequent complication of systemic sclerosis (SSc) and is often progressive and has a poor prognosis. A restrictive ventilatory defect could suggest ILD either alone or in combination with pulmonary arterial hypertension.

Nowadays, Early-SSc is well defined as preliminary stage of SSc. Patients who meet criteria for Early-SSc could benefit from an early diagnosis of pulmonary involvement.

Objectives: Our aim was to assess the pulmonary function in patients diagnosed of Early SSc.

Methods: Retrospective observational study of a wide and unselected series of patients diagnosed as Early-SSc from a single university hospital from 2012 to 2019. Patients were classified as Early-SSc following Le Roy criteria. Despite this, patients already did not meet 2013 ACR/EULAR classification criteria for SSc. We reviewed pulmonary function through conventional spirometry and diffusing capacity of lung for carbon monoxide (DLCO).

Results: We included 56 patients with a mean age of 52.3 ± 12.1 years (96.4% women; 3.6% men).

At the diagnosis of Early-SSc, no one of our patients evidenced a restrictive ventilatory pattern. DLCO was below normal limits in 18 patients (32.1%). Small airway obstruction expressed according decreased maximal (mid-) expiratory flow (MMEF) 25-75 was present in 24 patients (42.8%).

After a mean follow-up period of 38.3 ± 2.4 months, 29 (51.8%) patients fulfilled 2013 ACR/EULAR criteria. The average time between diagnosis of Early-SSc and achieve SSc classification was 24.4 ± 1.8 months. The remaining 27 patients continued classified as Early-SSc.

An analysis of the subgroup of patients which progressed to SSc showed that DLCO was decreased in 15 of those 29 patients (51.7%) and 18 of 29 patients (62.1%) presented decreased MMEF 25-75. Comparing with the subgroup of patients which not progressed to SSc were significant differences (Decreased DLCO: 51.7% vs 11.1%; $p=0.02$ and decreased MMEF 25-75: 42.8% vs 22.2%; $p=0.05$).

The analysis of pulmonary function of the subgroup of patients continued classified as Early-SSc after follow-up period did not show significative changes after follow-up.

Conclusion: In our study, a third of the patients classified as Early-SSc presented at diagnosis abnormal values of DLCO and/or signs of small airway obstruction without the presence of a restrictive ventilatory pattern. Moreover, this pulmonary dysfunction was significantly more frequent in patients who progressed to definitive SSc. Patients which remains classified as Early-SSc did not experience significative changes.

Our results support the concept that pulmonary function was impaired in Early-SSc and that I should probably be considered for future Early-SSc classification criteria.

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AB0598

THE INCREASING USE OF IMMUNOSUPPRESSANTS IN EARLY SYSTEMIC SCLEROSIS

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Background: Immunosuppression (IS) remains the main treatment for progressing skin involvement, active interstitial lung disease (ILD) and underlying inflammatory joint (IJ) or muscle disease in systemic sclerosis (SSc).

Objectives: This study investigated the pattern and trends in immunosuppressive agent use in patients with early SSc diagnosed before and after 2007 to determine whether the changes in the preferred type and combination of IS, timing and predictors of administration took place over the past decade.

Methods: 397 SSc patients from Canadian Scleroderma Research Group (CSRG) database (183dcSSc, 214lcSSc) who had baseline and follow-up visits within 3 years (1.8 ± 0.8) after disease onset were included: 82% females, age at diagnosis 53 ± 13 years, disease duration 1.6 ± 0.8 years. Organ involvement was assessed by modified Rodnan skin score, Medsger Disease Severity Score (DSS) and CSRG definitions using bivariate, chi-squared, ANOVA, and adjusted regression analyses.

Results: 115 dcSSc patients (63%) and 62 lcSSc (29%) received IS, most commonly methotrexate (MTX) (72% dcSSc and 52% lcSSc), followed by mycophenylate mofetil (MMF) and cyclophosphamide (CYC). Within the patients receiving IS, monotherapy prevailed (77% dcSSc and 68% lcSSc); CYC and azathioprine were the preferred choice of IS more frequently in lcSSc compared to dcSSc ($p<0.006$ and $p<0.02$, respectively). In dcSSc, IS were predominantly prescribed at years 2 and 3 after the onset of first non-Raynaud's phenomenon (RP) manifestation, when about half of the patients received IS. The proportion of lcSSc patients receiving IS was significantly lower and distributed more equally through the first three years. After 2007, dcSSc patients received IS more often (74% vs 50%, $p=0.001$), especially MTX ($p=0.02$) and MMF ($p<0.05$), and earlier (peaked at 2 years after disease onset)(Table 1).

Table 1. Proportion of patients receiving immunosuppressive treatment at each year after disease onset in SSc diagnosed before and after 2007.

Years after the first non-RP symptom	lcSSc				
	Before 2007		After 2007		
	Total N of pts seen at each year	% receiving immune suppressives	Total N of pts seen at each year	% receiving immune suppressives	P-value
1	13	15	47	17	>0.9
2	24	21	82	18	0.772
3	49	14	107	14	>0.9
	dcSSc				
1	24	29	40	43	0.286
2	51	26	65	65	0.0001
3	62	45	63	54	0.325

IS administration was associated with male gender, ILD, a-Scl-70 positivity, ACA-negativity and IJ disease in lcSSc, and with ACA-negativity and a higher mRSS in dcSSc. Multivariate logistic regression analysis showed that IS treatment could be predicted by ACA-negativity in lcSSc patients (Exp(B) 0.317, $p=0.012$) and younger age in dcSSc patients (Exp(B) 0.974, $p=0.002$).

Conclusion: Over the past decade, there has been a trend to prescribe IS more often, especially MTX, and earlier in dcSSc patients. MMF has gained favour over CYC. Autoantibody status was the most consistent predictor whether a patient is likely to take IS over the course of the disease.

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AB0599

TREATMENT OF REFRACTORY DERMATOMYOSITIS WITH TOFACITINIB

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Background: Dermatomyositis (DM) is a systemic, autoimmune disease affecting the skin and proximal skeletal muscles. A subset of DM patients present with subclinical or resolved muscle involvement but continue to have skin disease. In these cases, first and second line treatments including glucocorticoids are sometimes insufficient for controlling the disease, necessitating escalation of treatment. Several recent studies have investigated the response of Tofacitinib, an oral Janus Kinase inhibitor approved for the treatment of rheumatoid arthritis, in DM patients and patients with inflammatory skin diseases.

Objectives: Due to the reported ability of JAK inhibitors to suppress type 1 interferon (IFN) signaling, which is suspected to be upregulated in DM, we evaluated the efficacy of treatment with Tofacitinib in four refractory DM patients.

Methods: Four patients with dermatomyositis without evidence of current muscle involvement began treatment with Tofacitinib 11 mg daily after they had failed or had adverse effects to first and second line immunosuppressive agents. Their medical records were reviewed at 0, 3, and 6 months, with improvement measured using the Cutaneous Dermatomyositis Disease Area and Severity Index (CDASI) activity score. Throughout their treatment they were additionally monitored for improvement in markers of inflammation and the necessity for concomitant treatments. Patients were monitored for adverse effects to Tofacitinib treatment.

Results: All four patients within the case series showed significant improvement of their cutaneous disease activity (CDASI scores improved by 8-15 points) over the first 6 months, with three of the four having achieved minimal clinically improved difference of ≥ 5 point by three months. Based on the CDASI, three of the cases' disease classification changed from moderate-to-severe disease to mild disease. The last patient initially presented with mild disease. Other outcomes noted included improved pruritus in 3 patients and improvement of calcinosis in 1 patient. One patient was additionally able to stop concomitant treatment with prednisone and IVIG at 3 and 6 months, respectively. This patient had been on daily prednisone for 4 years. Only other patient on prednisone was on low dose of 3mg daily. The patients all had normal muscle exams prior to treatment with Tofacitinib. No worsening muscle involvement or adverse effects were noted with Tofacitinib use.

Conclusion: Tofacitinib is believed to play a role in the inhibition of IFN signaling pathways that are overactive in dermatomyositis. All four patients

Gender/ Age/ Ethnicity	DM Subtype / disease duration	Previous Treatment	Concomitant Treatment with TOF	CDASI Activity	CDASI Activity 3 months	CDASI Activity 6 months	Other Outcome
M/ 55/ Caucasian	Amyopathic DM; 5 years	AZA, HCQ, IVIG, MTX, MMF, Prednisone, RCI, RTX, TAC	HCQ	12	5	4	None
F/ 37/ Caucasian	Amyopathic DM; 7 years	AZA, HCQ, IVIG, MTX, MMF, Prednisone, RCI, TAC	Prednisone, IVIG, MTX	15	8	5	Pruritus and CRP improved
F/ 60/ Caucasian	Amyopathic DM; 4 years	AZA, MTX, Prednisone	Prednisone, MTX	17	15	5	Pruritus improved
F/ 47 African American	Classic DM; 5 years	AZA, colchicine, HCQ, IVIG, MTX, MMF, pamidro- nate Prednisone, RCI, RTX, TAC	IVIG, Colchicine	22	8	7	Pruritus and calcinosis improved

Abbreviations: CDASI, Cutaneous Dermatomyositis Disease Area and Severity Index; AZA, azathioprine; CRP, C-reactive protein; DM, Dermatomyositis; HCQ, hydroxychloroquine; IVIG, intravenous immunoglobulin; MTX, methotrexate; MMF, mycophenolate mofetil; RCI, repository-corticotropin injection; RTX, rituximab; TAC, tacrolimus; TOF, tofacitinib;

within this retrospective study showed significant improvement of cutaneous disease with Tofacitinib use.

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AB0600

THE EFFECTS OF HYPERBARIC OXYGEN THERAPY TO QUALITY OF LIFE AND STATE OF MICROCIRCULATION IN PATIENTS WITH SYSTEMIC SCLEROSIS - A PILOT STUDY

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Background: Many treatments have been tried in therapy systemic sclerosis (SSc) patients but use of hyperbaric oxygen therapy (HBOT) is very limited.

Objectives: To assess the effects of HBOT to quality of life and state of microcirculation in SSc patients.

Methods: 18 female patients aged 29-68 years (mean 57 years) with limited SSc and digital or leg ulcers were included in this work. The HBOT protocol comprised 20 sessions 5 day/weekly, 60 min, 100% oxygen at 2.2 ATA. The treated patients were evaluated at baseline and after 20 HBOT sessions. Evaluation consisted of physical examination, capillaroscopy, pulmonary function tests, biochemical analyses, socio-demographic and clinimetric questionnaires: Systemic Sclerosis Questionnaire (SySQ) and Health Assessment Disability index Questionnaire (HAQ-DI).

Results: Mean value [before: after, mean (range)] for SySQ [15.5 (4-48) vs 9.0 (3-31)], HAQ-DI [0.60 (0-2.88) vs 0.35 (0 -1.75)], erythrocyte sedimentation rate [21 (4-42) vs 12 (3-27)], forced vital capacity (96.61±14.44% vs 115.94±16.69%), diffusing lung capacity of carbon monoxide (73.61±6.63% vs 87.33±9.30%) significantly improved after HBOT sessions (p<0.001). There was no significant changes in the total number of capillaries (325 vs 338, p=0.235), mean number of enlarged capillaries (21 vs 27, p=0.182), giant capillaries (14 vs 14, p=0.235) and ramified/bushy capillaries (14 vs 13, p=0.178) before and after HBOT. All patients had digital ulcers, and 5 patients had bilateral lesions (digital and leg ulcers). Mean size of ulceration before HBOT was 12x11mm, and after therapy was 4x4mm (p<0.001). Three patients had digital gangrene. Amputation was not require for any.

Conclusion: Our data confirm the efficacy of HBOT in treating SSc patients. Further studies are required to evaluate the protocol and to understand the duration of the clinical effect.

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