measured by master instructor was 10.5 (9.0). Mean (SD) difference between skin scores by physicians and master instructor was 7.7 (9.5) units. In the univariable analysis, video education significantly reduced the difference from the gold-standard score ($\beta = -1.96$, 95% CI -3.83 to -0.10) whereas live demonstration did not showed additional enhancement in scoring skill. Effect of education program was significantly different according to the physician's status and patient's disease type (diffuse vs. limited). In addition, male patient, shorter disease duration and higher gold-standard skin score was associated with more accurate skin scoring irrespective of the education. In the multivariable analysis where above clinical factors were adjusted, video education also led to significantly accurate skin scoring (table 1). When the educational effect was stratified by individual site of examination, face and distal extremities showed greater enhancement of scoring accuracy whereas difference from gold-standard score in proximal extremities was rather increased. ICC of physicians' skin scores was acceptable over all scoring times (0.63 to 0.88) but was not significantly changed after the education.

Conclusions: The mRSS education program can significantly enhance the accuracy of mRSS, which is mainly achieved by video education.

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AB0775

CHARACTERISTICS OF PATIENTS WITH SCLERODERMA (SSC) TREATED WITH VARIOUS DRUGS IN THE CLINICAL ASSESSMENT AND TGF β AND IL13 CONCENTRATION IN COMPARISON TO THE HEALTHY GROUP

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Background: Scleroderma (SSc) is a rare multisystem chronic disease the treatment of which is still challenging. Until now, there is no effective therapy that can modify the overall disease course. However, the main aim of Ssc treatment is directed toward managing organs involvement and providing symptomatic relief. Effective drug therapy should inhibit three components of the disease: tissue fibrosis, vascular abnormalities and autoimmunity. Moreover, potential drug needs to be considered in the context of specific four subsets of the disease: fibroproliferative, inflammatory, limited, and normal-like. It was shown that various subsets could have different profile of specific cytokines: TGF β – associated with fibroproliferative and inflammatory type of disease and IL13 – mediator of fibrotic and vascular pathology.

Objectives: The aim of the study was to assess the level of TGF β and IL-13 in SSc pts with various treatment regimen comparing to healthy control.

Methods: 55 patients (71% were women) with SSc diagnosed according to EULAR/ACR 2013 criteria were divided into 5 treatment groups: 1st group – 8 pts treated with methotrexate (MTX), 2–13 pts with mycophenolate mofetil (MMF), 3–5 pts with cyclophosphamide (CYC), 4th group- 7 pts with azathioprine (AZA), 5th group- 22 pts without immunosuppressive treatment. All patients have been treated based on a scheme required for organs involvement in accordance with the update of EULAR recommendations from 2016. Mean age of patients was 54.56 $\pm$ 13.97. The blood and serum samples have been collected for basic examination. TGF β and IL13 concentration in serum was quantitated by ELISA. Differences in cytokine concentration were determined using non-parametric Kruskal-Wallis test. The level of statistical significance was set at $p < 0.05$. The modified Rodnan Skin Score (mRSS) examination were taken by one assessor at the beginning of the study and six months later. There were measured DLCO, HRCT, echocardiography and capillaroscopy.

Results: In 82% pts positive antinuclear antibodies have been revealed: in 16% positive results for CENPB were obtained and in 44% for Scl70. Capillaroscopy shown in 23% of pts early pattern, in 38% – active pattern, and in 27% – late pattern. 32% of SSc pts had confirmed pulmonary fibrosis, while 90% – Raynaud's syndrome. The median of mRSS in CENPB (+) pts was 5, ⁴⁻²⁴ in Scl70 (+) pts – 12. ²⁻³⁰ Statistically significant differences were found between IL13 and TGF β levels in patients treated with immunosuppressants and healthy subjects. There was no correlation between IL13 or TGF β with lung fibrosis progression or skin involvement.

Abstract AB0775 – Table 1. Comparison of serum level of IL 13 and TGF β in SSc pts

	MTX (n=8)	MMF (n=13)	CYC (n=5)	AZA (n=7)	patients without immunosuppressive treatment (n=22)	healthy controls n=16	p value
TGFβ							<0,0001
Minimum	179.6	175.4	203.5	96.15	104.4	44	
Median	226.6	272.3	273.2	272.3	202.5	192	
Maximum	414.6	385.3	359.5	356.6	389.9	323	
Mean	267.6	284.8	270.8	271.2	228.5	150.9	
Std. Deviation	90.23	67.15	60.52	90.23	79.71	70.58	
Std. Error	31.9	18.62	27.06	34.1	16.99	17.65	
IL 13							<0,0001
Minimum	0	0	0	0	0	156	
Median	90	172.7	0	172.4	175.8	219	
Maximum	185	191	187	191.3	224.4	389	
Mean	90.14	123.6	73.39	103.3	115	228.3	
Std. Deviation	96.42	85.98	100.5	96.81	96.1	53.53	
Std. Error	34.09	23.85	44.96	36.59	21.13	6.572	
		23.85	44.96	36.59	21.13	6.572	

Conclusions: In conclusion, our findings indicate that IL 13 and TGF β are characteristic cytokines in scleroderma, but these parameters did not correlate with severely progressive course of SSc.

Disclosure of Interest: None declared

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AB0776

MUSCLE ULTRASONOGRAPHY: A POTENTIAL NEW DIAGNOSTIC TOOL FOR INFLAMMATORY MYOPATHIES

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Background: Quantitative muscle ultrasound (QMUS) imaging has proven to be a useful, non-invasive technique to visualise normal and pathological skeletal muscle tissue. ¹ Electromyography (EMG) findings are not always disease specific in patients suspected of idiopathic inflammatory myopathies (IIM).

Objectives: To assess diagnostic value of QMUS in patients suspected for an IIM and to compare results with EMG.

Methods: In 57 patients, suspected for IIM, panel diagnosis blinded for QMUS was used as reference standard. QMUS results were used to classify patients according an ultrasound neuromuscular disorder (NMD) algorithm (normal/borderline/abnormal). The predictive value of QMUS and EMG was assessed in a two by two table and a multivariate logistic regression model.

Results: Twenty-two patients (39%) were diagnosed with IIM; 8 polymyositis, 4 dermatomyositis, 4 necrotizing myopathy, 3 inclusion body myositis and 3 non-specific myositis. Sixteen patients were classified with other NMD. We found an increased echointensity of the sternocleidomastoid, biceps, forearm flexor and tibialis anterior in the IIM group. Sensitivity, specificity, positive and negative predictive values (PPV/NPV) were 82%, 51%, 51%, 82% for ultrasound NMD algorithm and 63%, 64%, 50%, 75% for EMG. Multivariate analyses showed area under the curve (AUC) (0.81) (0.69–0.92) for ultrasound NMD algorithm, EMG (0.79) (0.67–0.92) and ultrasound NMD algorithm plus EMG (0.82) (0.70–0.93).

Predictor	Model A	Model B	Model C
Age ≥ 50 year	2.97 (0.65–13.59)	2.88 (0.62–13.28)	2.98 (0.62–14.21)
Serum CK ≥ 2 x upper limit	7.52 (1.57–36)	8.76 (1.55–49.55)	7.35 (1.22–44.21)
Muscle ultrasound			
Total echointensity of proximal muscles/measured muscle			
Total number of affected proximal muscles			
Distal muscles affected (yes/no)			
NMD algorithm:			
- No NMD	Reference:		Reference:
- Borderline presence of NMD (yes/no)	1.61 (0.13–18.87)		1.62 (0.13–19.57)
- Presence of NMD (yes/no)	3.01 (0.44–20.55)		3.01 (0.43–21.09)
EMG qualitative report:			
Negative myopathic results		Reference:	Reference:
Borderline myopathic results		1.00 (0.08–11.67)	0.94 (0.07–12.09)
Positive myopathic results		1.28 (0.23–7.13)	1.03 (0.17–6.04)
Cox and Snell R square	0.28	0.26	0.28
Nagelkerke R square	0.38	0.35	0.38
Hosmer Lemeshow Test	0.91	0.94	0.69
AUC (95% CI)	0.81 (0.69–0.92)	0.79 (0.67–0.92)	0.82 (0.70–0.93)

Conclusions: QMUS with NMD algorithm provides a fair diagnostic value for patients suspected for an IIM and is similar to EMG results. A sizeable NPV indicates a low rate of false negative QMUS results. In addition to the relevant PPV for the presence for NMD, QMUS could serve as a potential screening tool for clinicians to detect possible myopathies and to rule out the presence of IIM.

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AB0777

INFLAMMATORY MYOSITIS ASSOCIATED WITH MYASTHENIA GRAVIS WITH AND WITHOUT THYMIC PATHOLOGY: CASE SERIES AND LITERATURE REVIEW

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Background: Inflammatory myopathies (IM) and Myasthenia gravis (MG) are two well-recognised and distinctive neuromuscular diseases. The association of myasthenia gravis (MG) and inflammatory myositis (IM) is rare and often only one of the diseases is diagnosed. The coexistence of MG and IM might be associated with thymoma. Even less common is the association of IM (polymyositis or dermatomyositis) and myasthenia gravis in the absence of thymoma.

Objectives: Here, we report a case series of 6 patients with concurrent MG and IM who were followed at the Neuromuscular Disease Unit (NMDU) at a tertiary referral centre in Vancouver, British Columbia. We also conducted literature review on clinical characteristics, diagnostic challenge and management of this condition.

Methods: In this study, we retrospectively examined patients seen at NMDU from 2004 to 2017 who had diagnosis of concurrent MG and IM. We reviewed medical records to assess their clinical presentations, laboratory findings, imaging studies and electrophysiological features. The data is presented descriptively.

Results: We identified 6 patients with MG-IM overlap. Three patients had simultaneous onset of MG and IM, 2 of whom presented with myasthenia crisis and fulminant myositis. In the other 3 patients, MG was the initial presentation and IM occurred 3–11 years after MG.

Diagnosis of MG was confirmed with clinical features, electromyography and/or serology. All had symptoms of MG with predominant ocular or bulbar weakness. Among these 6 patients, 3 had underlying thymic pathology including two benign thymoma and one stage IV thymoma; all 3 patients had Acetylcholine Receptor (AChR) Antibody. Of the 3 patients with no thymic pathology by computed tomography (CT) or thymectomy, 1 had high positive AChR antibody and 2 were negative.

Four patients had biopsies confirming the diagnosis of dermatomyositis or polymyositis. The other 2 patients declined biopsy; however, their MRI and EMG findings were consistent with IM. Only one patient had typical dermatomyositis rash. Among the 3 patients with underlying thymic pathology, thymoma were resected; all 3 were treated with high dose glucocorticoid, IVIG, and methotrexate with complete remission after 2 years. Of the 3 patients with no thymic pathology identified, one patient (AChR+) was in remission on mycophenolate and passed away from pancreatic cancer; two patients (AChR-) had refractory MG and IM, and both responded to rituximab.

Conclusions: In summary, this is one of the largest case series with MG-IM overlap with or without thymic pathology. It is very important to recognise such association and the different pattern of muscle involvement because therapies may be adjusted to treat both conditions. In patients with thymic pathology, conventional disease modifying agents, IVIG and glucocorticoid in addition to thymoma resection appear to be effective. In patients with refractory MG and myositis who were AChR negative, rituximab may be effective.

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AB0778

CLINICAL CHARACTERISTICS AND PROGNOSIS OF POLYMYOSITIS AND DERMATOMYOSITIS ASSOCIATED WITH MALIGNANCY: A 25-YEAR RETROSPECTIVE STUDY

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Background: Previous studies indicate that malignancies in polymyositis (PM) or dermatomyositis (DM) patients are associated with high mortality. Hence, it becomes important clinically to identify patients who are at high risk of developing

malignancy. However, previous studies to characterise such risk factors for malignancy in patients with PM and DM were small size or the findings were not conclusive.

Objectives: This study investigated factors predictive of malignancy and prognosis in patients with PM and DM.

Methods: We conducted a retrospective study of PM and DM patients who were inpatients at our hospital between January 1992 and September 2016. The diagnosis of PM or DM was made according to Bohan and Peter criteria. The collected data included gender, age at onset, laboratory test results at presentation, clinical features and complications in all progress. Past history of hypertension or diabetes mellitus was determined upon diagnosis of myositis. All patients in this study were followed from the baseline visit until loss of follow-up or death or otherwise until censor date 1 September 2017. Fisher exact test and Mann-Whitney U test were used for group comparisons. Univariate and multivariate analysis of the predictors of malignancy associated with PM or DM were performed by logistic regression to identify independent risk factors. Patient survival was analysed by using Kaplan-Meier curve and log-rank test.

Results: Among 134 patients, 29 had cancer diagnosed between 2 years prior and 3 years following identification of PM or DM. Esophageal cancer (n=5, 14.7%) and lung cancer (n=5, 14.7%) were the most frequent. Univariate analysis showed that male sex (55.2% vs 23.8%, p=0.003), older age (64.6±11.3 vs 53.4±16.5, p<0.001), past history of diabetes mellitus (28.6% vs 2.9%, p<0.001), dysphagia (41.4% vs 17.3%, p=0.01) and absence of interstitial lung disease (37.9% vs 64.8%, p=0.01), arthralgia (17.2% vs 47.1%, p=0.005) and the Raynaud phenomenon (3.4% vs 23.1%, p=0.02) were associated with increased malignancy. Multivariate analysis showed that independent factors included male sex (OR=3.65, p=0.03), older age (OR=1.05, p=0.02), past history of diabetes mellitus (OR=10.4, p=0.005) and absence of interstitial lung disease (OR=0.25, p=0.03) (table 1). Survival was significantly lower in patients with malignancy than in patients without malignancy (p<0.001).

Abstract AB0778 – Table 1. Multivariate analysis of PM/DM with and without cancer

Variables	Adjusted OR	95% CI	P value
Male, n(%)	3.65	1.17–11.30	0.03
Onset age, years, mean±SD	1.05	1.01–1.10	0.02
Interstitial lung disease, n(%)	0.25	0.08–0.84	0.03
Diabetes mellitus	10.4	2.00–54.3	0.005
Arthralgia, n(%)	0.55	0.15–2.04	0.37
Dysphagia, n(%)	2.82	0.82–9.71	0.10
Raynaud phenomenon, n(%)	0.25	0.03–2.29	0.22

Abbreviation: OR, odds ratio; CI, confidence intervals.

Conclusions: This study is the first to associate a past history of diabetes mellitus with malignancy; 28.6% of PM or DM patients with malignancy had diabetes mellitus whereas 7.3 percent of cancer patients had diabetes mellitus.

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AB0779

LONG TERM FOLLOW-UP OF A SYSTEMIC SCLEROSIS GROUP TREATED WITH BOSENTAN

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Background: Prospective studies with Bosentan have shown short term efficacy, while it is not clear whether long-term treatment may be effective or whether ulcers may recur once treatment is discontinued.

Objectives: Our objective was to evaluate the long term efficacy and tolerability of bosentan in patients with systemic sclerosis (SSc) who develop digital ulcers (DU).

Methods: In the present prospective, observational, non-controlled study, we followed 26 SSc patients treated with Bosentan from Sept 2014 to Dec 2017 Number of DU, semiquantitative capillaroscopic scoring, VAS (visual analogue scale) for Raynaud, VAS for DU and HAQ were evaluated every 6 month. Results are presented as mean(sd). The difference between efficacy measures at follow-up visits was tested with the Wilcoxon's signed-rank test.

Results: The group included 26 patients, 16 females, 11 diffuse subsets, age was 48.08 (9.8) years, disease duration was 84.35 (76.04) months, number of DU was 4.27 (3.71), most of them had a late scleroderma pattern pattern (16/26).