years (0–11) after the diagnosis of SLE. At the time of the CVT diagnosis: no patients had a past medical history of thrombotic event or miscarriage or foetal loss; 7 patients had a lupus flare (5 lupus nephritis [1 class I, 1 class V, 2 class IV and 1 class III+V], 4 immune thrombocytopenia, 2 autoimmune hemolytic anaemia, 3 cutaneous lupus, 1 serositis and 1 arthritis); 7 patients were treated with corticosteroids, 4 with hydroxychloroquine and 4 with immunosuppressive drugs. Other potential precipitating factors of CVT were: 2 nephrotic syndromes, 2 anaemia and 1 hyperhomocysteinemia. CVT was symptomatic for 9 patients: 8 headaches, 3 epilepsy and 1 sensitive-motor deficit. The diagnosis of CVT was confirmed by magnetic resonance imaging (MRI) for 9 patients and cerebral angiography for 1 patient. The median delay between the onset of clinical symptoms and the diagnosis of thrombosis was 10 days.^{3–37} Nine patients presented a single localisation of CVT (superior longitudinal or lateral or cavernous sinus, or cortical cerebral vein). Only 1 patient had thrombosis of both lateral and sigmoid sinus. Cerebral infarction or haemorrhage was seen for 2 patients. Corticosteroids and immunosuppressant treatment were increased or introduced because of a concomitant lupus flare for 2 patients. All patients were treated with heparin followed by vitamin K antagonists for 7 patients or apixaban for 2 patients. One patient received long-term heparin. After a median survey of 19.5 (1–120) months: anti-coagulant drugs were stopped for 6 patients; recanalisation were complete in 7 of the 7 patients assessed on brain MRI; no patient had a residual neurological damage; only 1 patient had a new vascular event in the form of a brain haemorrhage. In addition to these 10 cases we found 17 cases from a literature review.

Conclusions: CVT is a rare event in SLE in absence of APS. In our cohort SLE was often active at the time of the CVT occurrence. Most of the CVT were limited in extension and severity. The outcome on anticoagulant treatment was favourable without residual neurological damage. The occurrence of CVT in SLE is not an indication by itself to increase or to introduce corticosteroids or immunosuppressive drug.

Disclosure of Interest: None declared

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Scleroderma, myositis and related syndromes

SAT0468 SCLERODERMA -INTERSTITIAL LUNG DISEASE: DOES MAINTENANCE THERAPY MAKE A DIFFERENCE

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Background: Interstitial lung disease (ILD) is the commonest cause of death among Scleroderma (SSc) patients. The evidence regarding efficacy of cyclophosphamide (CYC) in SSc-ILD results mainly from two high-quality RCTs.^{1,2} In the scleroderma lung study 1, after 12 months of therapy with oral CYC, a significant difference in mean forced vital capacity (FVC) was noted favouring CYC group. However, the effect waned off by the end of 24 months of follow up, indicating that maintenance therapy might help in preserving the initial benefits.³

Objectives: To study the long-term effect of maintenance therapy in SSc related interstitial lung disease (SSc-ILD)

Methods: This is a retrospective data analysis of forty-three patients with SSc-ILD divided into two groups, those who received maintenance therapy with either mycophenolate (MMF) or azathioprine (AZA) and the other group without any maintenance therapy following CYC induction. Response to treatment was classified as improved ($\geq 10\%$ increase in FVC from baseline), stable (change in FVC of 0%–10% from baseline) and worsened ($\geq 10\%$ decrease in FVC or fall of FVC to $<40\%$ of predicted). Results were expressed as percentages for categorical variables and mean with standard deviation (SD) for continuous variables. Chi square test was used for comparison of responses between two groups of patients. Following induction patients were divided into groups: those They were assessed at three years.

Results: Baseline characteristics of the study population are shown in table 1. Maintenance therapy was given in 29 (67.4%) patients and among them favourable outcome (improve +stable) was noted in 23 (79.3%) patients and unfavourable outcome was noted in 6 (20.7%) patients. Fourteen (32.6%) patients did not receive maintenance therapy and among them, favourable outcome was noted in 4 (28.6%) patients and unfavourable outcome in 10 (71.4%) patients. This difference in the rates of favourable outcome between both the groups was statistically significant ($p=0.001$). Figure 1 summarises the response rates of the patients of these two groups.

Abstract SAT0468 – Table 1. Baseline characteristics of the patients (n=43)

Characteristic	Maintenance group (n=29)	No maintenance group (n=14)
Females (%)	93	85.7
Mean age in years(SD)	40.9 (± 10.9)	42 (± 10.5)
Type of Scleroderma		
Limited cutaneous (%)	44.8	57.1
Diffuse cutaneous (%)	55.1	42.9
ANA positivity (%)	75.9	78.6
PAH (%)	34.5	28.6
Mean FVC (SD)	57.6 (± 13)	55.6 (± 15.2)

Conclusions: Maintenance therapy up to 3 years (either with AZA or MMF) following IV-CYC induction, resulted in a favourable outcome and helps in preserving the initial beneficial effects achieved with IV CYC.

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SAT0469 “INTERSTITIAL LUNG DISEASE IN IDIOPATHIC INFLAMMATORY MYOPATHIES: ULTRASOUND ASSESSMENT OF PLEURAL IRREGULARITIES AND COMPARISON WITH HIGH RESOLUTION COMPUTED TOMOGRAPHY”

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Background: Interstitial Lung Disease (ILD) is one of the most frequent and significant extra-muscular manifestations in Idiopathic Inflammatory Myopathies (IIM). Although high resolution computed tomography (HRCT) remains the gold standard technique for the evaluation of ILD, during the last years several authors proposed ultrasound (US) quantification of pleural irregularities (PIs) as a possible method to detect lung involvement in IIM patients.

Objectives: The objective of our study was to evaluate the prevalence of PIs and to compare the results obtained to the data provided by HRCT.

Methods: Thirtyseven patients (24 females, 13 males, mean duration of the disease 5.21 $\pm$ 7.6 and median age 62.24 $\pm$ 1.8 years) with a diagnosis of IIM according to Bohan and Peter criteria (17PM, 16DM, 1MCI, 3overlap syndromes) who required HRCT evaluation were enrolled. All patients underwent rheumatological clinical evaluation, including dyspnoea measurement by means MRC scale and disease activity assessment according to IMACS criteria (muscle enzymes, MMT8, VAS, HAQ, MYOACT). Patients were also asked to complete Patient Reported Outcome (PRO) questionnaires as Leicester Cough Questionnaire (LCQ). All the patients underwent a thoracic US evaluation in 53 anterior and posterior thoracic areas by Esaote MyLab Gold ultrasound device with 8–18 MHz

linear probe. We evaluated the aspect of the pleural profile assigning each space a score according to a 3 points scale (regular=0, mild irregularity=1, irregularity=2) and summed the score in each space to obtain the PIs total score. HRCT was assessed by an expert radiologist to obtain a semiquantitative evaluation of parenchymal involvement by Warrick score. In a subgroup of patients, thoracic US was repeated by the same operator after two days from the first evaluation and by a second operator to evaluate the intrareader and interreader agreement of the technique.

Results: The PIs total score obtained with lung US (24.70 ± 13.66) was higher in patients with thoracic crackles at clinical examination ($p=0.008$) and in patients with positivity for antisynthetase autoantibodies, particularly anti-Jo1 ($p<0.001$). A positive correlation was found between PIs total score and MYOACT total score ($r=0.36; p=0.027$), instead a negative one was found with LCC score ($r=-0.56; p=0.009$). Interestingly, a good correlation between PIs total score and the Warrick score ($r=0.65; p<0.001$) was found. From the analysis of ROC curve, we demonstrated that a cut-off of PIs total score <18.5 might be able to identify all patients with HRCT abnormalities (sensitivity 100%). Lung US was repeatable (inter-reader reliability: $\alpha=0.916$) and reproducible (intra-reader reliability: $\alpha=0.945$).

Conclusions: The US PIs total score is strictly correlated to clinical, serological and HRCT parameters and as the lung US is a non-invasive and relatively inexpensive technique, the results of our study suggest a possible role of this method in the screening of lung involvement in IIM patients. Follow up studies in a larger cohort of IIM patients are required.

Disclosure of Interest: None declared

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SAT0470 MYOSITIS, OFTEN SUSPECTED, IS ACTUALLY RARE IN PRIMARY SJÖGREN'S SYNDROME: DATA FROM THE FRENCH COHORT ASSESS

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Background: Myositis prevalence in primary Sjögren's syndrome (pSS) and whether it is associated with peculiar extra-muscular involvement and/or a biological profile is unknown.

Objectives: To refine prevalence and characteristics of patients with pSS and myositis.

Methods: In the national multicenter prospective cohort ASSESS (395 patients with pSS with a 5 year-prospective follow-up), patients with suspected myositis were identified. Their charts were reviewed and the patients were compared with the rest of the cohort.

Results: Myositis was suspected in 38 patients (2 men, 36 women, 58^{29-78} yr old) because of myalgia and/or exercise intolerance ($n=38$) along with high blood CK level ($n=28$). As compared with the rest of the cohort, they had higher patient-reported signs including pain VAS, dryness VAS and patient reported index ESSPRI (6.3 vs 5.3, $p=0.007$). In contrast, proportion of systemic involvements (76.3% vs 70.9%, $p=0.57$) and systemic disease activity measured by the ESSDAI, without taking the muscular domain into account (2 vs 3, $p=0.93$), were not different from patients without suspected myositis. Demographic characteristics and disease duration were also similar.

Electromyographic recording and/or muscle histology performed in these 38 patients objective muscular involvement in only 4 patients (1% of the cohort).

- Three patients were diagnosed with inclusion body myositis (sIBM). Accordingly, age at first sign was 49 years, $49-60$ muscle disease had progressive onset as suggested by the 11 years $1-28$ delay between first muscle sign and diagnosis. CK level was <1000 U/L. Immunomodulatory drugs had no effect on myositis, even though methotrexate, IVIg and rituximab were most frequently given than in patient without myositis (respectively 50% vs 12.4%; 75% vs 1.8% and 75% vs 4.9%; $p<0.0001$).

- One pSS patient with myositis was a 39 years old woman with polymyositis. She developed proximal muscle weakness, CK was 750U/L and muscle biopsy found endomysial infiltrate. Muscle force and CK level normalised with immunomodulatory drugs.

Compared to the rest of the cohort, patient with myositis had longer disease duration at inclusion (15 vs 5 year, $p=0.01$) and tended to be younger at pSS diagnosis (41.5 vs 53 year, $p=0.07$). They had more frequent history of systemic manifestations (100% vs 65.0%, $p=0.011$) but ESSDAI at baseline (2 vs 3, $p=0.56$) and the

biological characteristics were not different from patients without myositis. In contrast with the patients with suspected but not confirmed myositis, patient-reported signs (i.e. fatigue, pain, dryness and ESSPRI score) were similar to patients without myositis.

Conclusions: Myositis is frequently suspected in patients with pSS, especially when patients-reported signs are particularly disabling. Myositis occurs very infrequently (1% of the cohort), in patients with longer disease duration. sIBM was the most predominant subset of myositis in the present study.

Disclosure of Interest: None declared

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SAT0471 ABNORMAL NAILFOLD CAPILLAROSCOPIC PATTERNS ARE COMMON IN UNDERWEIGHT SUBJECTS WITH RAYNAUD'S PHENOMENON

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Background: Despite the extensive research on this subject, the exact pathogenesis of RP still remains incompletely understood. The current view on this rather complex phenomena is that the imbalance between vasoconstriction and vasodilatation is purely functional and that structural vascular changes do not occur in PRP patients. Although RP is a frequently occurring problem in underweight patients, microvascular changes have never been (structurally) investigated.

Objectives: The aim of the current study was to investigate the relationship between microvascular abnormalities and body mass index (BMI) in subjects with Raynaud's phenomenon (RP), without an underlying connective tissue disease.

Methods: Nailfold capillaroscopic patterns (NCP) were retrospectively assessed in 352 RP patients, without an underlying systemic disease (e.g. negative serology tests, and no signs of organ involvement). Patient characteristics were obtained and patients were divided by BMI category: underweight (BMI <18.5 kg/m²), normal weight (BMI 18.5–24.99 kg/m²), and overweight (BMI ≥ 25 kg/m²). Patterns were deemed abnormal if the mean capillary count was <20 per 3 mm, or mean number of dilated capillaries per finger was >3 , or any giant capillaries were observed.

Results: Approximately 70% of the underweight patients showed an abnormal NCP. This was irrespective of age and smoking behaviour. Underweight RP patients had the highest mean count of dilated capillaries (5.14 for BMI <18.5 kg/m²; 4.17 BMI for 18.5–24.99 kg/m²; 2.61 for BMI ≥ 25 kg/m², $p=0.002$) and giant capillaries (1.01 for BMI <18.5 kg/m²; 0.48 BMI 18.5–24.99 kg/m²; 0.23 BMI ≥ 25 kg/m², $p=0.024$). However capillary loss was not found to be significantly different ($p=0.458$). Furthermore, mean number of capillaries ($r=0.260$, $p<0.001$), mean number of dilated capillaries ($r=-0.225$, $p<0.001$) and mean number of giant capillaries ($r=-0.221$, $p<0.001$) were found to be associated with BMI.

Conclusions: Our findings indicate NCP abnormalities are more frequently observed in underweight individuals, indicating that microvascular changes may occur independently of an underlying connective tissue disease. These findings may alert clinicians that (perivascular) adipose tissue may play a crucial role in the occurrence of Raynaud's phenomenon.

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