Abstract FRI0428 – Table 2. Associations between mean number of capillaries at baseline and disease activity (ESSG score 2003) at FU (linear regression)

Predictors	B (95% CI)	p
m_nr/pat rater 2	-0.45 (-0.834,-0.07)	0.02
m_nr/3rd dom rater 2	-0.33(-0.62,-0.03)	0.03
m_nr/4th non-dom rater 2	-0.27 (-0.57,0.02)	0.07

Conclusions: The mean number of capillaries had a good association with the history of DUs and predicted disease activity at 6 months follow-up. The m_nr/pat performed better in our analysis than the m_nr/3rd dom and m_nr/4th non-dom, however these could be used alternatively in clinical practice as they are less time consuming.

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FRI0429 DISTINCT CLINICAL AND IMMUNOLOGICAL PICTURE OF MCTD PATIENTS WITH SKIN INVOLVEMENT

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Background: Mixed connective tissue disease (MCTD) is characterised by the co-existence of systemic lupus erythematosus (SLE), rheumatoid arthritis (RA), systemic sclerosis (SSc) and polymyositis/dermatomyositis (PM/DM) symptoms. The majority of patients have skin symptoms typical of at least one of the diseases making up the clinical picture MCTD. MCTD is characterised by an auto-reactive antibody response to RNP antigen and resultant formation of anti-U1RNP antibodies. Current knowledge on cytokine biology and their documented role in the pathogenesis of SLE, SSc and PM/DM suggests that also in MCTD some of them may affect clinical course, activity and/or degree of organ damage.

Objectives: To compare clinical and immunological characteristics of MCTD patients with/without skin involvement.

To identify clinical and immunological parameters increasing the risk for a specific (SLE- or SS-like) type of skin lesions and protecting against them.

Methods: 79 MCTD patients based Kasukawa' MCTD diagnostic criteria were included. The patients were divided into groups based on the presence of skin lesions typical for a given MCTD component: SLE- SSc- and DM-specific.

Results: Skin lesions were found in the majority of the MCTD patients (81%). The SLE-, SSc- and DM-specific skin symptoms were found in 54%, 61% and 5% of the patients, respectively. The group of patients with skin symptoms typical of DM was too small to distinguish it separately (4/79). The measures of disease activity (mean AI=10.6 vs 5.5; p=0.006) and MCTD-related damage (DI=4.1 vs 2.1; p=0.009) in patients with skin involvement were twice as high as in individuals with the intact skin. Furthermore, patients with skin involvement had higher mean serum concentrations of TNF- α (46.4 vs 2.3 pg/ml; p=0.013), and lower levels of IFN- γ (43.2 vs 120.8 pg/ml; p<0.001) than patients without any skin symptoms.

The following clinical and immunological parameters were shown to be independently associated with specific types of skin involvement in MCTD patients on multivariate logistic regression analysis:

Independent risk factors for:

- SLE-like skin changes: increased ESR (OR=8.9, 4.47 and 2.6, respectively), higher AI scores and swelling of the hands,
- SSc-like skin changes: higher DI scores (OR=1.522), and presence of anti-Ro60 antibodies (OR=15.903)

Independent protective factors for:

- SLE-like MCTD: chronic progressive course of the disease (OR=0.248) and higher serum concentration of IFN- γ (OR=0.988)
- SSc-like MCTD: acute onset of the disease (OR=0.155).

Conclusions: The course of MCTD in patients with skin involvement is more severe with specific panel of cytokine levels (increased TNF- α and decreased IFN- γ serum concentrations) is characteristic, as compared to patients with the intact skin.

In patients with SLE-like skin lesions MCTD is more often multiphasic, its clinical activity and levels of inflammatory markers are higher while serum concentration of IFN- γ is diminished. Patients with SSc-like skin lesions more often have chronic MCTD, associated with more severe organ damage and elevated serum levels of TNF- α .

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FRI0430 EFFECT OF ORAL NUTRITIONAL INTERVENTION ON NUTRITIONAL STATUS IN PATIENTS WITH SYSTEMIC SCLEROSIS

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Background: Patients with systemic sclerosis (SSc) are at risk of malnutrition, which ranges from 18% to 56% of cases.^{1,2} The high impact of nutritional status on clinical outcome has been shown for many diseases. The optimal nutritional treatment can lead to improvement or preservation of the current nutritional status and increases probability of long-term survival.³

Objectives: The aim of the study was to determine whether nutritional support has an impact on improvement of nutritional status in SSc patients.

Methods: The study included 61 patients with SSc and 49 healthy adults. Nutritional status was determined with subjective global assessment (SGA), body mass index (BMI), bioelectrical impedance analysis (BIA) and anthropometric measurements. Nutrition-related laboratory tests were measured. Appetite was assessed by simplified nutritional appetite questionnaire (SNAQ).

Results: Impaired nutritional status was confirmed in 16 patients with SSc (26,2%). Those patients had significantly lower SGA, BMI (p=0,0019), hand grip strength (p=0,0019), appetite (p=0,019) and BIA parameters such as lean tissue mass (p=0,013), intracellular water (p=0,0006), adipose tissue mass (p=0,04). In laboratory tests levels of haemoglobin, albumin and HDL cholesterol were significantly lower, while erythrocyte sedimentation rate (ESR) was higher (p=0,0025). Thirteen patients had dietary intervention (high-energy, high-protein, oral, liquid nutritional supplements) for 12 weeks. SGA (p=0,017) and hand grip strength (p=0,006) improved after nutritional treatment. BMI, appetite, BIA parameters, lipid profile and ESR after 12 weeks remained stable.

Conclusions: Assessment of nutritional status in SSc patients should be performed regularly, because inclusion of oral nutritional intervention may improve SGA and hand grip strength.

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FRI0431 DYSREGULATION OF LYMPHANGIOGENETIC FACTORS IN SYSTEMIC SCLEROSIS PATIENTS WITH PULMONARY ARTERIAL HYPERTENSION

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Background: Pulmonary arterial hypertension (PAH) continues to be a major complication in systemic sclerosis (SSc); indicating an unmet need for rational therapeutic targets. Recently, we found that chemokine CCL21 was upregulated in SSc and associated with PAH. CCL21 appears to be a key regulator of the expression and secretion of vascular endothelial growth factor-C (VEGF-C); a critical growth factor for lymphatic vessels. This is interesting as SSc is marked by lymphatic vessel abnormalities; and high blood levels of Angiopoietin-2, which is a known regulator of VEGF-C. Moreover, it was recently shown that VEGFR3, the cognate receptor for VEGF-C, is down-regulated in pulmonary arterial endothelial cells from human idiopathic PAH subjects with BMPR2 gene defects. Based on these observations, we reasoned that the observed upregulation of CCL21 in SSc-PAH could be linked to VEGF-C and Ang-2 dysregulation.

Objectives: Assess serum concentrations of CCL21, the VEGF family and Ang-2 in right heart catheterization (RHC) verified SSc-PAH patients and compare these to patients with borderline PAH, no PH and to healthy controls.

Methods: Sera from the prospective Oslo University Hospital SSc cohort (n=372) and healthy controls (n=100) were analysed for VEGF-A,C,D, CCL21 and Ang2 using Luminex kits from Millipore. Patients with an incident RHC (n=167) were included in the present study. PAH was defined as precapillary PH (mean

pulmonary arterial pressure, mPAP ≥ 25 mmHg) in the absence of significant interstitial lung disease (ILD), based on high resolution computed tomography and lung function tests. Borderline PAH was defined as mPAP of 20–24 mmHg in the absence of significant ILD. Patients with pulmonary hypertension (PH) due to other causes were excluded. Descriptive statistics and logistic regression analyses were performed and tested by the goodness-of-fit test with area under the curve (AUC).

Results: Mean age at onset was 54 ± 14.1 , at RHC 61 ± 10.9 years. The time from sera to RHC was 0.9 ± 3.1 years. In total, 73.7% (123) patients had limited cutaneous SSc, 46.7%⁴⁸ were positive for anti-centromere antibody and 80.2% (134) were females. Of these, 123 (73.7%) patients were included in the study, 28 patients with PAH, 45 borderline PAH and 50 patients with no PH. Mean VEGF-A, C, D, CCL21 and Ang2 levels are shown in figure 1A. VEGF-A was significantly higher in SSc patients compared to healthy controls ($p=0.001$), no significant differences between PAH and no PH patients were found. VEGF-D was decreased in SSc-PAH but not significantly. CCL21 and Ang2 levels were increased in patients with PAH, whereas in VEGF-C was significantly decreased in patients with PAH (figure 1 B-D). In univariable logistic regression analyses, VEGF-C (OR 0.99, 95% CI 0.997–0.998, $p=0.001$, AUC=0.79), CCL21 (OR 1, 95% CI 1–1.003, $p=0.050$, AUC=0.69) and Ang2 (OR 1, 95% CI 1–1.0001, $p=0.49$, AUC=0.67) were associated with PAH compared to no PH patients. Due to few event numbers, no multivariable analyses were performed.

Abstract FRI0431 – Figure 1. Serum levels of A) all; B) CCL21; C) VEGF-C and D) Ang2

Conclusions: The present study is the first to demonstrate dysregulation of lymphangiogenic factor expression of multiple targets in sera of SSc-PAH patients.

Disclosure of Interest: None declared

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FRI0432 SINGLE-PORT THORACOSCOPIC SYMPATHICOTOMY FOR TREATMENT RESISTANT RAYNAUD'S PHENOMENON. FIRST REPORT OF A NOVEL MINIMALLY-INVASIVE ENDOSCOPIC TECHNIQUE

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Background: Raynaud's phenomenon of the hands is a great burden in daily life and reduces quality of life in patients with or without an underlying connective tissue disease. Although vasodilatory treatment may be effective in some patients, complaints may be resistant to treatment, for which additional treatment options are very limited. In earlier years, thoracic sympathectomy by anterior or axillary thoracotomy has been shown effective, but with a great surgical burden and limited durability. In the 90 s, endoscopic procedures were introduced. However, these techniques needed multiple endoscopic ports and still performed a relatively

mutilating sympathectomy of the sympathetic ganglia. In our centre, the single-port thoracoscopic sympathectomy (SPTS) was developed, which is a new minimally invasive endoscopic technique with a limited surgical burden.

Objectives: The aim of the current pilot study was to evaluate feasibility and efficacy of SPTS in patients with treatment resistant Raynaud's.

Methods: This new technique entails only a single-port endoscopic procedure, during which a sympathectomy (figure 1) of the nerve is performed, thus sparing the ganglia. Hospital stay is limited to one day. The procedure has been developed for treatment of hyperhidrosis and performed in our hospital in over 550 patients without major adverse events with a success rate of 98%. In the current study, we aim to include 10 patients with treatment resistant Raynaud's, defined as unsatisfactory effect or contraindications of oral vasodilatory agents and iv prostaglandin analogues. Sympathectomy was performed on the left hand first and the effects were compared with the contralateral hand after 3 and 12 months. Major exclusion criteria were severe lung involvement or proximal vascular stenosis. The primary end point was Raynaud's Condition Score (RCS) and Quality of life (SF-36). Among others, cooling fingertip plethysmography (PPG) and laser doppler imaging (LDI) were used as secondary end points for objective assessment of hand perfusion.

Results: This is an interim report, and 4 patients have been included so far (age 20,29,32,56 years), male/female 3/1, 3 primary, 1 secondary), without a history of digital ulcers. No adverse events have been observed, apart from some hyperaemia in the first post-operative week. RCS significantly in each patient. Additionally, a clear improvement in hand perfusion following cooling was observed with PPG and LDI, as compared to the contralateral site. All 4 patients were satisfied and agreed to have their contralateral site operated on also. Patients will be followed in the outpatient clinic to assess long term efficacy.

Conclusions: Single-port thoracoscopic sympathectomy is a novel minimally-invasive technique which appears to be safe and feasible in patients with treatment resistant Raynaud's phenomenon. However, this study is ongoing and long-term efficacy needs to be established.

Disclosure of Interest: None declared

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FRI0433 ASSESSING MORTALITY MODELS IN SYSTEMIC SCLEROSIS RELATED INTERSTITIAL LUNG DISEASE

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Background: Interstitial lung disease (ILD) is a major cause of morbidity and mortality in systemic sclerosis (SSc). However, the severity of lung involvement can vary widely, and current evidence-based treatment options have modest benefit. Prognosis assessment is particularly important for initial management decisions at the time SSc-ILD is diagnosed. There are now multiple mortality models available for use in SSc-ILD which utilise patient's baseline parameters

Objectives: The Gender, Age, and Lung Physiology (GAP) model,¹ interstitial lung diseases –GAP (ILD-GAP) model,² and the Smoking history, Age, and Diffusion capacity of the Lung (SADL) model³ were compared using a systemic sclerosis-ILD (SSc-ILD) cohort to evaluate which best determined prognosis.

Methods: The models were applied to a cohort of patients with SSc (meeting the 2013 ACR/EULAR classification criteria) seen at a tertiary care centre within 1 year of ILD diagnosis from 2000–2013. Demographics, clinical characteristics,