increased (increase of FVC of more than 5%). The skin score was reliably decreased from 11.2 ± 9.3 to 6.2 ± 4.9 ($p < 0.0001$), FVC was increased from 76.9 ± 19.9 to $84.7 \pm 20.9\%$ ($p < 0.0001$), also the increase of DLCO from 46.3 ± 18.3 to $47.8 \pm 16.9\%$ from the proper amount was detected. The Valentini Disease Activity index reliably reduced from 2.93 ± 1.7 to 1.38 ± 1.2 points ($p < 0.0001$). Under the RTX therapy, the decrease of glucocorticoids dose was insignificant, but statistically reliable. Thus, prednisolone dose reduction from 11.8 ± 4.4 to 9.22 ± 3.2 mg/day was achieved. Clinically insignificant but reliable decline of eGFR from 100.2 ± 23 to 94.6 ± 22 ml/min/1.73m² was detected.

Conclusion: Thus, RTX has a positive influence on the basic parameters of SSC, mainly on skin damage and lung functionality, reduces the general disease activity. The influence of the RTX on kidney function requires further long-term study.

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AB0652

QTC INTERVAL PROLONGATION IN A SCANDINAVIAN COHORT OF PATIENTS WITH IDIOPATHIC INFLAMMATORY MYOPATHIES AND SYSTEMIC SCLEROSIS: CORRELATIONS WITH CLINICAL VARIABLES

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Background: Idiopathic inflammatory myopathies (IIM) are rare devastating diseases characterized by progressive muscle weakness and muscle fatigue. IIM frequently affects other organs pointing to IIM as a systemic inflammatory disease. Cardiac involvement is associated with poor prognosis. The symptoms are often subclinical and therefore overlooked. QTc prolongation has been detected in patients with systemic sclerosis (SSc). Autoantibodies are important diagnostic tools to confirm IIM and are present in approx. 60% of IIM patients. Autoantibodies are increasingly being recognized as markers for specific organ involvement. A biomarker for cardiac involvement has yet to be elucidated.

Objectives: The aim is to generate new knowledge about cardiac involvement in IIM detected by electrocardiography (ECG) and to evaluate possible associations between autoantibodies and cardiac involvement detected on ECG in a cohort of IIM patients compared with ECG changes in a cohort of SSc patients.

Methods: In a Scandinavian cohort, 263 IIM patients (130 polymyositis patients, 77 dermatomyositis patients, and 56 inclusion body myositis patients) and 102 SSc patients were investigated by ECG and basic cardiovascular and disease specific assessments according to international guidelines. IIM patients were tested for myositis specific autoantibodies (MSAs; anti-Jo-1, anti-PL-7, anti-PL-12, anti-OJ, anti-EJ, anti-SRP, anti-Mi-2, anti-MDA5, anti-TIF1 γ , anti-NPX2, anti-SAE1, anti-HMGCR) and myositis associated autoantibodies (MAAs; anti-PM/Scl75, anti-PM/Scl100, anti-Ro52, anti-Ku, anti-cN1A).

Results: Twenty two IIM patients (8.49%) had abnormal QRS duration versus one SSc patient (1.06%) ($P = 0.012$). SSc patients had significantly longer QTc duration than IIM patients (QTc = 432.7 ms $\pm$ 22.2 and 426.4 ms $\pm$ 23.6, respectively) ($P = 0.03$). Multivariate regression analysis revealed that increased C-reactive protein (CRP) ($P = 0.008$), gender ($P = 0.002$), and hypertension ($P = 0.007$) were associated with QTc duration in IIM patients. Likewise, pulmonary arterial hypertension was associated with QTc duration ($P < 0.001$) in SSc patients.

In analysis of pooled data for IIM patients and SSc patients, factors associated with QTc duration were increased CRP ($P = 0.005$), gender ($P = 0.001$), and hypertension ($P = 0.01$).

Conclusion: Both IIM patients and SSc patients had ECG changes though no particular pattern was shown. The findings support our hypothesis on cardiac involvement in IIM patients. No significant association was found between presences of either myositis specific or myositis associated autoantibodies and ECG changes. This could be due to the

relatively low number of each autoantibody. There is a need to conduct larger prospective studies to identify a possible autoantibody for cardiac involvement in IIM.

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AB0653

SERUM CARDIAC BIOMARKERS BUT NOT SUBCLINICAL CARDIOVASCULAR MAGNETIC RESONANCE ABNORMALITIES IN SYSTEMIC SCLEROSIS ASSOCIATE WITH THE DEVELOPMENT OF CARDIOVASCULAR EVENTS

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Background: Primary cardiac disease in systemic sclerosis (SSc-PCD) is associated with a poor prognosis when clinically evident. Subclinical SSc-PCD is described in up to 2/3 of SSc patients when sensitive methods are employed. The prognostic implication of these findings is not clear.

Objectives: To describe the prevalence of cardiovascular magnetic resonance (CMR) abnormalities, their association with clinical phenotype and cardiac biomarkers, and whether cardiac biomarkers and CMR predict the development of cardiovascular (CV) events.

Methods: Patients fulfilling the ACR/EULAR criteria for SSc, with no CV disease (CVD), diabetes and no more than 1 CV risk factor had 3Tesla CMR, including late gadolinium enhancement (LGE), myocardial perfusion, T1 mapping for T1 native and extracellular volume (ECV) quantification, and cardiac biomarkers measured. CV events were defined as episode of myocarditis, heart failure, rhythm disturbances and/or any echocardiographic abnormalities including systolic dysfunction, diastolic dysfunction > grade 1 or regional wall motion abnormalities. 47 healthy volunteers (HV) with comparable age and gender served as controls.

Results: 83 SSc patients were recruited, median (IQR) age of 54 (49, 54), 84% females and 33% dcSSc, 40% had ILD, 24% a history of digital ulceration (HxDU) and 29% were Scl70+.

CMR showed higher ECV% and T1 native, markers of diffuse fibrosis in SSc compared to HV [mean (SD) 30 (4) vs 25 (3), $p = 0.000$; 1241 (76) vs 1206 (55), $p = 0.003$]. Lower myocardial perfusion reserve (MPR) was also noted in SSc patients [mean (SD) 2 (0.8) vs 3 (1), $p = 0.000$]. None of the HV had LGE-fibrosis, whilst 16 (20%) of the patients had a non-ischemic LGE pattern. Left ventricular (LV) volume and function were similar between HV and SSc patients.

Presence of LGE associated with hsTnI and lower forced vital capacity ($\rho = -0.283$, $p = 0.032$; $\rho = -0.217$, $p = 0.5$). Multivariate analysis revealed an association of T1 native with mRSS and CRP ($B = 0.5$, $p = 0.02$; $B = 0.218$, $p = 0.049$). ECV% associated with NTproBNP and HxDU ($B = 0.032$, $p = 0.034$; $B = 0.310$, $p = 0.005$). 73/83 had ≥ 1 year available clinical and echocardiography follow up data with a mean (SD) duration of 24(20) months. 12/73 had a CV event judged to be SSc-PCD. LV mass associated with CV events ($p = 0.043$) (Table 1), whilst univariate regression analysis showed that both hsTnI ($OR = 1.014$, $p = 0.014$) and NTproBNP ($OR = 1.005$, $p = 0.026$) but none of the CMR variables associated with the development of CV events.

Conclusion: Asymptomatic SSc patients with no overt CVD demonstrate both focal and diffuse fibrosis, which are associated with cardiac biomarkers and markers of SSc disease activity. Whilst hsTnI and NTproBNP associated with the development of CV events, none of the measures of fibrosis did although a higher LV mass was noted in this

group. The prognostic implications of subclinical CMR findings remain unclear and would benefit from longer follow-up.

Abstract AB0653 Table 1. CMR measures in SSc patients with CV events versus SSc patients with no CV events

CMR variable	SSc patients with CV events n=12 Mean (SD)	SSc patients with no CV events, n=61 Mean (SD)	p value
LVEDV/BSA (ml/m ²)	83 (17)	76 (15)	0.158
LVESV/BSA (ml/m ²)	33(12)	30(9)	0.222
LVSF/BSA (ml/m ²)	50(7)	47(7)	0.259
LV mass/BSA (g/m ²)	49(10)	43(11)	0.043*
LVEF%	61(7)	62(5)	0.494
ECV%	31(2)	30 (4)	0.572
T1 native(ms)	1210 (78)	1238 (72)	0.262
LGE, n Scar mass	1 0.80 (1.68)	9 0.41(1.14)	0.678 (X ² test)
MPR	1.8(0.5)	2.2(0.8)	0.359 0.375

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AB0654 NAILFOLD MICROVASCULAR CHANGES IN MIXED CONNECTIVE TISSUE DISEASE: DIFFERENCES WITH SYSTEMIC SCLEROSIS

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Background: Detailed nailfold capillary abnormalities have not been described in mixed connective tissue disease (MCTD) as well as their changes over time, unlike systemic sclerosis (SSc) which microvascular damage has been classified and validated by nailfold videocapillaroscopy (NVC) in specific and progressive scleroderma patterns ("Early", "Active", "Late") (1-5).

Objectives: Aim of this study was to retrospectively compare detailed MCTD nailfold capillary abnormalities with those of SSc patients at first visit, and to monitor MCTD capillary changes over time.

Methods: Ten patients (mean disease duration 6.4±4.2 years, mean age 50±19 years), affected by MCTD (Kasukawa's criteria) were enrolled. Patients with either anti-extractable nuclear antigen positivity different from U1RNP or anti-dsDNA antibodies were excluded. Main capillary parameters (scores of enlarged capillaries, giant capillaries, microhemorrhages, capillary ramifications, as well as absolute number of normal and total capillaries per linear millimetre) were assessed in MCTD patients at their first NVC analysis (T0), and during a three year follow-up. Furthermore, NVC parameters were compared at T0 with 10 random SSc patients matched for disease duration (6.4±4.2 years) and age (51±17 years). Statistical analysis was performed by non parametric tests.

Results: The capillaroscopic scores of enlarged capillaries, giant capillaries and microhemorrhages were found significantly higher in SSc versus MCTD patients at T0 (2.50±0.5 vs 1.90±0.6 p=0.04, 1.63±0.7 vs 0.70±0.7 p<0.02, 1.25±0.7 vs 0.70±0.7 p=0.05, respectively). Moreover, the absolute number of both total capillaries and normal capillaries were found significantly lower in SSc versus MCTD patients (5.8±1.9 vs 7.6±1.6 p=0.04 and 0.45±1.0 vs 3.03±2.9 p=0.009). On the contrary, no statistically significant difference was observed at T0 for the other capillary parameters (including capillary ramifications) between the two groups of patients. With the limit of a different pharmacological background among

patients, no statistically significant variation of the scores, as well as of the absolute value of the above reported capillary parameters was observed during the 3 years of follow-up in MCTD patients. No statistically significant correlation was observed between capillary parameters and MCTD clinical aspects (Raynaud phenomenon, dysphagia, dyspnoea, sclerodactyly, sicca syndrome, teleangiectasias and arthralgia) at first visit and during follow-up.

Conclusion: In a limited cohort of MCTD patients with an average disease duration of 6.4 years and a follow-up of three years, the nailfold microvascular damage does not seem to be significantly progressive. Patients with MCTD seem to show less enlarged/giant capillaries, and larger absolute number of total and normal capillaries than SSc patients.

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AB0655 SCLERONET®: A PATIENT-CENTERED MULTIDISCIPLINARY APPROACH TO SYSTEMIC SCLEROSIS

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Background: Systemic Sclerosis (SSc) is a rare connective tissue disease which may affect almost the whole body¹. Because of its clinical complexity and heterogeneity, a multidisciplinary approach is helpful to patients.

Objectives: The project aims at providing evidence that a multidisciplinary approach allows to early detect and rightly treat diseases linked to SSc determining an improvement of the Quality of Life (QoL) for patients.

Methods: GILS, the Italian SSc Patient Organisation, selected 4 expert centers in Milan (where over one thousand patients are treated), among those able to diagnose and cure SSc in its different and difficult aspects. Inside these hospitals excellences have been identified: immuno-rheumatologists (overall management of SSc), cardiologists (cardio-pulmonary involvement and Pulmonary Arterial Hypertension - PAH), maxillofacial and plastic surgeons (lipofilling), odontologists (difficult dental cure due to microstomia), dermatologists (prevention of skin cancer), ophthalmologists (dryness of the eye), vasculars (digital ulcers - DUs), pneumologists (lung fibrosis), gynaecologists (dryness of the vagina) and nutritionists (malabsorption). Specialists can share, care and cure inside this net. A monthly meeting is held with patient representatives (PRs). No private data are disclosed. Hospitals General and Sanitary Managers must give a positive evaluation every year in order to continue ScleroNet® project.

Results: The monthly meeting allows to understand strength and threads and find the way to improve ScleroNet®. The PRs point out the unmet needs², to keep ScleroNet® patient-centered and to focus the attention on the QoL whose improvement has been described by patients but not yet supported for all interventions by questionnaires.

Common inclusion and exclusion criteria have been established. Cardiologists performed 110 assessments through EKG, echocardiography and cardiopulmonary exercise testing. These tests gave the following diagnosis: 16 non-ischemic cardiomyopathy (after BNP and troponin dosage as well as cardiac MRI) got an immunosuppressive therapy; 22 PAHs; 34 diastolic dysfunctions; 23 respiratory disease. The rest did not receive a specific diagnosis but still in follow up.

70 perioral lipofillings have been carried out: body weight rose by 2 to 3 kg after 4 months in patients with low BMI thanks to a greater mouth opening. Moreover, a better elasticity of the perioral area led to better oral hygiene, speech and self-perception.

115 patients before Anti-TNF therapy have been screened to prevent non-melanoma skin cancer; 6 the diagnosis.

10 lipofillings for DUs nonresponsive to pharmacological therapy. UCLA/MUST questionnaires were given to selected patients in order to evaluate the GI involvement and undergo a nutritionist evaluation.

A non-rheumatologist made his first SSc diagnosis due to the personal increased knowledge.

Conclusion: ScleroNet® provides a fruitful practical support to patients in order to receive the best care and cure inside the experts centers in a