patients (6.3%) presented with only positive anti-centromere. CP was normal in 191 patients, while in 45 patients scleroderma pattern was observed and in 19 patients nonspecific pattern suggestive of SAD was seen. Capillaroscopic findings: in each of the 23 patients diagnosed with SSc, 21 patients had scleroderma pattern. Of the 6 patients with SLE, CP was normal in 4 patients, while one patient had abundant tortuosity. Likewise, the 6 patients with Sjögren's syndrome, only one patient had a nonspecific pattern suggestive of SAD. Of the 7 patients with MCTD, four had normal CP, two had a scleroderma pattern and one had tortuous pattern. The 2 patients with overlap (myopathy-scleroderma and Lupus-scleroderma) both had a pathological CP with scleroderma pattern.

Conclusion: Our study demonstrates that the evidence of scleroderma pattern is very specific of SSc. However, in certain diseases such as MCTD, overlap syndromes and inflammatory myopathies, we can also see this pattern. In other SAD, CP is usually normal or may show some nonspecific changes.

References: The contribution of capillaroscopy to the differential diagnosis of connective autoimmune diseases. Best Practice & Research Clinical Rheumatology Vol. 21, No. 6, pp. 1093–1108, 2007

Disclosure of Interests: None declared

DOI: 10.1136/annrheumdis-2019-eular.4133

AB0674

RETROSPECTIVE STUDY OF A COHORT OF PATIENTS WITH SYSTEMIC SCLEROSIS IN A TERTIARY CARE HOSPITAL

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Background: Systemic sclerosis (SS) is an autoimmune disease of unknown etiology, characterized by the presence of fibrosis and vasculopathy in skin and multiple internal organs such as the lungs, the kidneys and the digestive tract. The course of the disease is unpredictable and could remain relatively stable or have a rapid evolution. Multiple studies have been carried out to determine the clinical characteristics and survival prognosis on SS patients.

Objectives: To analyze the demographic characteristics, clinical features, treatment and prognosis in a SS disease cohort.

Methods: We performed an observational and retrospective study of patients with SS. The patients had been attended by the Rheumatology department of a tertiary care hospital. We collected demographic, clinical and analytical variables, as well as treatment and prognosis. We classified the disease employing the LeRoy and Medsger, VEDOSS criteria and 2013 ACR/EULAR criteria.

Results: Of our 43 patients, 36 (83.7%) were female and only 7 (16.3%) were male. The average age was 60.4 years (SD 15.6), the average age at diagnosis was 53.3 years (SD 17.6) and the mean time of evolution of the disease was 7.9 years (SD 6.3). Of all the patients, 3 patients (6.9%) died, the mean age at death being 53.6 years (SD 23.7) and the mean time from diagnosis to death of 19 years (SD 10.1). The most frequently occurring presentation was limited SS (with 18 patients (41.9%)), followed by preescleroderma with 14 patients (32.6%), diffuse SS with 6 patients (14%) and escleroderma sine escleroderma with 2 patients (4.7%). Three patients (7%) were labeled with MCTD. SS was associated with other autoimmune diseases in 20% of patients. Five (11.6%) patients developed neoplasms throughout the course of the study. The rest of the clinical characteristics are listed in Table 1 and 2, as well as the strength of association of these with the type of SS, calculated using chi square and Fischer test.

Conclusion: Worthy of noting in our cohort is the absence of scleroderma in more than 40% of our patients, probably because the new criteria have allowed us to diagnose the disease at an earlier stage and also due to the scarce frequency of puffy fingers with respect to other larger series. Digestive involvement was the most frequent visceral manifestation, followed by pulmonary manifestations, specifically interstitial lung disease (ILD). Despite the small sample size, lung disease was significantly associated with the two forms of systemic sclerosis, but not with the antibody pattern. Both the ILD and the pulmonary arterial hypertension (PAH) were more frequent in patients with SSd. Mortality in all cases was due to interstitial lung involvement. As is frequently described, SS is associated with other systemic autoimmune diseases, constituting an overlap syndrome.

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Disclosure of Interests: None declared

DOI: 10.1136/annrheumdis-2019-eular.7361

AB0675

DECREASED CAPILLARY DENSITY IS ASSOCIATED WITH DECREASED HAND MOBILITY IN SYSTEMIC SCLEROSIS

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Background: Nailfold videocapillaroscopy (NVC) is a simple method to evaluate capillary density and morphology. Systemic sclerosis (SSc) is characterized with decreased hand mobility and joint contractures evolving in hands in early disease stage. Movement ability (MA) of hands can be measured with different methods including Hand Mobility Index in Scleroderma (HAMIS), hand anatomic index (HAI), delta finger-to-palm index (Δ FTP), and number of joints with decreased range of motion (joint contractures-JC).

Objectives: To examine correlation between capillary density and MA of hands in patients with SSc.

Methods: 136 patients with SSc underwent detailed examination of MA and capillary density measured with videocapillaroscopy. 70 patients had limited cutaneous SSc (lcSSc) and 66 had diffuse cutaneous SSc (dcSSc). 48 patients had ≤ 5 years of disease duration (early SSc) and 88 patients had disease duration > 5 years (late SSc) calculated from the first non-Raynaud symptoms.

Results: DcSSc patients had significantly higher HAMIS score than lcSSc patients (4.5/1.5; 6.5/ vs. 2/0.5; 4.5/; $p < 0.01$). HAMIS score showed negative correlation with capillary density in the entire cohort (rho: -0.269, $p < 0.01$) and in dcSSc patients (rho: -0.487; $p < 0.0001$), but not in lcSSc ones.

HAI showed a significantly higher value in lcSSc patients than in dcSSc patients (3.79/3.18; 4.39/ vs. 3.19/2.64; 3.76/; $p < 0.01$). Significant positive correlation between HAI and capillary density was found in dcSSc patients (rho: 0.377, $p < 0.01$).

DcSSc patients showed significantly lower Δ FTP value than lcSSc patients (73.88/65.63; 87.00/ vs. 82.81/73.63; 90.5/; $p < 0.05$). Positive correlation was found between capillary density and Δ FTP in the entire cohort, in dcSSc patients even if dcSSc was examined separately as early dcSSc and late dcSSc. No significant correlation was found between capillary density and Δ FTP in lcSSc patients.

The number of JC was significantly higher in dcSSc patients than in lcSSc patients (16/0; 28/ vs. 0/0; 11/; $p < 0.01$), and no statistically significant difference was found between early SSc patients and late SSc patients. Strong negative correlation was found between JC count and capillary density in the entire cohort (rho -0.376; $p < 0.0001$), in dcSSc patients (rho -0.501; $p < 0.0001$), in early SSc patients (rho -0.583; $p < 0.0001$) and late SSc patients (rho -0.262 $p < 0.05$), but not in lcSSc patients.

Conclusion: Decreased capillary density correlated with decreased hand mobility and higher number of JCs. Tissue ischemia may play role in the development of joint contractures and it might be a contributing factor to decreased hand mobility impairment in SSc.

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Disclosure of Interests: None declared

DOI: 10.1136/annrheumdis-2019-eular.2627

AB0676

BODY COMPOSITION IN MYOSITIS PATIENTS AND THE ASSOCIATION WITH DISEASE SPECIFIC FEATURES, PHYSICAL ACTIVITY AND PLASMA LEVELS OF INFLAMMATORY CYTOKINES

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Background: Skeletal muscle, pulmonary, and articular involvement in idiopathic inflammatory myopathies (IIM) limit the mobility/self-sufficiency of patients, and can have a negative impact on body composition.

Objectives: To assess body composition and physical activity of IIM patients and healthy controls (HC) and the association with selected inflammatory cytokines in IIM.

Methods: 54 patients with IIM (45 females; mean age 57.7; disease duration 5.8 years; PM, (22)/DM, (25)/IMNM, (7)) and 54 age-/sex-matched HC (45 females, mean age 57.7) without rheumatic/tumor diseases were included. PM/DM patients fulfilled Bohan/Peter criteria for PM/DM. We assessed body composition (densitometry: iDXA Lunar, bioelectric impedance: BIA2000-M), physical activity (Human Activity Profile, HAP questionnaire), disease activity (MITAX and MYOACT activity score), muscle involvement (manual muscle test, MMT-8 and functional index, FI2) and plasma levels of 27 cytokines (commercial multiplex ELISA kit, Bio-Rad Laboratories). Data are presented as mean±SD.

Results: Compared to HC, patients with IIM had a trend towards significantly increased body fat% (BF%) as assessed by iDXA (39.9±7.1 vs. 42.4±7.1 %, p=0.077), but significantly decreased lean body mass (LBM) as assessed both by iDXA (45.6±8.1 vs. 40.6±7.2 kg, p=0.001) and BIA (52.6±8.8 vs. 48.7±9.0 kg, p=0.023), and increased extracellular mass/body cell mass (ECM/BCM) ratio (1.06±0.15 vs. 1.44±0.42, p<0.001), reflecting deteriorated nutritional status and worse muscle predispositions for physical activity. Compared to HC, IIM patients had significantly lower bone mineral density (BMD: 1.2±0.1 vs. 1.1±0.1 g/cm², p<0.001). Disease duration negatively correlated with BMD (r=-0.392, p=0.004) and LBM-BIA (r=-0.272, p=0.047). Disease activity assessed by both MITAX and MYOACT positively correlated with LBM-BIA (MITAX: r=0.294, p=0.031; MYOACT: r=0.335, p=0.013) and LBM-DXA (MITAX: r=0.341, p=0.012; MYOACT: r=0.368, p=0.007), similarly as with basal metabolic rate (BMR; MITAX: r=0.336, p=0.014; MYOACT: r=0.351, p=0.010), and fat free mass (FFM; MITAX: r=0.338, p=0.014; MYOACT: r=0.356, p=0.009). CRP was positively associated with BF% assessed both by DEXA (r=0.276, p=0.035) and BIA (r=0.306, p=0.025). Higher BF%-DEXA was associated with worse physical endurance (FI2: r=-0.311, p=0.026) and worse ability to perform physical activity (HAP: r=-0.292, p=0.032). MMT-8 score negatively correlated with ECM/BCM ratio (r=-0.385, p=0.006). Plasma levels of IL-1ra positively correlated with body mass index (BMI, r=0.359, p=0.009), BF% (r=0.363, p=0.009), and visceral fat (VF, r=0.409, p=0.003). Plasma levels of MCP-1 positively correlated with ECM/BCM ratio (r=0.387, p=0.005). Increased plasma levels of IL-10 were associated with decreased LBM (r=-0.473, p=0.014).

Conclusion: Compared to healthy age-/sex-matched individuals we found significant negative changes in body composition of our IIM patients, which are associated with their disease activity and duration, inflammatory status, skeletal muscle involvement, and physical activity. Plasma levels of several inflammatory cytokines were associated with alterations of body composition in IIM patients and highlight the potential role of inflammatory status on impaired body composition during the course of IIM.

Acknowledgement: Supported by AZV NV18-01-00161A, MHCR 023728 and GAUK 312218

Disclosure of Interests: Sabina Oreska: None declared, Maja Špiritović: None declared, Petr Česák: None declared, Ondrej Mareček: None declared, Hana Štorkánová: None declared, Hana Smucrova: None declared, Barbora Heřmáňková: None declared, Kateřina Kubínová: None declared, Martin Klein: None declared, Lucia Vernerová: None declared, Olga Růžicková: None declared, Karel Pavelka: None declared, Ladislav Šenolt Grant/research support from: AbbVie, Consultant for: AbbVie, Bristol-Myers Squibb, Celgene Corporation, Merck Sharp and Dohme, Novartis, Pfizer, Roche, UCB, Amgen, Takeda, Speakers bureau: AbbVie, Amgen, Bristol-Myers Squibb, Celgene Corporation, Eli Lilly, Merck Sharp and Dohme, Novartis, Pfizer, Roche, UCB, Heřman Mann Consultant for: Pfizer, Eli Lilly, Sanofi, Speakers bureau: AbbVie, Roche, Pfizer, MSD, Eli Lilly, Sanofi, Jiří Vencovsky Consultant for: Samsung, Speakers bureau: AbbVie, Novartis, Pfizer, Sanofi, Eli Lilly, Biogen, UCB, MSD, Werfen, Roche, Michal Tomčík: None declared

DOI: 10.1136/annrheumdis-2019-eular.4126

AB0677

TREATMENT OF IDIOPATHIC INFLAMMATORY MYOPATHIES – A SINGLE CENTRE EXPERIENCE

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Background: There is no consensus or recommendations for the treatment of idiopathic inflammatory myopathies (IIM).

Objectives: We explored how we treated incipient IIM patients in our secondary/tertiary rheumatology centre.

Methods: We retrospectively included cases with IIM diagnosed between January 2010 and June 2018, who were followed at least 6 months. The remission inducing treatment and the treatment at the last follow-up were recorded.

Results: During the 102-month period we identified 102 IIM cases (71.6% female, median (IQR) age 62.7 (52.2–72.1) years): 27.5% dermatomyositis, 22.5% anti-synthetase syndrome, 14.7% myositis in an overlap syndrome, 13.7% immune mediated necrotizing myopathy, 11.5% polymyositis, 6.9% cancer associated myositis, 2.9% inclusion body myositis (IBM), 1% unspecified myositis. 94 (92.2%) patients received glucocorticoids (GC), 18/94 (17.6%) as monotherapy. The remaining 81 (79.4%) were treated with additional immunomodulatory agents (Table 1), most commonly methotrexate. In addition to therapy presented in Table 1, 13 (12.7%) also received human immunoglobulins (IVIG), one (1%) received plasma exchange therapy.

Of the 8 (7.8%) cases who were not treated with GC 3 received methotrexate, 1 rituximab, and 4 no additional treatment (1 paraneoplastic polymyositis received chemotherapy, 2 IBM, 1 mild polymyositis in elderly, multimorbid patient).

In the first 6 months following diagnosis, 11 patients died (3 due to infection, 2 as a consequence of IIM, 1 due to haemorrhagic shock, 5 of unknown causes), and 5 were lost to follow-up. The remaining 86 patients were followed for a median (IQR) 37.7 (20.2–62.6) months. At last follow-up 49 (57%) patients were still receiving GC, 8 (9.3%) in monotherapy. 9 (10.5%) cases were off GC and other maintenance treatment. Maintenance treatment at last follow-up is presented in Table 2. Treatment was changed in 13 (15.1%) patients (due to progression of pulmonary involvement in 3, active myositis in 4, pulmonary involvement progression and active myositis in 1 and due to intolerance to medication in 6 cases). In addition to therapy presented in Table 2, 2 (2.3%) patients received IVIG (1 concurrently with rituximab and mycophenolic acid due to recalcitrant anti-synthetase syndrome and 1 due to IBM complicated with dysphagia).

Table 1. Induction treatment of idiopathic inflammatory myopathy

Type of treatment	N (%)
Cyclophosphamide; median dose (range)	23 (22.5); 6 (2–12) g
Methotrexate	43 (42.2)
monotherapy	30 (69.8)
+ cyclosporine A	8 (18.6)
+ cyclosporine A and rituximab	4 (9.3)
+ chloroquine	1 (2.3)
Rituximab monotherapy	4 (3.9)
Azathioprine	3 (2.9)
Mycophenolic acid	3 (2.9)
Cyclosporine A monotherapy	1 (1.0)