

ARA+SSc had a significantly higher ratio compared to ATA+ and ACA+pts (mean 61.24±41.40 [45.78 – 76.70] vs 39.62±17.99 [29.66 – 49.58] and 37.77±10.20 [32.12 – 43.42] $\mu\text{mol}/\text{mmol}$ respectively, $p<0.05$). Kyn/Trp ratio was significantly correlated with SRC ($p<0.05$). We found a direct correlation with mRSS ($r=0.269$, $p<0.05$), peak mRSS ($r=0.276$), urate level ($r=0.376$), CRP ($r=0.285$) and ESR ($r=0.320$). Conversely, Neo levels, although significantly higher in SSc compared to HC (mean 12.63±9.30 [10.21 – 15.06] vs 7.11±3.31 [4.74 – 9.48] nmol/L, $p<0.05$), were not significantly different in diffuse compared to limited SSc, but were higher in ARA+ compared to ACA+ and ATA+ patients (mean 14.93±11.52 [10.54 – 19.31] vs 10.81±4.95 [8.07 – 13.56] vs 10.02±6.83 [6.24 – 13.80] respectively, $p<0.05$). Neo levels significantly correlated with PAH. A direct correlation ($p=0.05$) was found with CRP ($r=0.471$) and ESR ($r=0.430$).

Conclusions: These data suggest that Kyn/Trp ratio and Neo levels may reflect aetio-pathogenetic mechanisms in SSc and be elevated in the subgroup of dcSSc that are ARA+, or those manifesting SSc complications associated with ARA+. A specific IFN-gamma signature could be thought to be responsible for the higher levels found in patients although larger studies are required.

References:

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Disclosure of Interest: None declared

DOI: 10.1136/annrheumdis-2017-eular.3786

SAT0336 MALNUTRITION AND SARCOPENIA IN A LARGE COHORT OF PATIENTS WITH SYSTEMIC SCLEROSIS

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Background: Systemic sclerosis (SSc) is an autoimmune disease that may affect gastrointestinal tract, leading to malabsorption and malnutrition. Previous studies defined this complication with no widely accepted criteria. No thorough evaluations of sarcopenia are available.

Methods: 141 SSc consecutive outpatients have been enrolled. A thorough history, blood samples and body composition by densitometry were collected. Malnutrition was defined accordingly to recently published and widely accepted ESPEN criteria (1); sarcopenia was diagnosed in patients with a reduced skeletal muscle index (2).

Results: The table summarizes cohort's characteristics. Malnutrition was diagnosed in 9.2% (CI95%: 4.4–14.0%). Malnourished patients were more often treated with steroids ($p=0.039$), had worse gastrointestinal symptoms accordingly to UCLA questionnaire ($p=0.007$), lower physical activity accordingly to International physical activity questionnaire ($p=0.028$), longer disease duration ($p=0.019$), worse predicted DLCO/VA and FVC ($p=0.009$, respectively) and worse disease severity accordingly to Medsger severity score (DSS) ($p<0.001$ for total, $p=0.001$ for lung and $p<0.001$ for gastrointestinal tract). In multivariate analysis only FVC ($p=0.006$) and disease severity ($p=0.003$), in particular lung involvement as defined by DSS ($p=0.013$), were confirmed to be worse in malnourished patients. Z-scores were significantly lower in malnourished patients at lumbar site $p=0.033$, even after correcting for possible confounders. Sarcopenia was diagnosed in 20.7% (CI95% 14.0–27.4%); 11/29 sarcopenic patients were also malnourished and 6/29 were cachectic (i.e. sarcopenia + systemic inflammation). Sarcopenic patients had worse DLCO/VA ($p=0.003$) and lung ($p=0.005$) involvement accordingly to DSS than non-sarcopenic ones; cachectic had even lower value ($p=0.016$ for both). Sarcopenic patients had also longer disease duration ($p=0.033$).

Table 1. Patients' characteristics

Age	63 (13)
Sex (female) [§]	119 (84.4)
Diffuse disease subset [§]	44 (31.2)
Disease duration (year)	13.3 (7.2)
Interstitial lung disease [§]	39 (27.7)
Pulmonary arterial hypertension [§]	12 (8.5)
Active disease accordingly to Valentini [§]	27 (19.1)
FVC predicted (%)	103 (23)
DLCO/VA predicted (%)	75 (21)
mRSS*	8 (7)
Medsger severity score*	5 (3)
Erythrocyte sedimentation rate (mm/h)	26 (16)
C-reactive protein (mg/l)*	3 (0)
Endothelin receptor antagonists [§]	16 (11.3)
Prostanoids (any) [§]	130 (92.2)
Steroids [§]	23 (16.3)
Immunosuppressive treatment [§]	35 (24.8)

*Expressed as media (IQR); [§]Expressed as absolute valute (%).

Conclusions: Malnutrition defined with widely accepted diagnostic criteria was found to be lower than previously reported (3–7) using screening tool or non-validated criteria. Sarcopenia was found to be somewhat common, although no previous study on comparable cohorts are available. Lung involvement and

function was shown to be significantly linked with nutritional status and may not be explained only by muscle weakness given the absence of correlation between muscle weakness and FVC but only with DLCO/VA.

References:

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Disclosure of Interest: None declared

DOI: 10.1136/annrheumdis-2017-eular.3420

SAT0337 IMPACT OF ORGAN INVOLVEMENT ON PATIENT-REPORTED OUTCOMES IN PATIENTS WITH IDIOPATHIC INFLAMMATORY MYOPATHIES

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Background: Idiopathic inflammatory myopathies (IIM) are associated with considerable morbidity, primarily related to severe muscle weakness and visceral involvement, resulting in disability and impaired quality of life¹. Results from the OMERACT Myositis Special Interest Group indicate that there is insufficient knowledge on patient-reported outcomes (PROs) in IIM².

Objectives: To analyse the association between organ involvement and PROs in IIM patients, taking the presence of autoantibodies into account.

Methods: Data of IIM patients, recorded in the National Database of the German collaborative arthritis centres between 2007 and 2014, were analysed. Physician-reported data on myositis disease phenotypes, organ involvement and antibody status were linked with PROs on functional status (FFbH, range 0–100, 100 indicating full capability), and numerical rating scales (0–10) for pain, fatigue, general health, physical and emotional well-being and coping. Multivariable linear regression analysis was used to investigate the impact of phenotype, organ involvement and autoantibodies on PROs, adjusted for sex, age and disease duration.

Results: A total of 142 IIM patients - 60 polymyositis (PM), 46 dermatomyositis (DM), 15 antisynthetase syndrome (ASS), 12 overlap (OL), 9 others - with mean disease duration of 7.4 years were included. 85% showed muscular, 36% skin involvement, 22% arthritis, 28% interstitial lung disease, 17% dysphagia and 9% cardiomyopathy. Visceral (lung, cardiac or gastrointestinal) manifestation was present in 46% (PM), 54% (DM), 100% (ASS), and 80% (overlap). While moderate to severe (4–10) fatigue was predominately reported in overlap (64%) and ASS (70%), pain was more frequent in overlap (55%) and emotional discomfort was reported most frequently in ASS (57%). For all PROs, worse outcomes were documented in patients with visceral manifestation. Myositis-specific autoantibodies, predominantly Anti-Jo1, were present in 63% of the patients, and were associated with more frequent visceral manifestation (73% vs. 46%), especially interstitial lung disease (50% vs. 15%), and arthritis (32% vs. 13%), but less skin involvement (26% vs. 49%). DM and PM subtypes showed almost identical coefficients for fatigue, physical well-being, general health and coping, while PM was associated with higher emotional strain. Pulmonary hypertension had a severe impact on pain, functional status and daily activities. Cardiomyopathy was associated with impaired general health, arthritis with poorer scores for coping, physical and emotional well-being.

Conclusions: IIM patients with distinct subtypes differ considerably regarding the frequency of organ involvement and self-reported dimensions of disease burden. Anti-Jo1 positivity is associated with higher visceral organ involvement and arthritic manifestations and may therefore also indicate a higher patient-reported disease burden.

References:

- [1] Marie I. *Curr Rheumatol Rep* 2012;14:275–85.
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Acknowledgements: The database is funded by unconditional grants from the German Collaborative Arthritis Centres and from a consortium of 11 pharmaceutical companies to the German Academy for Continuing Medical Education in Rheumatology.

Disclosure of Interest: None declared

DOI: 10.1136/annrheumdis-2017-eular.3822

SAT0338 SURVIVAL IN A TURKISH INFLAMMATORY MYOSITIS COHORT: A SINGLE-CENTRE STUDY

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Background: Inflammatory myositis is an uncommon group of diseases that