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] μmol/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:
[1] Ozkan Y et al. Correlation between DAS-28 and neopterin in early rheumatoid arthritis. Clin Rheum. 2012.
[2] Krcmova KJ at al. Fast and sensitive HPLC method for the determination of neopterin, kynurenine and tryptophan in amniotic fluid, malignant effusions and wound exudates. Bioanalysis. 2015.

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

C. Caimmi[1], P. Caramaschi[1], A. Venturini[1], E. Bertoldo[1], M. Ferrari[2], M. Rossini[1]. [1]*Rheumatology Unit;* [2]*Internal Medicine, University of Verona, Verona, Italy*

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 value (%).

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:
[1] Cederholm et al. Clinical Nutrition 2015;34;335.
[2] Baumgartner et al. Am J Epidemiol 1998; 147:755.
[3] Baron et al. J Rheumatol 2009;36:2737.
[4] Krause et al. Ann Rheum Dis 2010;69:1951.
[5] Murtaugh et al. Clin Nutr 2013;32:130.
[6] Caporali et al. Clin Nutr 2012;31:666.
[7] Rosato et al. Nutrition 2014;30:204.

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

D. Huscher[1], K. Albrecht[1], J. Richter[2], J. Henes[3], T. Alexander[4], A. Zink[1,4]. [1]*Epidemiology, German Rheumatism Research Centre, Berlin;* [2]*Policlinic & Hiller Research Unit for Rheumatology, Medical Faculty, Heinrich-Heine-University Duesseldorf, University Clinic, Duesseldorf;* [3]*Department of Rheumatology and Clinical Immunology, Eberhard Karls-University of Tuebingen, Tuebingen;* [4]*Department of Rheumatology and Clinical Immunology, Charité University Hospital, Berlin, Germany*

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 life[1]. Results from the OMERACT Myositis Special Interest Group indicate that there is insufficient knowledge on patient-reported outcomes (PROs) in IIM[2].

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.
[2] Regardt M et al. Report from OMERACT 12. J Rheumatol. 2015;42:2492–5.

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

E. Er Gülbezer, F. Yargucu Zihni, G. Karabulut, G. Keser, Y. Kabasakal. *Rheumatology, Ege University Medical Faculty Internal Medicine, Izmir, Turkey*

Background: Inflammatory myositis is an uncommon group of diseases that

can be associated with significant morbidity and mortality related to systemic involvement or treatment-related complications.

Objectives: This study aimed to describe the clinical features and survival of patients with inflammatory myositis in our centre.

Methods: We performed a single-centre, retrospective study on patients with inflammatory myositis (polymyositis [PM], dermatomyositis [DM], or Anti-Jo1 syndrome) seen from 2000 to 2014, noting their demographic data, clinical features and outcome until December 2016. The primary outcome assessed was all-cause mortality. Cumulative mortality rates were estimated using the Kaplan-Meier test; the Log Rank (Mantle-Cox) test was used to compare subgroup differences in survival.

Results: Seventy-four patients (19 PM, 28 DM, 27 Anti-Jo1 syndrome) were available for the study. Median age at diagnosis was 47 years (min 17, max 75) and median follow-up time was 93 months (min 4, max 311). Sixty-one patients (82, 4%) were female and 13 (17, 6%) were male. Malignancy was found in 4 patients (2 invasive ductal adenocancer of breast, 1 over cancer and 1 non-small cell lung cancer) and they were all female DM patients. Nineteen patients (25,7%) died at the end of the follow-up.

The 19 patients with PM consisted of 15 female and 6 male, with a median age at diagnosis of 45 years (min 22, max 74) and median follow-up time of 88 months (min 5, max 204). The 28 patients with DM consisted of 24 female and 4 male, with a median age at diagnosis of 52 years (min 17, max 75) and median follow-up time of 80 months (min 4, max 288). The 27 patients with Anti-Jo1 syndrome consisted of 22 female and 5 male, with a median age at diagnosis of 50 years (min 22, max 63) and median follow-up time of 117 months (min 5, max 311).

Overall survival rates of the whole group were 91%, 83%, and 76% for 1, 5, and 10 years, respectively. The survival rates at 1, 5 and 10 years from the diagnosis were respectively 88%, 82%, %82 for PM, %88, 80%, %80 for DM and 96%, 88%, 74,5% for Anti-Jo1 syndrome. But there was no significant difference between the survival rates of the diagnose groups (p=0,734). Also there wasn't a significant difference between the survival rates of sex and age groups (p=0,503), (p=0,112). But the survival rates were significantly lower in patients with the time from diagnose less than 8 years (p=0,000).
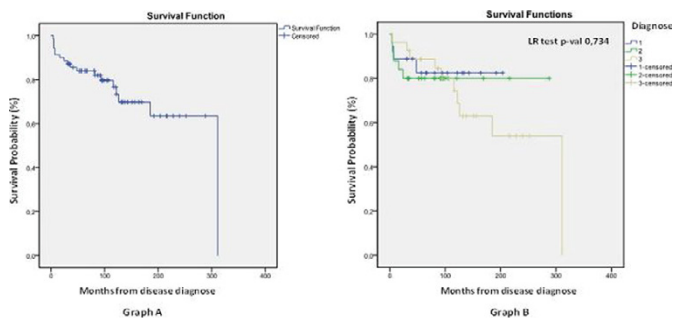

Figure 1. Survival probability of all patients with inflammatory myositis (Graph A) and according to the diagnose* (Graph B). *1, Polymyositis; 2, Dermatomyositis; 3, Anti-Jo1 syndrome

Conclusions: Our study involved 74 patients followed up for a median of 7, 7 years and is one of the largest cohorts of patients with inflammatory myositis in Turkey. Survival was quite similar with the literature. Usually mortality has been expected in the first years after the diagnosis.

Disclosure of Interest: None declared
DOI: 10.1136/annrheumdis-2017-eular.5956

SAT0339 **LUNG TRANSPLANTATION IN PATIENTS WITH INTERSTITIAL LUNG DISEASE ASSOCIATED WITH ANTISYNTHETASE AND ANTI-MDA5 SYNDROMES. EXPERIENCE FROM A REFERENCE SPANISH HOSPITAL**

E. Trallero-Araguás [1], C. Berastegui [2], M. López-Corbeto [1], I. Bello [3], A. Román [2], A. Selva-O'Callaghan [4]. [1]*Rheumatology Unit;* [2]*Pneumology Department;* [3]*Thoracic Surgery Department;* [4]*Internal Medicine Department, Hospital Vall d'Hebron, Barcelona, Spain*

Background: Interstitial lung disease (ILD) is the most characteristic feature and prognosis determinant of patients with antisynthetase (AS) and anti-MDA5 syndromes. Despite immunosuppressive treatment, ILD sometimes progresses to an end-stage lung disease, for which lung transplantation (LT) is the only therapeutic option. There is scarce data about post-LT outcome in this group of patients.

Objectives: To describe clinical characteristics and post-LT outcome of patients with ILD associated to AS and anti-MDA5 syndromes included in the LT program of the Vall d'Hebron Hospital of Barcelona.

Methods: We performed a review of patient records listed in the LT program.

Results: From 1990 to 2016 ten patients (5 women) with ILD related to AS or anti-MDA5 syndromes were included in the LT program (3% of patients with ILD accepted for LT). Nine patients (2 anti-MDA5, 4 anti-Jo1, 2 anti-PL12 and 1 anti-PL7) received LT while 1 patient (anti-Jo1) was still in list at the end of this study. Median age (range) of disease diagnosis was 39 years (25–55). Six patients had

clinical myopathy [2 dermatomyositis (DM) and 4 polymyositis] whereas 1 patient was diagnosed with amyopathic DM. Four patients had associated pulmonary hypertension. Time between disease diagnosis and patient inclusion in LT list was higher in anti-Jo1 patients [median (range) 8.8 years (8.3–17.6)] than in the rest of the cohort [anti-MDA5 <1 year; antiPL12 0.7 and 3.9 years; anti-PL7 1.3 years] (p<0.05). Four patients underwent bilateral LT and 5 unilateral LT. Three patients received an urgent LT (2 anti-MDA5 and 1 anti-Jo1). Six patients presented an histologic pattern of UIP and 3 of NSIP. Early complications (<45 days) included: primary graft disfunction in 3 patients; phrenic paresis in 2 patients and infection in 6 patients. One patient suffered an acute rejection (AR). Six patients presented late complications (>45 days): one patient developed an acute cellular rejection (ACR) and an autoantibody mediated rejection 8 and 10 months from LT respectively; 4 patients suffered chronic lung allograft dysfunction (CLAD), 2 of whom are still alive; Infection was diagnosed in 6 patients. One patient developed a squamous skin carcinoma. No flare of myopathic or lung disease was observed after LT in any case. Four patients (44%) died: 2 of an acute respiratory failure in the immediate postoperatory period (1 caused by suture dehiscence and 1 by refractory ACR); 1 of an invasive aspergillosis, 17 months after LT; and 1 of CLAD after 54 months. Median follow-up (range) of the rest of the cohort was 35 months (8–70). All patients who survived more than 45 days recovered an optimal functional capacity for daily activities with no request for long-term oxygen therapy.

Conclusions: LT should be considered a valid option to treat patients with end-stage or severe and rapidly progressive ILD associated to AS and anti-MDA5 syndromes. An early remission to LT referral centers for evaluation should be considered especially in non-Jo1 patients. No relapse of myositis or ILD was observed after LT. Mortality could not be attributed to the primary disease.

Disclosure of Interest: None declared
DOI: 10.1136/annrheumdis-2017-eular.6320

SAT0340 **SUBCLINICAL CARDIOVASCULAR DISEASE IN SCLERODERMA: A STUDY WITH CARDIOVASCULAR RISK CHARTS, CT CORONARY CALCIUM SCORE AND CAROTID ULTRASONOGRAPHY**

I. Sanz Pérez [1], F. Martínez Valle [1], A. Guillén del Castillo [1], N. Pizzi [2], A. Roque Pérez [3], H. Cuéllar Calàbria [3], A. Fernández Codina [1], E. Callejas Moragas [1], V. Fonollosa Pla [1], C.P. Simeón Aznar [1]. [1]*Internal Medicine;* [2]*Nuclear cardiology;* [3]*Radiology, Hospital Universitari Vall d'Hebron, Barcelona, Spain*

Background: Recently published population-based cohort studies had shown a high prevalence of cardiovascular (CV) disease in Systemic Sclerosis (SSc) patients.

Objectives: The aim of this study is to compare three different methods to measure CV risk in patients with scleroderma.

Methods: We conducted a cross-sectional study in a single center that included 43 SSc patients without CV events. We used both CV risk assessment charts SCORE for populations with low CV risk and REGICOR algorithm adjusted to Spanish subjects. Coronary Compute Tomography (CT) with coronary arterial calcium score (CACscore) was performed considering several cut-off points as predictors of CV risk. Carotid doppler ultrasound was performed to measure the Carotid Intima Media Thickness (CIMT) and for the detection of cholesterol plaques, according to Mannheim consensus criteria.

Results: Risk factors and SSc related features are described in table 1. None of the patients were catalogued as high risk according to SCORE chart (>5%). According to REGICOR chart, 17 patients (39,5%) were catalogued as intermediate risk and none as high risk. Twenty-two patients (51,2%) had carotid plaques (CP) and the CACscore of these patients was 283.4. In patients without CP CACscore was 53,2 (p<0.05).

Based on the presence of CP we performed ROC curve with CACscore. The AUC was 0.778. The best cut off point was 28 with a sensibility of 71% and a specifity of 82%. Kappa's coefficient was 0.54. Twenty patients (46.5%) had CACscore>28. Compared to patients with CACscore<28 statistical significance was found on CIMTmax (0.93 vs 0.80mm; p<0.01), presence of CP (79% vs 25%, p<0.01) and number of CP (2.55 vs 0.43, p<0.01). There was no statistical significance on CIMTm (0.71 vs 0.64mm p: 0.11).

Female	37 (82%)
Age	59 (33–83)
Disease duration	19 (3–57)
Limited cutaneous SSc	32 (74,4%)
Diffuse cutaneous SSc	10 (23,3%)
Sine Scleroderma SSc	1 (2,3%)
Arterial Hypertension	26 (60,5%)
Diabetes mellitus	5 (11,6%)
Statin treatment	22 (51,2%)
Smokers	5 (11.6%)
Digital ulcers	24 (55,8%)
Pulmonary Hypertension*	6 (14%)
Interstitial lung disease	20 (46.5%)
Altered conduction in Electrocardiogram	15 (34,9%)
Left Ventricular diastolic dysfunction**	32 (74,4%)
Left Ventricular systolic dysfunction**	3 (7%)

*Mean pulmonary artery pressure >25 mm Hg at right heart catheterization. **Measured by echocardiography.