changes. This finding supports the notion that this treatment modality normalises specific serum protein imbalances implicated in SSc pathogenesis.

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Disclosure of Interest: None declared

DOI: 10.1136/annrheumdis-2018-eular.3731

SAT0487 A COHORT STUDY OF MACROVASCULAR INVOLVEMENT IN PATIENTS WITH SYSTEMIC SCLEROSIS

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Background: It is well known that systemic sclerosis (SSc) affects microvessels, but data on macrovascular involvement are still lacking or debatable.

Objectives: Aim of this study was to estimate the prevalence of atherosclerotic plaques and their possible determinants in a large cohort of SSc patients.

Methods: One-hundred and four outpatients with SSc were enrolled. Data about disease characteristics and cardiovascular risk factors (diabetes, hypertension, dyslipidemia, smoke) were collected and patients underwent a full ecocolor Doppler ultrasonography of arteries of the neck and lower limbs (LL).

Results: Mean age of our cohort was 62±13 years and 17 (16.3%) patients were male. Seventy patients (67.3%) had a limited subset and the mean disease duration was 12.7±6.8 years. A previous history of digital ulcers was found in 27 cases (26.0%). Thirty-eight patients (36.5%) were on ongoing or previous treatment with immunosuppressive drugs.

Regarding cardiovascular risk factors, 22 (21.2%) had hypertension, 7 (6.7%) diabetes, 52 (50%) dyslipidemia and 45 (43.3%) were active or past smokers. Fifty-seven (54.2%) patients had plaques at carotids, 1 (1%) at LL and 37 (35.9%) at UL. Prevalences were higher in older patients, as expected.

Patients with carotid plaques were older ($p<0.001$), with hypertension ($p=0.057$), a limited disease subset ($p=0.005$), a more severe disease accordingly to Medsger severity score (0.048), worse renal function ($p=0.012$), higher glucose blood levels ($p<0.001$), homocysteinemia ($p=0.006$) and ESR ($p=0.004$) and less often on immunosuppressors ($p=0.048$) but more often on steroids ($p=0.050$). Patients with LL plaques were older ($p<0.001$), male ($p=0.003$), treated with statins ($p=0.056$), with a worse renal function ($p=0.001$), higher glucose blood levels ($p=0.038$) and homocysteinemia ($p=0.006$).

In multivariate analysis with all variables with a $p<0.010$ in univariate, patients with carotid plaques were found to be older ($p=0.003$) and with a limited disease subset ($p=0.012$; OR 5.6, CI95% 1.5 to 21.5). These data were confirmed even after correcting also for other well-known risk factors for atherosclerosis. No variables were found to be statistically different between patients with and without LL plaques.

Conclusions: in this study we performed one of the most complete evaluation of macrovascular involvement in one of the most numerous cohort of SSc patients present in literature. The prevalence of carotid and LL plaques did not seem to be higher in SSc patients as compared to what reported in healthy subjects. Intriguing is that patients with limited disease have an increased risk of having carotid plaques even after correcting for possible confounders.

Disclosure of Interest: None declared

DOI: 10.1136/annrheumdis-2018-eular.3929

SAT0488 ORGAN INVOLVEMENT AND ILD PROGRESSION IN SCLERODERMA PATIENTS WITH ANTI-TOPOISOMERASE I SPECIFICITY AND LIMITED CUTANEOUS FORM

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Background: The great variability of clinical manifestations in systemic sclerosis (SSc) yields the need of defining prevalence and progression of organ

involvement in the different subsets of the disease. Despite the classical association between anti-topoisomerase-I (SCI70) positivity with diffuse cutaneous SSc (dSSc), and anti-centromere (ACA) with limited cutaneous SSc (lSSc), a population of patients with anti-SCI70 and lSSc has been described.¹ Interstitial lung disease (ILD) is nowadays the leading cause of mortality in SSc; its progression can range from a self-limiting form to a rapidly progressive lung involvement, despite immunosuppressive treatment.

Objectives: The aim of our study was to evaluate the prevalence of different organ involvement in three groups of patients: ACA-lSSc, SCI70-dSSc e SCI70-lSSc. Our second endpoint was to investigate possible differences in ILD onset and progression between SCI70-lSSc and SCI70-dSSc patients.

Methods: Consecutive 260 patients attending the Rheumatology Unit of Padova University of Padova were included and retrospectively evaluated. Clinical, biochemical and functional parameters concerning pulmonary, cardiac and articular involvement were collected in all patients, with an average follow-up time of 15 years. As for lung involvement, spirometric indices (FVC e DLCO) and HRCT at ILD onset and at last follow up were considered (median ILD duration 8 years^{4,12}). Time between SSc onset and ILD first evidence was defined as "scleroderma free ILD". ILD grading was determined according to Kazerooni score. ILD progression was defined as either an HRCT score worsening of at least 2 points, or as a significant progression of spirometric indices (10% and 15% for FVC and DLCO respectively).²

Results: 150 patients with ACA-lSSc, 58 with SCI70-dSSc and 52 with SCI70-lSSc were included in the study. SCI70-lSSc patients presented more often with pulmonary and articular involvement with respect to ACA-lSSc patients (50% vs 6.9% and 42.9% vs 23.6% respectively), and less often compared to SCI70-dSSc. In SCI70-lSSc, cardiac and gastrointestinal involvement, BEV and digital ulcers were less prevalent with respect to SCI70-dSSc ($p<0.05$). SCI70-lSSc patient had a longer "scleroderma free-ILD" compared to SCI70-dSSc. At ILD onset pulmonary function was worse in SCI70-dSSc group than in SCI70-lSSc group ($p=0.02$ e $p=0.009$ for FVC e DLCO respectively), even in the absence of a significant difference between HRCT scores (12.73 ± 5.25 vs 10.25 ± 4.93 , $p=0.284$). Total ILD progression was significantly higher in SCI70-dSSc ($p<0.0001$).

Conclusions: Our study shows that SCI70-lSSc patients appeared to have a different prevalence of targeted organ involvement and ILD progression with respect to both SCI70-dSSc and ACA-lSSc. Since the correct classification of SSc patients is extremely important in view of a tailored treatment, our data suggest that it could be worth to identify patients with SCI70 and limited cutaneous form as a different and specific subset.

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Disclosure of Interest: None declared

DOI: 10.1136/annrheumdis-2018-eular.7232

SAT0489 ASSESSMENT OF PERSISTENT ORGAN DAMAGE ACCORDING TO IMACS (INTERNATIONAL MYOSITIS ASSESSMENT AND CLINICAL STUDIES) MYOSITIS DAMAGE INDEX IN 92 PATIENTS WITH IDIOPATHIC INFLAMMATORY MYOSITIS

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Background: Number of work regarding long term organ damage caused by idiopathic inflammatory myopathies (IIM) and risk factors associated with organ damage have been understudied and never reported from Turkey; recently a new tool as developed for that purpose.

Objectives: In this study we aimed to evaluate by long term organ damage and risk factors, associated with these prospectively in IIM patients.

Methods: IIM patients ($n=110$) who has been followed up for at least six month by our clinic and fulfilling Bohan and Peter criteria were recruited. Demographic data, clinical and serological features, treatment and final clinical status was recorded. IMACS Myositis Damage Index (MDI) was determined twice in 92 patients (71% female) at the time of diagnosis from the records and at the last clinical visit prospectively.

Results: Mean age of the 92 patients during the diagnosis was 46±14.7. Out of 92 patients 69% had dermatomyositis, 23% had polymyositis, 8% had necrotizing

autoimmune myopathy and inclusion body myositis. Mean follow up was 82 months. Frequencies of dysphagia, respiratory muscle involvement and interstitial lung disease were 29.5;34% respectively. Twenty-one percent of the patients had associated malignancy. The mean daily prednisolone dosage and total amount was 7.5 mg/day and 9000 mgs. Mortality was 13%. Initial mean MDI at the time of diagnosis was 1.6±3.0 (range, 0–14) and the last DMI score recorded was 6.1±4.7 (range, 0–21). After the last assessment the proportion of patients without damage was 8% and whose score was 4 were 37%. The last DMI score was significantly higher than the first DMI score ($p<0.001$). The last DMI in females and patients with calcinosis were significantly high ($p=0.002$; $p=0.007$). The last DMI score and disease duration were weakly correlated ($r=0.35$ $p=0.001$). A moderately significant correlation was found between the last DMI score, the duration of glucocorticoid use and the total dose used ($r=0.48$ $p<0.001$; $r=0.45$ $p<0.001$).

Conclusions: Our long term follow up data showed that persistent organ damage assessed by DMI and mortality were high in patients with IIM and over half of patients developed severe damage. Organ damage was detected in some patients at presentation and DMI scores were significantly increased during the follow up. DMI scores were found high in females and in patients with calcinosis. There were significant correlation between disease duration, the duration of glucocorticoid use, the total dose used and DMI scores. Current treatments and strategies have been insufficient at improving the prognosis of patients with IIM and new treatment strategies and drugs are needed.

Disclosure of Interest: None declared

DOI: 10.1136/annrheumdis-2018-eular.4872

SAT0490 INCIDENCE AND PREVALENCE OF SYSTEMIC SCLEROSIS IN AN ITALIAN ALPINE VALLEY DURING A 18-YEAR LONG PERIOD

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Background: Not much information is available on the variations in the incidence and prevalence of Systemic Sclerosis (SSc) over time.

Objectives: To investigate the epidemiology of SSc in Valcamonica, an Alpine valley in northern Italy (Brescia Province, Lombardy region), during a 18-year-long period.

Methods: All patients with diagnosis of SSc living in Valcamonica between 1999 and 2016 were identified by capture/recapture method using: 1) clinical databases of the only secondary Rheumatology Unit present in the valley (Esine Hospital), and of the only tertiary referral centre for this area (Spedali Civili, Brescia); 2) administrative data (ASST Valcamonica, Esine), extracting all records with the ICD-10 code for SSc (710.1). Clinical charts were reviewed and patients included in the analysis when either the 1980 ARA or the 2013 ACR/EULAR classification criteria for SSc were satisfied. Incidence was calculated using the number of new cases observed as the numerator, and the Valcamonica population as the denominator for each year; to study temporal changes, mean yearly incidence during 3 different 6 year intervals was calculated (table 1). Prevalence rates were estimated at 4 different time points dividing the number of living patients by the number of individuals in the population (table 2). Survival, incidence and prevalence rates were expressed with 95% confidence intervals. Incidence and prevalence rates observed in different periods were compared using ANOVA test. Demographic data were expressed as the medians (IQR).

Results: General population with age over 14 years living in Valcamonica varied during the evaluated period between 85 168 (1999) and 91 245 inhabitants (2011). Fifty-six patients with SSc fulfilling the 2013 ACR/EULAR criteria, and in 68% of them also the 1980 ARA criteria were identified (Female 85.7%, Male:

14.3%; Caucasian: 96.4%, African 3.6%; age at diagnosis: 58;^{47–80} deaths: 13 (8 because of SSc); survival: at 5 years: 89.4% (76.2–95.5), at 10 years: 84.4% (69.9–92.3); diffuse SSc: 17.9%, limited SSc: 82.1%; anticentromere: 62.3%, anti-topoisomerase I (topo I): 20.8%, anti-RNA polymerase III (RNAP3): 5.4%; anti-topo I and RNAP3: 1.9%; Anti-Th/To: 1.9%; interstitial lung disease: 23.1%; group 1 pulmonary arterial hypertension 19.6%; renal crisis 5.4%). No significant variation of incidence was observed during the period of time evaluated in the study (table 1). Analysis of prevalence revealed a continuous increase of the disease using both the 1980 ARA criteria or 2013 ACR/EULAR criteria ($p<0.001$ for both comparisons; table 2). The prevalence on 2016 was 50.9 per 1 00 000 persons aged >14 years, one of the highest ever recorded by SSc epidemiology studies. However, notably, the prevalence in Valcamonica during the first years of the 21 st century was similar to that reported by previous other studies performed in Italy in the same years.^{1–2}

Abstract SAT0490 – Table 1. Incidence of SSc in Valcamonica expressed as cases per 100,000 adults aged over 14-years

SEX	STUDY PERIOD	MEAN POPULATION >14 YEARS	NEW CASES	INCIDENCE (CI 95%)
MALE AND FEMALE	1999-2004	86,205.5	12	2.3 (1.2-4.1)
	2005-2010	89,448.5	14	2.6 (1.4-4.4)
	2011-2016	90,843	16	2.9 (1.7-4.8)

Abstract SAT0490 – Table 2. Estimates of the prevalence of SSc in Valcamonica expressed as cases per 100,000 adults aged over 14-years

SEX	YEAR	POPULATION >14 YEARS	LIVING CASES	PREVALENCE (CI 95%)
MALE AND FEMALE	1999	85168	14	16.4 (9-27.6)
	2005	88011	26	29.5 (19.3-43.3)
	2011	91245	37	40.6 (28.6-55.9)
	2016	90441	46	50.9 (37.2-67.8)
FEMALE	1999	43755	13	29.7 (15.8-50.8)
	2005	44751	22	49.2 (30.8-74.4)
	2011	46149	31	67.2 (45.7-95.3)
	2016	45868	42	91.6 (66-123.8)
MALE	1999	41413	1	2.4 (0-13.5)
	2005	43260	4	9.3 (2.5-23.7)
	2011	45096	6	13.3 (4.9-29)
	2016	44573	4	9 (2.5-23)

Conclusions: Although the incidence of SSc did not change significantly over time in this area, the disease prevalence constantly increased, with recruitment of new cases exceeding the number of deaths.

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Disclosure of Interest: None declared

DOI: 10.1136/annrheumdis-2018-eular.4239