an inflammatory disease (overlap, infection, neoplasia), and showed a tendency to an association with mortality. This association became statistically significant when considering DD plasma levels as a quantitative variable ($p < 0.001$) and remained significant after adjustments (age, coexistence of an inflammatory disease) ($p = 0.003$).

Conclusions: DD plasma levels are associated with macrovascular involvement and can be helpful in predicting medium-term mortality in our SSC patients. Levels of plasma DD can be modified by systemic inflammation.

Disclosure of Interest: None declared

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SAT0496 INCIDENCE, CARDIOVASCULAR EVENTS AND MORTALITY OF ADULT INFLAMMATORY MYOPATHIES IN SOUTH KOREA: A NATIONWIDE POPULATION-BASED STUDY

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Background: The inflammatory myopathies (IMs), including dermatomyositis (DM) and polymyositis (PM), are rare autoimmune diseases characterised by progressive proximal muscle weakness, elevated muscle enzyme and various organ involvement. Studies on epidemiology and mortality of IMs at the national level are rare.

Objectives: We conducted a nationwide population study of incidence, cardiovascular events (especially acute myocardial infarction, ischaemic stroke, haemorrhagic stroke) and survival of IMs in South Korea over the course of 11 years.

Methods: We used data from the Rare Intractable Disease (RID) registry and Health Insurance Review and Assessment (HIRA) service, which include information on all IMs patients diagnosed based on uniform criteria between 2005 and 2015. Survival data from Statistics Korea linked to HIRA-RID database were used in our survival analysis.

Results: In this study, total incident cases between 2005 and 2015 were 3014 (1,860 DM patients, 1,154 PM patients) and the mean annual incidence rate was $7.16/10^6/\text{year}$ (DM was $4.42/10^6/\text{year}$, PM was $2.74/10^6/\text{year}$). The female to male ratio of DM was 2.2:1 and that of PM was 1.9:1. Cardiovascular events occurred in 155 patients during the study period. Of the patients with DM, 25 were diagnosed with AMI (SIR 1.3, 95% CI 0.9–1.9), 39 with ischaemic stroke (SIR 1.4, 95% CI 1.0–1.9), 18 with haemorrhagic stroke (SIR 1.3, 95% CI 0.8–2.0). The results of PM were similar with those of DM. Of 155 patients, 63 (40.6%) died and had a high mortality rate. 640 of 3014 patients with IMs died during the study period; 290 and 350 were male and female, respectively. The survival curve for IMs is shown in Fig.1. The DM patients showed a 5 year survival rate (YSR) of 76.8%, and a 10-YSR of 68.9%, and the PM patients showed a 5-YSR of 79.3% and a 10-YSR of 68.3%.

Abstract SAT0496 – Figure 1. Survival curves of dermatomyositis and polymyositis

Conclusions: This is the first study of incidence and mortality of IMs over 3000 cases by the national registry in South Korea. In this nationwide population-based study of IMs, we found still high mortality and low survival and increased cardiovascular events related mortality compared than general population.

Disclosure of Interest: None declared

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SAT0497 FOLLOW-UP OF DISEASE ACTIVITY IN HUNGARIAN PATIENTS WITH IDIOPATHIC INFLAMMATORY MYOPATHY

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Background: Assessing disease activity in patients with myositis is an important goal. A method called Disease Activity Core Set Measures was established by International Myositis Assessment and Clinical Studies Group (IMACS) for evaluating activity of myositis. This assesses the manifestations of myositis which are thought to be reversible that result directly from the inflammatory process.

Objectives: In this prospective study authors aimed to monitoring disease activity in Hungarian myositis patients treated at the Autoimmune Outpatient Clinic, Department of Clinical Immunology, University of Debrecen. First, disease activity data of poly (PM) – and dermatomyositis (DM) patients were compared. Secondly, by patients with active disease, the correlations between the components of Disease Activity Core Set Measures were studied.

Methods: During the three-year follow-up period, at every single visit, authors evaluated disease activity using factors of Disease Activity Core Set Measures and patients filled Health Assessment Questionnaire (HAQ). Only those patients who appeared at least three times during the follow-up period were selected. Statistical analysis was made using SPSS 17.0 statistical software.

Results: Collection of data from 219 patients (age: 44.93 years, female: male ratio 2.98:1) happened in a total of 1101 outpatient visits. The 107 PM patients had statistically significant higher physician and patient visual analogue (VAS) and myositis disease activity assessment tools scores than the 61 DM patients, with the exception of cardiovascular disease activity. Activity of skin lesions was significant higher in DM patients. By the 44 patients, who had an active disease, the MMT scores gave significant negative correlations with HAQ scores ($R = -0.536$ and $p < 0.001$) and CK levels ($R = -0.387$ and $p < 0.001$). The physician and patient VAS gave strong negative correlations with MMT scores ($R = -0.714$ and $p < 0.001$ and $R = -0.730$ and $p < 0.001$, respectively) and positive correlations with HAQ scores and CK levels (physician VAS vs. HAQ $R = 0.691$ and $p < 0.001$; patient VAS vs. HAQ $R = 0.629$ and $p < 0.001$; physician VAS vs. CK $R = 0.622$ and $p < 0.001$; patient VAS vs. CK $R = 0.615$ and $p < 0.001$).

Conclusions: As far as we know, this is the first study which compares the activities of PM and DM patients based on the IMACS system. According to our data, we can conclude that PM patients had more severe muscle symptoms and extra-muscular manifestations. Secondly, as seen in other previous studies, calculations in patients with active myositis showed a clear correlation between the most important components of Disease Activity Core Set Measures.

Disclosure of Interest: None declared

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SAT0498 ABNORMAL ELECTROCARDIOGRAPHIC FINDINGS IN A SCANDINAVIAN COHORT OF PATIENTS WITH IDIOPATHIC INFLAMMATORY MYOPATHIES

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Background: Idiopathic inflammatory myopathies (IIMs) are characterised by progressive muscle weakness and muscle fatigue. IIMs frequently appear with other organ involvement, e.g. the heart. Cardiac affection is often subclinical but associated with poor prognosis which makes early detection critical.

Presence of autoantibodies is common in IIMs and autoantibodies are important biomarkers to confirm the diagnosis of IIMs. Autoantibodies may even predict organ involvement, e.g. the common myositis specific autoantibody anti-Jo1, which is a marker of lung involvement. So far, an autoantibody predicting cardiac involvement has yet to be identified.

Objectives: The aim of this study was to identify and estimate presence of cardiac involvement detected by electrocardiography (ECG) and to evaluate possible associations between ECG changes and autoantibodies in a Scandinavian cohort of patients with IIMs.

Methods: In a Scandinavian cross-sectional study, 241 patients with polymyositis (PM), dermatomyositis (DM), or inclusion body myositis (IBM) and 48 healthy controls (HCs) were investigated by ECG, basic cardiovascular assessments, and

autoantibody profile including myositis specific autoantibodies (MSAs) and myositis associated autoantibodies (MAAs).

Results: Compared to HCs, patients with IIMs more frequently had prolongation of QTc ($p=0.037$) and QRS ($p=0.031$). All patient groups had significantly longer QTc and QRS duration than HCs. In multivariate regression analysis of patients with IIMs, increased CRP ($p=0.006$) was associated to increased QTc. Pooled data for patients with IIMs and HCs showed an association between diagnosis of IIM and increased QTc duration ($p<0.001$). An association between presence of any MAA and QRS duration appeared in patients with IIMs ($p=0.019$). In pooled data for patients with IIMs and HCs, diagnosis of IIM was associated with QRS duration ($p=0.001$).

Table: Multivariate regression analyses of explanatory factors for QTc duration (ms)

Risk factor	OR	95% CI	P-value	OR	95% CI	P-value
Gender, male	0.46	0.28 0.75	0.002	0.46	0.29 0.72	0.001
Age at inclusion, per year	1.02	0.99 1.04	0.068	1.02	1.00 1.04	0.047
Hypertension, no vs. yes	1.63	0.93 2.86	0.086	1.58	0.97 2.60	0.069
Former/present smoker, no vs. yes	1.35	0.84 2.17	0.213	1.57	1.02 2.41	0.041
Increased CRP, no vs. yes	2.17	1.25 3.75	0.006	1.98	1.15 3.40	0.013
BMI, kg/m ²			NS			NS
Dyspnea (NYHAI-IV), no vs. yes			NS			NS
Presence of MSAs/MAAs, no vs. yes			NS	NA	NA	NA
HAQ (Health Assessment Questionnaire), 0–3			NS	NA	NA	NA
IIM, no vs. yes	NA	NA	NA	4.11	2.21 7.63	<0.001

Conclusions: Patients with IIMs, no matter of clinical subgroup, had a higher occurrence of cardiac abnormalities detected by ECG than HCs. Increased CRP and presence of any MAA were associated with increased QTc and QRS duration, respectively. These results support our notion of possible associations between inflammation and autoimmunity and cardiac affection in patients with IIMs. There is now a pressing need to set up a larger prospective study to validate the present findings.

Disclosure of Interest: None declared

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SAT0499 ASSOCIATION BETWEEN SYSTEMIC SCLEROSIS AND OTHER SYSTEMIC AUTO-IMMUNE DISEASES: STUDY IN TWO UNIVERSITY HOSPITALS COHORTS

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Background: Association between systemic sclerosis (SSc) and another autoimmune systemic disease (AISD) in the same patient seems to be more frequent than each disease's prevalence would explain.

Objectives: The aim of our work was to describe patients presenting an overlap syndrome from 2 french cohorts and to compare their characteristics with patients presenting SSc alone.

Methods: This was a retrospective observational study conducted in two french auto-immune diseases reference centre (Strasbourg and Bordeaux). Patients responding to the 2013 ACR-EULAR scleroderma classification criteria for SSc were screened for concomitant AISD. Patients satisfying 2010 ACR-EULAR diagnostic criteria for rheumatoid arthritis (RA) and/or 2016 ACR-EULAR classification criteria for Gougerot-Sjögren syndrome (GSS) and/or 2012 SLICC systemic lupus erythematosus (SLE) classification criteria were included in our study. Patient, disease, and treatment characteristics were retrospectively retrieved from medical records and were compared to a SSc control cohort for Bordeaux University Hospital.

Results: A population of 534 SSc patients was studied. Thirty-four (6.4%) patients were identified as having overlap syndrome. There was 21 (62%, prevalence 3.9%) patients with RA, 14 (41%, prevalence 2.6%) with GSS and 4 (12%, prevalence 0.7%) with SLE (5 patients had 2 AISD).

There were 24 (71%) patients with limited cutaneous SSc. Median Rodnan was 6 (extreme 0–42), 13 (38%) patients had interstitial pneumonia and 9 (26%) presented lung fibrosis. Three patients had pulmonary arterial hypertension (PAH) confirmed with catheterism. Seventeen patients (50%) had anti-centromere Ab, 11 (32%) had anti-Scl70 Ab whereas none had anti-RNA-polymerase III Ab (investigated in 24 patients).

Concerning RA patients, 17 (81%) were ACPA-positive and 17 (81%) had erosive disease. Only 6 (29%) were in remission according to Boolean criteria and 13 (62%) had a DAS28-CRP $\leq$ 3.2 suggesting difficult-to-treat RA. Patients with GSS

all presented sicca syndrome, 8 (57%) had a Chisholm grade $\geq$ 3 on accessory salivary gland biopsy and 12 (86%) had positive anti-SSA Ab. In patients with SLE, 3 (75%) had positive anti-DNA Ab and one had a grade IV kidney disease. Three (75%) patients with SLE had a SLICC organ-damage score $\geq$ 5 suggesting severe SLE.

Compared with our control cohort, patients with overlap syndrome had higher frequency of corticosteroid, immunosuppressive and biologic therapy use (85.3% vs 45%; 70.6% vs 31.3%; 52.9 vs 3.8%; $p<0.0001$ for all comparisons).

Conclusions: Association of SSc and another auto-immune systemic disease is present in more than 6% of patients. These patients might have a more severe disease than usual SSc patients requiring prompt diagnosis and adequate treatment.

Disclosure of Interest: None declared

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SAT0500 MYOCARDIAL INVOLVEMENT AT MAGNETIC RESONANCE IN PATIENTS WITH SYSTEMIC SCLEROSIS AND MINOR ARRHYTHMIAS: ASSOCIATION WITH CLINICAL FEATURES AND IMPACT OF TREATMENT

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Background: Systemic sclerosis (SSc) is an autoimmune fibrotic disease characterised by variable clinical manifestations based on the predominant organ involvement. With the exceptions of acute fulminant myocarditis with pericardial effusion and heart failure, myocardial involvement in SSc is clinically silent until arrhythmias appear, while representing the major cause of sudden death or atrio-ventricular (AV) blocks in SSc. There are no reliable screening algorithm to discriminate myocardial involvement nor established treatments.

Objectives: To identify early/clinically silent myocardial involvement in SSc and define a possible treatment.

Methods: We used Holter electrocardiography (ECG) to investigate our cohort of patients with SSc ($n=221$, all fulfilling the ACR classification criteria) for ventricular ectopic beats (VEB)/AV blocks/unexplained tachycardia as sensitive signs of myocardial involvement. In 24 patients (women 23%–95.8%, anti-centromere-ACA 16%–66.6%, anti-topoisomerase- α Scl70 3%–12.5%, anti-RNA polymerase III 3%–12.5%, anti-Ku 2%–8.3%, limited skin disease-ISSc 12%–50%, diffuse skin disease 5%–20.8%, *sine scleroderma* 7%–29.2%, median age 66.5 years, IQR 57.8–72.5), never treated with anti-fibrotic agents, we performed heart magnetic resonance (hMR) searching for myocardial oedema (on T2 STIR sequences, using the signal ratio between myocardium and skeletal muscle with a cut-off value of ≥ 1.9 for oedema) or fibrosis (presence of any late gadolinium enhancement (LGE) with intramyocardial or subepicardial pattern). Patients with myocardial oedema at hMR were treated with mycophenolate mofetil (MMF) 2 g/day or with azathioprine (AZA) 100 mg/day and underwent a control hMR after six months.

Results: In 10/24 (42%) of the SSc cases with Holter ECG alterations (8 VEB, 1 tachycardia, 1 type II AV block) we observed SSc myocardial involvement at hMR. In more detail, 6 patients had myocardial oedema at T2 STIR sequences (including cases as follows: 1 α Scl70-positive with ISSc and lung fibrosis, 2 ACA-positive with ISSc, 3 ACA-positive *sine scleroderma*), and 4 only fibrosis (1 Scl70-positive with ISSc and lung fibrosis, 1 ACA-positive with ISSc, 1 ACA-positive *sine scleroderma*, 1 anti-Ku-positive *sine scleroderma*). At 6 months of medical treatment, myocardial oedema disappeared in 3 patients treated with AZA and in 1 treated with MMF, the additional 2 patients are still receiving MMF treatment at the time of analysis.

Conclusions: We observed that 42% of patients with SSc and minor arrhythmias on 24 hour Holter ECG have clinically silent myocardial involvement at hMR; those with potential reversible disease (oedema rather than fibrosis) favourably respond to immunosuppressants. Detectable serum ACA and the absence of skin involvement are over-represented in this subgroup of patients.

Disclosure of Interest: None declared

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SAT0501 EARLY VERSUS LATE-ONSET SYSTEMIC SCLEROSIS: ARE THERE CLINICAL AND IMMUNOLOGICAL DIFFERENCES?

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Background: The clinical course of Systemic Sclerosis (SSc) depends on subtype, organ involvement and age. Peak age at onset of SSc is between 30 and 50