PBC or other autoimmune liver disease or manifested elevated liver function tests.

Conclusion: The expression of the PDC antigenic components is variable both in SSc patients with AMA positive PBC and in SSc patients AMA positive without PBC or altered liver function tests, but we could not identify a clinical significance of this variability. It may be necessary to maintain a strict follow-up of these patients and to perform longitudinal studies to determine the prognostic value of this variable expression of PDC components in the onset of PBC.

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FRI0317

NOVEL CLASSIFICATION OF IDIOPATHIC INFLAMMATORY MYOPATHIES BASED ON DISTINCTIVE FEATURES AND AUTOANTIBODIES: ANALYSIS OF 67 KOREAN PATIENTS

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Background: Since Bohan and Peter first described their diagnostic criteria for idiopathic inflammatory myopathies (IIM) in 1975, new discoveries such as myositis-specific and myositis-associated autoantibodies (Abs) have been made.

Objectives: To investigate correlations between specific myositis Abs and their frequencies and clinical associations across different IIM groups, collectively demonstrating the utility of the new clinicoserologic classification in Korean adult patients with IIM.

Methods: We conducted a multicenter cohort study including 67 adult patients (age ≥ 18 years) who have been diagnosed as IIM by ENMC criteria. Immunoblot assay with Euroline strip (EUROIMMUN, Germany) was performed using the sera of definite dermatomyositis (DM, n=36), definite polymyositis (PM, n=25), amyopathic DM (n=4), DM sine dermatitis (n=1), and immune mediated necrotizing myopathy (IMNM, n=1). Patients were classified based on three classifications: 1) novel clinicoserologic classification suggested by Troyanov et al. in 2017. 2) 2017 EULAR/ACR classification criteria. 3) 2004 European neuromuscular center (ENMC) criteria. Associations of myositis Abs and clinical subsets of IIM were investigated.

Results: The distribution of the various IIM differed strikingly from those using the 3 classifications (Fig1). According to the 2004 ENMC classification and 2017 EULAR/ACR classification criteria, DM and PM was the most and the second frequent entities (DM: 55.2%, 56.7%; PM: 35.8%, 37.3%). But, using the new clinicoserologic classification, overlap myositis (OM) is the major type of IIM and the frequency of PM is significantly decreased. Anti-ARS Abs specificity included anti-Jo-1 (16.4%), -OJ (4.6%), -EJ (6.2%), -PL-7 (3.1%), and -PL-12 (4.6%). Interstitial lung disease was closely associated with anti-MDA5, and anti-ARS Abs, while DM-specific skin lesion was frequently observed in patients with anti-TIF1γ and anti-ARS Abs. Seven patients with cancer-associated DM were identified. They were positive for anti-TIF1γ (5/7) and anti-SRP (3/7) (table 1).

Table 3. Associations between myositis autoantibodies and distinctive clinical features

	Anti-ARS N=25	Anti-SRP N=24	Anti-MDA5 N=14	Anti-TIF1γ N=9	Anti-MG N=8	Anti-SAE N=3	Total N=87
ILD	20*	5	11*	0	3	1	36
Malignancy	1	3	0	5*	1	0	7
DM specific skin lesion	8*	6	10	9*	5	2	39
Dysphagia	2	2	2	3	2	1	16

*P<0.05, *P<0.001

Fig 1. Distribution of 67 patients according to 3 classifications for idiopathic inflammatory myopathies

Conclusion: Novel classification based on distinctive features and new myositis Abs reflects the clinical phenotype of IIM better. Establishment of a system routinely available to screen myositis Abs is needed. This will be beneficial to provide more precise diagnosis and proper management for patients with IIM.

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FRI0318

THROMBOCYTOPENIA AND MULTI-ORGAN INVOLVEMENT MIGHT BE POOR PROGNOSTIC FACTORS FOR SYSTEMIC SCLEROSIS CARDIOMYOPATHY

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Background: Systemic Sclerosis (SSc)-cardiomyopathy is associated with a high mortality in SSc. It is important to identify potential prognostic factors for SSc-cardiomyopathy.

Objectives: To describe the clinical characteristics and identify potential prognostic factors for patients with SSc-cardiomyopathy.

Methods: We retrospectively reviewed the clinical data of SSc-cardiomyopathy patients hospitalized at Sun Yat-Sen Memorial Hospital from January 1992 to November 2018. SSc-cardiomyopathy was defined as structurally and functionally abnormal of heart muscle in SSc patients with cardiac symptoms. Coronary artery disease, hypertension, valvular disease and congenital heart disease were excluded. The Chi-square test was used to compare the differences between the death group and the survival group.

Results:

1. There were 903 SSc patients recruited and 20 patients (2.2%) of them were SSc-cardiomyopathy. Among these 20 patients, 65% were females, age was 52.8±13.2 years, disease duration was 3.1±2.3 years, duration from SSc onset to cardiomyopathy occurred was 2.8±2.3 years.
2. SSc-cardiomyopathy developed in 18 patients (90%) with diffuse cutaneous SSc (dcSSc), and in 2 patients (10%) with limited cutaneous SSc (lcSSc). Eleven patients (55%) had pulmonary fibrosis, 7 patients (35%) had pulmonary arterial hypertension (PAH), 5 patients (25%) had scleroderma renal crisis (SRC) and esophageal involved respectively, 6

patients (30%) manifested thrombocytopenia and 4 patients (20%) manifested anemia.

- Cardiac symptoms included shortness of breath (n=18), palpitation (n=6), edema of lower extremity (n=4), fatigue (n=5) and/or cough (n=3). All patients were found to have cardiomegaly by echocardiography (UCG) or chest radiography. Left ventricular ejection fraction ($47 \pm 15\%$) were decreased. Ventricular wall motion abnormality was found in 5 patients (25%) by UCG. 15 patients (75%) manifested cardiac arrhythmia in electrocardiogram, with frequent premature ventricular contractions were the most common (n=10, 50%). Myocardial injury was found in 14 patients (70%) by electrocardiogram. Myocarditis was found in 7 patients (35%) by myocardial enzyme assay.
- Nine patients (45%) died, the causes of death were heart failure (n=4, 45%), sudden death (n=3, 33%) and ventricular tachycardia (n=2, 22%). 5 patients (55.6%) died within 1 month after the onset of cardiac symptoms. 3 patients died within 2 month, and one patient with implantable cardioversion defibrillation (ICD) died 2 years after ICD implanted. Ten patients survived in a mean 6 (1-10) years follow-up.
- Six patients (67%) manifested thrombocytopenia and 4 patients (44%) manifested anemia in the death group. However, only one patient (9.1%) in survival group manifested thrombocytopenia (p=0.007) and anemia (p=0.069). There were more organs and systems involved in the death group, compared to the survival group (4 ± 2 vs 2 ± 1 , p=0.044). 4 patients had SRC in death group, while 2 patients had SRC in survival group. 6 patients had pulmonary fibrosis in death group, while 5 patients in survival group (Table 1).

Table 1. Comparison between the death group and the survival group in Systemic Sclerosis cardiomyopathy patients.

	Death group (n=9)	Survival group (n=11)	p
Thrombocytopenia, n (%)	6(66.7)	1(9.1)	0.007
Anemia, n (%)	4(44.4)	1(9.1)	0.069
Pulmonary fibrosis, n (%)	6(66.7)	5(45.5)	0.343
Pulmonary arterial hypertension, n (%)	3(33.3)	4(36.4)	0.888
Scleroderma renal crisis, n (%)	4(44.4)	2(18.2)	0.202
Esophageal involvement, n (%)	3(33.3)	2(18.2)	0.436
Anti-SCL 70 positive, n (%)	5(55.6)	5(50)	0.653
Myocarditis, n (%)	4(44.4)	3(27.3)	0.423
Numbers of organ involved, mean (SD)	4(2)	2(1)	0.044

Conclusion: Thrombocytopenia and multi-organ involvement might be poor prognostic factors for patients of SSc-cardiomyopathy.

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FRI0319 ESOPHAGEAL EROSIONS PREDICT PROGRESSION OF LUNG DISEASE IN PATIENTS WITH SYSTEMIC SCLEROSIS

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Background: Interstitial lung disease (ILD) is the leading cause of death in Systemic Sclerosis (SSc) but its pathogenesis is not fully understood. Esophageal disease is common in SSc and micro aspiration of both acid and non-acid gastroesophageal reflux could be involved in the pathogenesis of ILD. Esophagogastroduodenoscopy (EGD) is an essential tool to evaluate disease severity of upper gastrointestinal tract involvement in SSc.

Objectives: The objective of the present study is to assess the role of EGD in predicting pulmonary functional deterioration in SSc patients.

Methods: One hundred and fifty patients with SSc and suspected esophageal involvement underwent EGD. Pulmonary function tests were performed at baseline and after a 36-months follow-up. Patients were characterized for disease phenotype, BMI, smoking exposure and medication history. A significant ILD progression was defined as a relative decline $\geq 10\%$ of FVC or a concomitant decline $\geq 5\%$ of FVC and $\geq 15\%$ of DLCO.

Results: One hundred and thirty-six patients (90.5%) were female with a mean age of 55.6 ± 13.8 years and 12.1% were active smokers. Fifty

patients (33.3%) had a diffuse cutaneous disease; 37.4% and 40.8% were positive for anti-centromere and anti-Scl70 antibodies respectively. The mean disease duration from the first non-Raynaud symptom was 5.9 ± 6.7 years. Sixty-one patients (40.8%) showed EGD signs of reflux esophagitis. Among them, 31.3% had an erosive form (9.5% grade A, 15.6% grade B, 4.8% grade C and 1.4% grade D according to Los Angeles classification). At the baseline, 23.1% of the patients had a FVC $\leq 80\%$ and 45.6% had a DLCO $\leq 50\%$. Patients with erosive esophagitis did not differ in terms of sex, age, duration and disease variant, positivity for anti-centromere, skin score values, FVC and DLCO at baseline compared to patients without erosions, but had a lower prevalence of anti-Scl70 (28.3 vs 52.5%, p=0.005) and active smoking (20.0 vs 8.4%, p=0.05). At follow-up, patients with esophageal erosions showed a greater relative decrease in FVC ($3.4 \pm 9.3\%$ vs $1.7 \pm 12.0\%$; p=0.013) without significant differences in the DLCO change. Overall, 11.0% of patients presented pulmonary disease progression. The presence of esophageal erosions was associated with a significantly greater risk of lung disease progression (OR 5.3, 95% CI 1.7-16.8, p=0.004) after paired correction for sex, age, duration of disease, auto-antibodies, skin involvement variant, baseline FVC and DLCO, smoke exposure and therapy with immunosuppressants, proton pump inhibitors, prokinetics, antiplatelet agents and prostanoids.

Conclusion: SSc patients with erosive esophagitis present a higher risk of progression of interstitial lung disease. This evidence supports a role of micro-aspiration of gastric contents in the development of inflammation and fibrosis of the airways.

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FRI0320 CN-1A AUTOANTIBODIES ARE SPECIFIC FOR SPORADIC INCLUSION BODY MYOSITIS

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Background: Sporadic inclusion body myositis (sIBM) is a subgroup of the idiopathic inflammatory myopathies (IIMs) and is characterized by both degenerative and autoimmune features. Unlike other IIMs, myositis-specific autoantibodies had not been found in sIBM patients until recently.

Objectives: We aimed to establish the prevalence and clinical associations of anti-CN-1A in a large Danish cohort with connective tissue diseases (CTD).

Methods: In a cross-sectional study design, a total of 568 participants (183 IIMs (55 sIBM, 128 non-sIBM: dermatomyositis (DM), polymyositis (PM), immune-mediated necrotizing myopathy (IMNM)), 164 systemic lupus erythematosus (SLE), 121 systemic sclerosis (SSc), and 100 blood donors (BD)) were tested for the presence of: a) CN-1A autoantibodies using a commercial Anti-CN-1A ELISA and b) myositis specific and associated autoantibodies (anti-Jo-1, anti-PL-7, anti-PL-12, anti-OJ, anti-EJ, anti-SRP, anti-Mi-2 α / β , anti-MDA5, anti-TIF1 γ , anti-NPX2, anti-SAE1, anti-PM-Scl75, anti-PM-Scl100, anti-Ro52, anti-Ku) using a commercial lineblot kit. The patients were classified according to the ACR classification criteria for IIMs (2017), SLE (1997) and SSc (1980), respectively. Clinical features were compared between anti-CN-1A positive and anti-CN-1A negative sIBM patients using two-sample t-test and Mann-Whitney test for continuous normally and non-normally distributed data and Chi-square test for categorical data, as appropriate.

Results: CN-1A antibodies were predominantly found in IIM patients. In the sIBM cohort, 24 patients (43.6%) were anti-CN-1A positive vs. 25 (19.5%) in the non-sIBM myositis cohort and 17 (10%) in the SLE cohort. None of the participants in the SSc or the BD cohorts had