- 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.

Disclosure of Interests: None declared

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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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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