Background: Patients with systemic sclerosis (SSc) are at risk of developing pulmonary hypertension (PH) of different etiologies. Two-dimensional and Doppler echocardiography is the reference screening tool and it is recommended yearly even in the absence of symptoms; however, elevated values of systolic pulmonary artery pressure (sPAP) are not detectable at rest in the early phases of the disease.

Objectives: The aim of the study was to determine whether early signs of right ventricular or right atrial dysfunction at speckle-tracking echocardiography might identify patients who are going to develop PH during follow-up.

Methods: We studied a consecutive cohort of 113 patients with a diagnosis of SSc according to the 2013 ACR/EULAR SSc classification criteria, undergoing screening for PH and who were followed-up regularly at a third-level centre of reference for SSc.

All patients underwent both the baseline echocardiographic examinations and at least one echocardiographic examination during follow-up in the referral center. Standard 2-D and Doppler echocardiography was performed with a Vivid 7 or Vivid E9 ultrasound system (GE Medical Systems, Norway); images and clips were saved for offline analysis on a GE workstation (EchoPAC PC SW). Clinical, laboratory and radiographical parameters, standard in the care of SSc, such as hemoglobin; albumin; CRP; ESR; platelet count; creatinine; fSG; ILD; CTD; ANA; anticentromere; anti-topoisomerase; and/or all-cause mortality during long term follow-up and after adjustment for classical cardiovascular risk factors.

RESULTS:

1. Disease duration: Mean age was 57.87 (SD 14.87) and mean disease duration was 12.14 years (SD 5.87).
2. Anti-Scl70 positivity was observed in 21 patients (44.7%) while anti-centromere positivity in 16 (34%). Pulmonary function tests and echocardiographic PAPs are resumed in Table 1. One out of 47 (2.1%) patients developed PAH. This was significantly lower in comparison to PAH prevalence (194 out of 1636, 11.8%) in Morriseoe et al [p 0.036].

Conclusion: The low prevalence of PAH recorded in this group of patients affected by SSc complicated by DUs suggests a protective role of the combination of Bosentan and Sildenafil against PAH, one of the most severe vascular manifestations of the disease. This data should be confirmed by prospective and controlled studies on larger populations.

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Our cohort consisted of 87 SSc patients, of which 64 (73.6%) were women, with a mean age at disease onset of 54.5 years (SD±19) and a median disease duration of 4.0 years (IQR 8.3). The median dp-ucMGP level at baseline was 634 pmol/L (IQR 393), which is greatly increased compared with 400 HCs (mean dp-ucMGP=393 pmol/L). Only one patient had prevalent CVD before onset SSc. Nine patients were lost to follow-up, in the remaining 78 SSc patients, 26 (33.3%) patients suffered from first CVD event during the study period, with a median time of 10.5 (IQR 15.2) years from onset SSc disease, corresponding to an incidence rate of 29.9 per 1000 person-years. Cardiovascular risk factors were not significantly different between patients with and without CVD. Odds ratios for sex (OR 2.44; 95%CI 0.89-6.72) and hypertension (OR 2.53; 95%CI 0.86-7.46) tended to predict CVD, but the association did not reach statistical significance. However, Kaplan-Meier analysis showed that elevated dp-ucMGP levels (>634 pmol/L) were associated with an increased risk for CVD and/or death during the first 10 years follow-up (log-rank test, P=0.006).

Conclusion: This study shows increased dp-ucMGP levels in SSc patients compared to age-matched HCs, indicating dp-ucMGP as a biomarker of disease. We confirm the high risk of CVD in SSc patients but traditional cardiovascular risk factors did not predict development of CVD. In contrast, high dp-ucMGP levels revealed an increased risk for CVD and/or death in SSc. It is still unclear what caused the increased dp-ucMGP levels in SSc patients, but given the strong, inverse association between dp-ucMGP and vitamin K status, a vitamin K deficiency is proposed. Whether this is caused by malabsorption or inflammation requires further research.

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Figure 1. Time to first CVD and/or death from onset SSc

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SuffleRATES IN PATIENTS WITH SYSTEMIC SCLEROSIS ARE AFFECTED BY CONCURRENT CARDIOPULMONARY DISEASE, GENDER, AND AUTOANTIBODY PROFILE: A 15-YEAR SINGLE CENTER EXPERIENCE

Keywords: Autoantibodies, Epidemiology, Systemic sclerosis

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Background: Systemic Sclerosis (SSc) is a systemic autoimmune disease with a high mortality rate [1-3].

Objectives: The purpose of this study was to assess long-term survival rates of patients with SSc in a single rheumatology center and identify factors affecting the survival rates.

Methods: Retrospective analysis of unselected adult patients with SSc, diagnosed and followed up in the Rheumatology Clinic of the University Hospital of Larissa in Central Greece for 15 years (2007-2022). Demographics and clinical characteristics were retrieved from medical records. While autoantibodies (auto-Abs) were assessed by indirect immunofluorescence and a SSc autoantibody line blot immunoassay (Euromimmun), Overall Survival (OS), 5-year and 10-year survival rates were calculated. The effect of age at disease onset, gender, clinical parameters, and autoantibodies were evaluated using Kaplan-Meier analysis.

Results: A total of 226 patients (85.8% female, mean age 61.9 years) were included. The mean ± SD duration of the disease was 12.2±4.9 years. The 5-year and 10-year survival were 95.9%, and 88.1%, respectively. In the majority of patients, the cause of death was related to SSc. The two main causes of death were interstitial lung disease (ILD) and heart disease (pulmonary arterial hypertension and/or arrhythmias). Older age at disease onset was independently associated with worse survival (59.9± 11.9 vs 47.3±14.6 years, p<0.05) and male patients had statistically worse OS compared to female patients (13.1 years vs 28.4 years, p=0.001). Patients with autoAbs against Th/To (12.1 years vs 28.2 years, p=0.03) against ILD had worse OS compared to autoAb-negative patients (12.1 vs 28.2 years, p =0.013, and 12.7 vs 28.8 years p=0.007, respectively). There was a trend for a better survival in patients with anti-centromere autoAbs (29.4 vs 26.2 years, p=0.064).

Conclusion: Our results show that the overall survival rates of patients with SSc have improved over the years than those reported in the past. Cardiopulmonary diseases still remain the main causes of death, while male sex is associated with worse prognosis. Additional prognostic factors have emerged from autoAb profiles.

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PULMONARY INVOLVEMENT IN VERY EARLY SYSTEMIC SCLEROSIS (VEDOSS): REPORT FROM A SINGLE CENTER

Keywords: Lungs, Systemic sclerosis

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Background: Previous work has described interstitial lung disease (ILD) in very early systemic sclerosis (VEDOSS) [1], but its progression over time is unknown. This can contribute decisively to the delineation of screening and monitoring strategies that have established guidelines.

Objectives: To evaluate the prevalence of ILD and its course in a cohort of patients who persist in a VEDOSS state, followed in a tertiary systemic sclerosis unit, over 10 years.

Methods: Retrospective observational study of patients persistently fulfilling VEDOSS criteria [2], with sequential measurement of diffusion lung capacity for carbon monoxide (DLCO) and forced vital capacity (FVC) at 0, 1, 5 and 10 years. Chest CT at baseline was considered, when available. A DLCO or FVC <80% of predicted values was considered indicative of lung involvement [1]. A decline of