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Table 1. The frequencies of non-vascular pulmonary manifestations in patients with Takayasu' arteritis

n (%)	All Patients n=197	Patients with PAH n=22	p
Symptoms	24 (12.2)	3 (13.6)	0.049
Cough/Dyspnea	4 (2)	3 (13.6)	0.000
Hemoptysis			
Pulmonary involvement in	10 (5.1)	5 (22.7)	0.000
CT	4 (2)	1 (4.5)	0.000
Pulmonary infiltrates	1 (0.5)	1 (4.5)	0.000
Nodules/cavities	1 (0.5)	1 (4.5)	0.000
Pulmonary hemorrhage	5 (2.5)	2 (9.1)	0.000
Pleural effusion			

(PAH: pulmonary arterial hypertension, CT: computed tomography)

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Scleroderma, myositis and related syndromes

FRI0300 IMPACT OF INTERSTITIAL LUNG DISEASE IN SYSTEMIC SCLEROSIS IN A COMPLETE, NATIONWIDE COHORT

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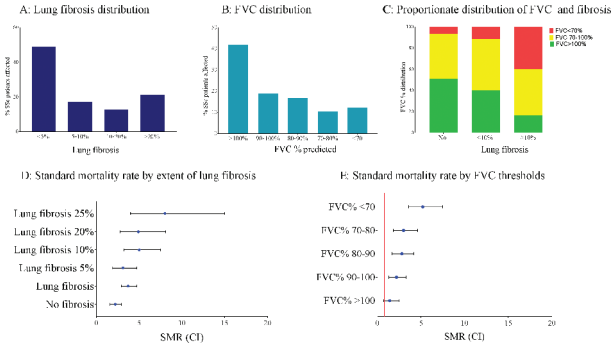
Background: Interstitial lung disease (ILD) represents a clinical challenge in systemic sclerosis (SSc) and associates with high mortality. The presence of severe lung fibrosis is a strong predictor for early mortality. There is substantial progress in SSc-ILD research, but precise, population-based data on cumulative incidence, range of severity and predictive value of clinical risk factors are lacking. Such data are vital for clinical decision making, and highly warranted as background information for appropriate development of screening and management strategies for SSc-ILD.
Objectives: To assess cumulative incidence of ILD, range of ILD severity and mortality risk predicted by baseline pulmonary function tests (PFT) and ILD extent by CT in a complete, nationwide SSc cohort.
Methods: The Norwegian SSc cohort study (Nor-SSc) includes all the 630 incident and 185 prevalent SSc patients from 2000-2012 meeting SSc classification criteria. A baseline PFT was recorded in 703 (86%) patients, and 650 (80%) had high resolution computed tomography (HRCT) images available for analyses. Extent of fibrosis was scored on 10 sections from every HRCT and expressed as percentage of total lung volumes. For the survival and mortality analyses, all Nor-SSc patients diagnosed from 2000-2012 (the 630 incident cases) were included and compared with 15 age- and gender matched controls per patient drawn from the national population registry. Vital status was available for all patients and controls at study end (January 2018). Descriptive statistics and standardized mortality rates (SMR) were estimated.
Results: Of the 815 patients in the total Nor-SSc cohort, 682 (84%) were female and 629 (77%) had limited cutaneous SSc. Mean age at SSc diagnosis was 53 yrs, with mean time from SSc onset to diagnosis of 3.8 yrs. We observed ILD on HRCT in 324/650 patients (50%), and the majority of these had <5% lung fibrosis (Figure 1A). Mean FVC at baseline was 94% of expected value, and nearly half of the patients (42%) had an FVC>100% (Figure 1B). Proportionate distribution of FVC values in patients with no lung fibrosis, <10% lung fibrosis of total lung volume and >10% lung fibrosis is shown in Figure 1C. During the mean 8.6 yrs observation period of this study, 148 of the 630 incident SSc patients died, corresponding to an overall SMR of 2.4. Separate analyses of the 650 patients with baseline HRCT data showed that the SMR correlated with presence and extent of lung fibrosis, from SMR 2.2 in patients with no fibrosis to SMR 8 in patients with >25% lung fibrosis (Figure 1D). Correspondingly, we found that the SMR changed across patient groups stratified by baseline FVC%, with increased mortality evident already in the FVC 90-100% group (Figure 1E).
Conclusion: The results from this population based SSc cohort study provide new, unbiased data regarding the impact of ILD. Our results indicate a dose-response relationship between lung fibrosis extent and SMR; and between FVC% and SMR. Importantly, this relationship was evident even in groups with limited lung fibrosis and groups with normal range FVC%, strongly suggesting that all SSc patients should be screened with PFT and HRCT at baseline, to diagnose ILD early and tailor further management.



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FRI0301 GASTROINTESTINAL ADVERSE EVENTS IN PATIENTS WITH SYSTEMIC SCLEROSIS-ASSOCIATED INTERSTITIAL LUNG DISEASE (SSC-ILD) TREATED WITH NINTEDANIB: DATA FROM THE SENSCIS TRIAL

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Background: In patients with idiopathic pulmonary fibrosis (IPF), nintedanib has a manageable adverse event (AE) profile characterised predominantly by gastrointestinal (GI) events. The efficacy and safety of