Background: Systemic sclerosis associated interstitial lung disease (SSc-ILD) is the leading cause of scleroderma-related mortality.

Objectives: This work identifies factors associated with SSc-ILD decline on pulmonary function testing (PFT).

Methods: This single center cohort identified 312 patients with ILD as determined by high resolution chest. 184 patients (59% of 312) completed baseline and serial PFTs (with at least two follow-up PFTs) and were included in this analysis. Mixed linear models were fit to assess the decline in the percent predicted forced vital capacity (ppFVC) over time. Demographics, disease factors, autoantibodies, and ILD features were included in the univariate mixed linear model; those achieving a p-value <0.20 were included in the multivariable mixed linear model. Patients were followed longitudinally, with survival as an endpoint identified using the National Death Registry Index, reviewing death certificates, and hospital records.

Results: The 184 patients were an average of 53.2 (12.1) years old; the median [QR] disease duration from the first non-Raynaud’s phenomenon symptom was 1.8 [0.7, 4.8] years. SSc subtype was diffuse in 55.4% (n=102), limited in 32.6% (n=60), overlap syndrome in 8.2% (n=15), and SSc sine scleroderma in 3.67% (n=7). Serologies were positive for anti-topoisomerase I (ATA), anti-centromere and anti-RNA polymerase III in 31.4% (n=53/169), 10.4% (16/154) and 22.9% (25/109) respectively. Mean ppFVC was 70.8 (18.9) and ppDLCO 572 (20.8). Whole lung involvement (WLILs) ≥20% on visual read was found in 49.3% of subjects (74/150 (49.3%)) where quantification was available. Over a median of 4.2 (4.6, 8.8) years, 21 patients (11.4%) died. The ppFVC declined a mean of 0.28/year in the overall group. There were differences in terms of ppFVC decline/year between patients who died in the first 2 years (n=10, -8.28%), 2-5 years (n=5, -3.89%), after 5 years (n=6, -1.0%) and patients who were still alive (n=163, -0.13%). The primary cause of death was ILD (6/21, 28.6%); those who died in the first 2 years most often died from progressive ILD (4/6, 67%). Factors significantly associated with decline in ppFVC on univariate analyses, included longer disease duration (ref. < 6 years, +0.0021), ATA positivity (ref. negative, P=0.0081), and WLILs ≥20% (ref. <20%, P=0.0484). In multivariate analysis the only statistically significant variable associated with decline in ppFVC was year at positivity.

Conclusion: In a large single center cohort of SSc-ILD, ATA positivity is a risk factor for developing progressive SSc-ILD, consistent with other SSc-ILD cohorts. Stratifying patients by survival demonstrates that lung function declines dramatically in those who died within 2 years, whose main cause of death was progressive ILD. These data support the growing need to identify risk factors for disease severity and risk for progression, and to target intervention in patients irrespective of sex and esophageal involvement.

Disclosure of Interests: None declared

DOI: 10.1136/annrheumdis-2022-eular.3648