European Respiratory Society
Annual Congress 2013

Abstract Number: 1262

Publication Number: P5119

Abstract Group: 1.5. Diffuse Parenchymal Lung Disease

Keyword 1: Imaging **Keyword 2:** Interstitial lung disease (connective tissue disease) **Keyword 3:** Lung function testing

Title: Scleroderma lung disease: A scleroderma physiologic index (SPI) derived from lung physiology and oxygen saturation

Dr. Katerina 13795 Antoniou kantoniou@med.uoc.gr MD [1], Prof. David 13796 Hansell pneumon@med.uoc.gr MD [2], Dr. Nicole 13797 Goh pneumon@med.uoc.gr MD [2], Dr. Katerina 13798 Samara kat_samara@hotmail.com MD [1], Dr. Elizabeth 13799 Renzoni pneumon@med.uoc.gr MD [2], Dr. Maher 13802 Toby pneumon@med.uoc.gr MD [2], Prof. Nikos 13803 Siafakas siafak@med.uoc.gr MD [1] and Prof. Athol 13816 Wells Athol.Wells@rbht.nhs.uk MD [2]. [1] Thoracic Medicine, Medical School, University of Crete, Heraklion, Greece and [2] Interstitial Lung Disease Unit, Royal Brompton Hospital, London, United Kingdom .

Body: The quantitation of disease severity in Scleroderma Lung (SSc-ILD) using pulmonary function tests is often confounded by emphysema. We identified the scleroderma physiologic index (SPI) with readily available measures (spirometry, diffusion capacity and oxygen saturation).Consecutive patients with a clinical/computed tomography diagnosis of scleroderma lung (n = 292) were divided into group I (n = 146) and group II (n = 146). Cohort 1 (derive cohort) and 2 (test cohort) did not differ in outcome, PFTs, age, gender, smoking status at baseline. There were 55 /146 deaths in cohort 1 and 50/146 deaths in cohort 2. The formula has been derived in cohort 1 and then calculated in test cohort 2. The SPI was derived in group I (by fitting pulmonary function tests against disease extent on CT) and was tested in Group II (test cohort). The formula for the SPI was as follows: 384.0 – (0.37x percent predicted diffusing capacity for carbon monoxide [DLCO]) – (0.59x percent predicted FVC) + 0.72x percent predicted FEV1) = (3.46 x saturation). In the test cohort, the SPI (median 40.9, range 9.2 to 82) was strongly predictive of mortality (HR 1.07, CI 1.04 -1.10, p<0.0005). SPI calculated from final formula and shown to predict survival more strongly than DLco, FVC, FEV1, Po2, Aag, satn, Kco or CPI. The SPI was always predictive of mortality and after inclusion of the SPI, no other variable was independently predictive of mortality. The SPI provides an easily calculated pulmonary function index, validated against mortality and potentially useful in the parametric quantification of disease severity, both for clinical purposes and to adjust for disease severity in clinical research.