Abstract Group: 4.3. Pulmonary Circulation and Pulmonary Vascular Disease
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Title: Clinical characteristics of scleroderma patients with and without pulmonary hypertension

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Body: Background: Scleroderma is a connective tissue disorder which is often complicated with pulmonary hypertension (PH). Aim: To investigate potential differences in the clinical characteristics of scleroderma patients with and without PH, estimated by resting Doppler echocardiography. Material-methods: A consecutive population of 82 scleroderma patients (11.9% male; 49.8 years old and 88.1% female; 54.9 years old), who were evaluated at the respiratory physiology laboratory of the Respiratory Failure Unit, constituted the study population. Patients underwent spirometry, diffusion capacity measurement and maximum cardiopulmonary exercise testing on a cycle ergometer. Data on anthropometric measurements, thorax computed tomography (CT) imaging and blood serology were also recorded. Results: Twenty eight patients (34.15%) presented with PH (systolic pulmonary arterial pressure >35 mmHg). Patients with PH had also lower FEV1 %predicted (p=0.001), FVC %predicted (p=0.002), IC %predicted (p=0.01), TLC %predicted (p=0.03), FRC %predicted (p=0.001), and DLCO %predicted (p<0.001) and exhibited more often pulmonary fibrosis in CT scan, versus those without PH (p=0.03). As expected, those with PH had lower maximum oxygen uptake %predicted compared to the ones without PH (p=0.02), while no difference was noted in the autoantibodies subtype (antisclero-70 or anti-centromere) between the two groups. Conclusion: In the current study most patients with PH also presented with respiratory restriction. It seems that pulmonary fibrosis plays an important role in the presentation of PH in scleroderma patients.