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Title: Pulmonary function involvement in scleroderma

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Body: Scleroderma is a systemic disease characterized by a severe inflammatory process with exuberant fibrosis. The pulmonary involvement (interstitial disease and/or pulmonary hypertension) is one of the first causes of mortality and morbidity. Aim: To describe the functional involvement of the pulmonary disease in scleroderma. Subjects and methods: Scleroderma patients with pulmonary involvement who performed pulmonary function tests (spirometry, diffusion capacity DLco, pulmonary volumes), walking tests, chest X-ray and echocardiography in 2012. Results: 24 patients were studied, mean age 52 years, 23 women; 9 had limited cutaneous scleroderma, 12 had interstitial radiologic involvement; 16 patients had exertional dyspnoea and 5 of them exercise desaturation. Five patients had normal pulmonary function testing. Although the mean values were normal for volumes and flows (FVC 88%, FEV1 85%, TLC 84% of the predicted values), 8 patients had restriction. Mean DLco was 68.7% (range 44–101%); decrease of DLco was found in 19 patients (79%); in 11 of them this was the only resting functional abnormality. Among patients with diffusion capacity impairment, only 14 had exertional dyspnoea and only 9 had pulmonary radiologic abnormalities. Echocardiography was performed in 15 patients and 2 of them had pulmonary hypertension with normal pulmonary volumes and impaired diffusion capacity. Conclusion: Complex pulmonary function tests including DLco are necessary for scleroderma patients with pulmonary involvement. The DLco is abnormal in the majority of cases, even without exertional dyspnoea or restriction. The echocardiographic evaluation is useful even in patients with mild decrease in DLco, due to the worse prognosis of pulmonary hypertension.