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Title: Observational retrospective study on the treatment of pulmonary hypertension (PH) associated to pulmonary disease in patient with PH out of proportion

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Body: Background: ERS guidelines recommend the use of specific drugs in Idiopathic Pulmonary Arterial Hypertension. Patients with out of proportion PH due to lung diseases should be enrolled in clinical trials targeting PAH-specific drug therapy. Objective: Aim of our retrospective study is evaluate the effects of treatment of PH associated to lung disease in patients with PH out of proportion on the exercise capacity and hemodynamic parameters. Methods: Fifteen subjects affected by PH out of proportion secondary to chronic hypoxia and miscellaneous (group III and V) were enrolled in our study. First line specific drug therapy was chosen randomly between bosentan or sildenafil. In some patients association of two or more drugs was performed. Spirometry, functional class assessment and six minute walking test (6MWT) were performed at baseline and after 6 and 12 months of therapy. Cardiac heart catheterization was performed at baseline and after 12 months. Results: In our patients there were no changes in NYHA functional class, no statistical difference of distance and dyspnea at 6MWT after 6 and 12 months of treatment. Concerning hemodynamic parameters we observed a decrease of mean pulmonary artery pressure (PAP m) of 8.900 mmHg (p=0,1289), a decrease of pulmonary resistance (PVR) of 113.50 dyne x sec x cm-5 (p=0,0488) and an increase of cardiac index (CI) of 0.7840 l/min/m2 (p=0,1055). Conclusions: The improvement of hemodynamic parameters and preservation of clinical status in our patients suggest the efficacy of treatment with specific drugs in PH due to lung disease in patients with PH out of proportion.