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Title: Effects of oral dual ERA therapy on pulmonary function testing and 6MWT in patients with idiopathic pulmonary fibrosis and pulmonary hypertension

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Body: PH can be present in 37% to 59% of patients with IPF and is a predictor of mortality, which appears to be the cause of additional burden of exercise capacity. The aim of study was to investigate the role of oral dual ERA therapy in 2 groups of patients with PH associate to IPF with or without Bosentan therapy during 24 months. The primary endpoint was the change of exercise capacity up to month 24, measured by a modified 6MWT.We evaluated retrospectively 16 patients: 8 pts (6 M; mean age of 72 yrs old) with moderate-severe PH and IPF (mPAP measured by RCH estimated as a mean value of 38 ± 4.8 mmHg), treated with Bosentan (PH-IPF ERA group); 8 pts (5 M; mean age of 70 yrs old) with moderate PH and IPF not treated with Bosentan (PH-IPF control group).At baseline, there were differences about hemodynamic and pulmonary functional test profile between two groups of pts. At T 24, the 6MWT increased in PH-IPF ERA group showing a mean increase of +150 mt, while in the PH-IPF control group we observed a reduction of 41% compared to the 6MWT values baseline (p 0,0003).In PH-PID group treated with ERA, FVC% and DLCO% were reduced of 8% and 32%, respectively, while in PH-IPF control group were reduced of 6,6% and 22% compared to the baseline (p 0,009).The NYHA functional class was decreased in group treated with ERA (3,5 vs 2; p 0,009) and improved in control group (2,12 vs 3,25; p 0,007). In patients with moderate-severe PH and idiophatic pulmonary fibrosis, treated by Bosentan, there was a clinical improvement, measured by a six minute-walk test and a change in NYHA functional class, without affecting pulmonary functional test.