A Young Patient With Pulmonary Arterial Hypertension

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Introduction: Pulmonary hypertension (PH), a serious disease that could lead to death. Due to its non-specific symptoms it might be diagnosed with 2.5 years delay. Here we reported an early diagnosed young patient with PH.

Case: 30 year old female admitted with progressive dispnea for five years. TTE revealed high (70mmHg) pulmonary artery systolic pressure (PASP) and no left heart pathology. After the evaluation she had the diagnosis of idiopathic pulmonary arterial hypertension. In her hysical examination atteunated 2nd heart sound was auscultated. Pro-BNP was high (260ng/ml). Right heart cathaterisation (RHC) revealed PAPmean: 54mmHg, PCWP: 10mmHg. 6MWD was 320m and she desaturated during the test. Bosentan was started . She had benefit with initial treatment (6MWT 520m) but she had progression at 30th month (6MWD:310m, Pro-BNP:324ng/ml, PASP:110mmHg). Inhaler ilioprost was added. At 60th month sildenafil was added due to progression of dispnea. At 72th month progression with bilateral pretibial edema in lower extremities was detected (FC III, 6MWD:280m, Pro-BNP:382ng/ml, PABmean:75mmHg). Intravenous epoprostenol infusion was considered due to clinical detoration. At 90th month her FC was I and 6WMD increased to 500m with iv epoprostenol 25ng/kg/min.

Conclusion: Early diagnosis of PH improve the survival of the patients. If there is no satisfying answer is gained during the monotherapy, it should be switched to combination therapy. The patients with progressive dispnea under the treatment of combination therapy, IV epoprostenol treatment should start without any delay.