[Treatment of idiopathic pulmonary arterial hypertension.]

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Abstract: Background. In the past 5-10 years, drug treatment of idiopathic pulmonary arterial hypertension has evolved considerably. Experience and results from use of such updated treatment in Norway has not been reported. Material and method. 32 patients newly diagnosed with idiopathic pulmonary arterial hypertension, were consecutively assessed with respect to hemodynamics and physical capacity. The results after three months were compared with those after 12 months. Observed survival was compared with estimated survival from the time when only conventional treatment was available. Results. The patients (78% women) were 42 ± 14 years, had dyspnea in NYHA class 2.9 ± 0.4 and a maximal oxygen uptake of 12.0 ± 3.9 ml/kg/min (37 ± 13% of the expected). Updated treatment led to significantly improved hemodynamics and physical capacity, which persisted during follow-up. During 43 ± 31 months follow-up, seven patients died while two underwent bilateral lung transplantation. Observed transplantation-free survival was 81% after one, two and three years, while that for estimated transplantation-free survival was 70%, 58% and 49% respectively. Interpretation. Treatment of idiopathic pulmonary arterial hypertension with updated treatment improves hemodynamics and thereby symptoms. Mortality remains high, but is probably lower than it was when only conventional treatment was available.

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