

Results of Treatment with Endothelin Receptor Antagonist and Prostacyclin Analogs for Patients with Pulmonary Arterial Hypertension (PAH)

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Introduction: The short and long term consequences of treating PAH patients with endothelin antagonist and prostacyclin analogs are evaluated in this study.

Methods: 25 PAH Patients (16 girls, 9 boys), followed from 2001 to 2012, are included in this study. Patients have been evaluated pre and post treatment with the following tests: cardiac catheterization(flows ratio, average pulmonary arterial and systemic arterial pressure, pulmonary and systemic vascular resistance index), echocardiography (right ventricular pressure), functional capacity (NYHA) and 6-minute walking distance (6-MWT).

Results: The average age of patients is 12.5 years(2-28years) with an average treatment start age of 9 years . The average weight is 30 kg . The average duration of treatment is 18 months and the average period between two hemodynamic studies is 24 months . 16% of the cases have idiopathic pulmonary arterial hypertension (IPAH), while %84 of the cases have innate heart disease with shunt. 32% of the patients have down syndrome. 52% of the cases have been treated with 52% prostacyclin analog (Ilioprost) while 48% have been treated with endothelin antagonist (Bosentan). The treatment of 3 patients (%12) have been switched from Ilioprost to Bosentan (1 due to disphony and resistance to treatment,1 due to resistance to treatment and 1 due to treatment nonconformity) There have been statistically significant drops in average pulmonary arterial pressure, PVRI and NYHA functional capacity when compared to pre-treatment levels, while a statistically significant increase in 6-WMT is observed (p-value=0.05). There is not a significant difference detectable pulmonary arterial pressure detectable via echocardiography. The difference in clinical and hemodynamic parameters between patients treated with Ilioprost and those treated with Bosentan is not found to be statistically significant

Conclusion: Specific PAH treatment improves quality of life of patients while the difference in the effects of various medications to the clinical and hemodynamic parameters is not significant. The medication preference should therefore be based on the age of the patient, the conformity of the medication to the patient and/or the family of the patient, ease of use, the period it takes until the medication starts becoming effective and side effects of the medication.