Results of Treatment With Endotelin Reseptör Antagonist and Prostacylin Analogs for Patients With Pulmonary Hypertension(PAH)

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INTRODUCTION

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

Table
Pre and post treatment values with the following tests: cardiac catheterization 6MWT and NYHA

Parameters	*Pretreatment	*Posttreatment	p ; Z
Mean PAP (mmHg)	71 (44-108)	65 (41-95)	0.002 ; -3.118
PVRI (WUm^2)	20.23 (6.66-46)	15.98 (4.05-38.6)	0.001 ; -3.296
PVRI/SVRI	0.89 (0.38-1.68)	0.84 (0.17-1.47)	0.007 ; -2.677
6-MWT (m)	340 (240-420)	470 (340-540)	0.000 ; -3.631
NYHA (functional class)	2.52 (1-3)	2.48 (1-3)	0.001 ; -3.317

PAP: Pulmonary arterial pressure, PVRI: Pulmonary vascular resistance index, SVRI: Systemic arterial pressure index, 6-MWT: 6 minutes walk test, (*)Median(min-max)

PATIENTS &METHOD

Patients with diagnosis of PAH (n=25; 16 girls and 9 boys), followed between 2001 and 2012, were included in the study. Patients were evaluated in pre and post treatment period 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 test (6-MWT). The age range of patients was 2-28 years (median: 12.5 years), average age of treatment beginning was 9 years (range: 2-17 years). The weight range was 6-60 kgs (median: 30 kgs). The average duration of treatment was 18 months (range: 6 -120 months) and the average period between two hemodynamic studies was 24 months (range: 6-156 months).

RESULTS

Patients with idiopathic pulmonary arterial hypertension (IPAH) were 16% of all cases, while 84% had congenital heart disease with shunt and 32% of the patients had Down syndrome. Patients treated with prostacyclin analog (Ilioprost) were 52% of the cases, while 48% were treated with endothelin antagonist (Bosentan). The treatment of 3 patients (12%) were switched from Ilioprost to Bosentan (one due to disphony and resistance to treatment, one due to resistance to treatment and one due to treatment nonconformity). There were statistically significant decreases in average pulmonary arterial pressure, PVRI and NYHA functional capacity, when compared to pre-treatment levels; and a statistically significant increase in 6-WMT was observed (p-value=0.05) (Table). There was not a significant difference in pulmonary arterial pressure via echocardiography (p>0.05). The difference in clinical and hemodynamic parameters between patients treated with Ilioprost and those treated with Bosentan was not statistically significant (p>0.05).

CONCLUSION

Specific PAH treatment improves quality of life of patients, while the difference in the effects of various medications on 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.