

Switch from bosentan to macitentan in children and adolescents with pulmonary arterial hypertension during a hospitalization period

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Introduction: Macitentan is the most effective of the known endothelin receptor antagonists approved for the treatment of pulmonary arterial hypertension (PAH) in adult patients. Randomized controlled studies have shown that macitentan improved functional class, long-term outcomes, reduced disease progression and mortality, as compared to previous therapy. In children, the application experience is limited to a few small studies.

Materials and methods: during the period from June 2018 to December 2018, 8 patients, including 3 boys and 5 girls aged 13.4+ 7 years, including 2 – LAG-CHD, 6 – ilag, who received therapy with bosentan and sildenafil for 2 to 7 years, were switched from bosentan+sildenafil to the macitentan and sildenafil therapy. The reason for the change of therapy was the worsening of the patient's condition in the presence of the maximum therapeutic doses of sildenafil and bosentan. All but one patient received 10 mg of macitentan a day.. One girl of 9 years with idiopathic PAH received 5 mg of macitentan per day. Duration of treatment in cardiology department was from 7 to 10 days. All patients underwent standard laboratory tests, liver ferments analysis, ECG, ECHOCARDIOGRAPHY, 6-minute walk test, cardiopulmonary test.

Results: the functional class decreased from 3.1+0.64 to 2.6+/- 0.74 (p=0.17), the distance at 6 minutes walk test increased from 390+99 to 494+111 m (p=0.067), the excursion of the fibrous ring of the tricuspid valve increased from 17.5+4.8 to 20.5+4.9 mm per second (p=0.23)

Conclusions: the change of therapy in children with primary pulmonary hypertension and pulmonary hypertension associated with congenital heart disease was performed. Combination therapy with sildenafil + bosentan was switched to sildenafil+macitentan. There was a decrease in the liver enzymes level. This switching is safe and effective. This is confirmed by short-term observation in a cardiological department. Such a switchover can improve the condition of children with PAH without joining the therapy of the third specific drug.