

Effect of bosentan in adults with pulmonary arterial hypertension due to congenital heart disease with and without Down's syndrome.

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Background. Oral bosentan is effective in pulmonary arterial hypertension (PAH) related to congenital heart disease (CHD). In literature, the effect of bosentan in patients with Down's syndrome is largely unknown. Aim of the study was to evaluate the long-term effects of bosentan in adult patients with CHD-related PAH with and without Down's syndrome.

Methods. WHO functional class, resting oxygen saturation, 6-minute walk test (6MWT) and hemodynamics were assessed at baseline and after 24 months of bosentan therapy in patients with CHD-related PAH with and without Down's syndrome.

Results. Ninety-five consecutive patients were enrolled: 25 with and 70 without Down's syndrome. After 24 months of bosentan therapy, both with and without Down's syndrome patients showed an improvement in WHO functional class (Down: 2.4 ± 0.5 vs 2.8 ± 0.6 , $p=0.005$; controls: 2.5 ± 0.5 vs 2.9 ± 0.5 , $p=0.000002$), 6-minute walk distance (Down: 281 ± 66 vs 232 ± 69 m, $p=0.0007$; controls: 383 ± 75 vs 336 ± 81 m, $p=0.00003$), and hemodynamics (pulmonary flow, Down: 3.7 ± 1.5 vs 3.2 ± 1.3 l/m/m², $p=0.006$; controls: 3.2 ± 1.3 vs 2.5 ± 0.9 l/m/m², $p=0.0005$; pulmonary to systemic flow ratio, Down: 1.3 ± 0.6 vs 0.9 ± 0.3 , $p=0.003$; controls: 1.0 ± 0.6 vs 0.8 ± 0.2 , $p=0.012$; pulmonary vascular resistance index, Down: 15 ± 9 vs 20 ± 13 WU.m², $p=0.007$; controls: 20 ± 10 vs 26 ± 15 WU.m², $p=0.002$). We did not find any difference in the efficacy of therapy between the two groups.

Conclusions. Bosentan was safe and well tolerated in adult patients with CHD-related PAH with and without Down's syndrome during 24 months of treatment. Clinical status, exercise tolerance, (evaluated by 6MWT), and pulmonary hemodynamics (evaluated by right heart catheterization), improved, regardless of the presence of Down's syndrome