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Title: Efficacy and safety of oral bosentan in patients with Down's syndrome and pulmonary arterial hypertension due to congenital heart disease

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Body: Aim. To evaluate the long-term effects of oral bosentan in adult patients with pulmonary arterial hypertension (PAH) due to congenital heart disease (CHD) with and without Down's syndrome. Methods. WHO functional class, 6-minute walk test (6MWT) and hemodynamics were assessed at baseline and after 12 months of bosentan therapy in CHD-PAH patients with and without Down's syndrome. Results. Seventy-four consecutive patients were enrolled: 18 with and 56 without Down's syndrome. After 12 months of bosentan therapy, both groups showed an improvement in WHO functional class, 6-minute walk distance and hemodynamics.

Clinical and haemodynamic variables before and after oral bosentan treatment in patients with and without Down's syndrome

	Down's syndrome (n=18)			No Down's syndrome (n=56)		
	Basal	Follow-up	p	Basal	Follow-up	p
WHO functional class	2.9±0.6	2.5±0.5	0.005	2.9±0.5	2.5±0.5	0.000002
Travelled distance (m)	239±74	288±71	0.0007	343±86	389±80	0.00003
mPAP (mmHg)	66±21	60±17	0.06	74±18	73±21	0.6
QP (l/min/m2)	3.5±1.4	4.0±1.6	0.006	2.8±1.0	3.5±1.4	0.0005
QP/QS	1.0±0.4	1.4±0.7	0.003	0.9±0.3	1.1±0.7	0.012
PVRi (WU.m2)	20±13	15±9	0.007	26±15	20±10	0.002

mPAP, mean pulmonary arterial pressure; QP: pulmonary cardiac index; QP/QS, pulmonary to systemic cardiac index ratio; PVRi, pulmonary vascular resistances index.

No differences in the efficacy of therapy were observed 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 12 months of treatment. Clinical status, exercise tolerance, and pulmonary hemodynamics improved, regardless of the presence of Down's syndrome.