

Subcutaneous treprostinil in pediatric patients with severe pulmonary hypertension.

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Background: Intravenous epoprostenol is first-line treatment for functional class (FC) III/IV Pulmonary Hypertension (PH), but infusion system complications can increase morbi-mortality. Subcutaneous (s.c) Treprostinil (stable prostacyclin analogue) infusion can avoid central catheters complications. Nevertheless, there is limited information about efficacy/safety of this treatment in children. Methods: We report treprostinil use in seven pediatric patients , with mean age of 3.7 ± 4.5 years (3 months - 11 years) and weight of 13.6 ± 13.9 (3 to 39 kg), with severe pulmonary hypertension (Pulmonary/systemic pressure ratio $104\% \pm 7$, PVRI $12,5 \pm 5,7$ WU/m², PVRI/SVRI $1 \pm 0,1$) in functional class III-IV ($3,6 \pm 0,4$), in spite of combined treatment with sildenafil and bosentan. The etiologies of PH were: idiopathic (n=2), residual PH after CHD surgery (n=2), Eisenmenger (n=1), PH associated to metabolic disease (n=2). Results. Treatment was initiated in seven patients and stopped after two weeks in three (one case due to local intolerance and in two, after definitive diagnosis of metabolic disease with neurologic impairment). In the four patients in which treatment was continued, mean treprostinil dose was 31 ± 3.4 ng/kg/min. Mean follow-up was 15 ± 4 months (11- 19 months). At the end of follow-up, FC had improved from III-IV ($3,6 \pm 0,4$) to I-II ($1,2 \pm 0,5$), and in two patients, (3 and 8 months old, with idiopathic and postoperative PH respectively) pulmonary pressure estimated by eco had dropped to near normal levels. Observed side effects were: four days of significant local pain treated with ibuprofen or paracetamol (n=1), and mild epistaxis (n=1). Conclusions: 1) Tolerance to s.c. treprostinil was good in most of our patients (Only 1/7 patients (14%) didn't tolerate s.c. treprostinil due to local pain. 2) Subcutaneous treprostinil was safe and effective in PH pediatric patients in FC III/IV in combined treatment with sildenafil and bosentan.