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Targeted pulmonary vasodilator therapy in children.
Experience of a single reference center.

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Introduction: The management of pulmonary arterial hypertension (PAH) in children is complex because most evidence-based practices derive from adult PAH studies.
Aims/Methods: We retrospectively analyzed all consecutive patients (pts) followed in a PAH consultation that began targeted therapy between 2003 and 2015.
Results: We analyzed 41 pts, of which 54% were female. PAH was idiopathic on 12.2%, associated with congenital heart diseases on 46% and with other diseases on 4.9%. Separately we formed a group with pts that underwent Glenn/Fontan surgery (36.6%), all with high pulmonary vascular resistance (PVR). All pts began targeted therapy (mean age 6.1 5.8 years): 61% with bosentan (B); 17% with sildenafil (S), 19,5% double therapy with B+S or a prostacyclin (P) and 2.4% triple therapy with B+S+P.
Evaluation before PAH therapy implementation showed: 1) Functional class III or IV on 78%, right heart failure (RHF) on 51.2% and syncope on 8.1%; 2) Mean of 6-minute walk test (6MWT): 408.6 105.2 meters, performed in 22% of pts; 3) Mean BNP: 748.3 1704.5 pg/mL; 4) On echocardiogram: mean PASP: 80 16.4 mmHg; right ventricle (RV) dilatation on 51.2% and pericardial effusion on 2.4%; 4) CATH: mean right atrium pressure (RAP): 7.1 4.6 mmHg, mPAP: 44.6 22.8 mmHg, cardiac index: 3.2 1.5 L/min/m2, PVR: 16 1.5 U Wood and SvO2: 63.9 10.8%; all pts have negative AVT; 5) 46.7% of pts had high pressure on Glenn/Fontan.
The mean follow-up time after PAH diagnosis was 4.8 5.5 years. Two pts were submitted to percutaneous atrial septostomy and one to an attempting of Potts shunt. After at least 3 months of targeted therapy, we found, although without statistical significance, improvement in most of the pts, on the functional class, 6MWT, mean BNP value, mPAP and on the pressure of Glenn/Fontan circuit. Overall the global mortality was 24.4%. At 5 years, the Kaplan-Meier estimates a survival of 65.5%.
Conclusions: Paediatric PAH is a progressive disease whose mortality rate remains high even with early targeted therapy implementation. Pts with Fontan circulation may benefit of early targeted therapy as form of prevention of circuit failure.