

## **Pulmonary arterial hypertension in congenital heart anomalies with left-right shunt**

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Introduction: Pediatric cardiology is mainly engaged in congenital heart anomalies (CHA), of which two thirds are anomalies with left-right (LR) shunt. Pulmonary arterial hypertension ( PAH ) present a secondary increase in pulmonary arterial pressure at rest > 25 mmHg or > 30 mmHg during loading, during the catheterization. In clinical practice elevation of pulmonary vascular resistance (PVR) leading to irreversible changes in pulmonary vasculature that are contraindications for operative treatment.

Aim: to evaluate incidence of the pulmonary arterial hypertension in congenital heart anomalies with left-right shunt

Methods: selective group patient with PAH based on clinical examination, ECG, X ray, 2D echo and final diagnostical catheterisation were divided in two groups : fixed and nonfixed PAH

Results: In the period between april 1997. and november 2011 was performed a total of 216 pediatric cardiac catheterization, (44% of all surgical - 489 patients), 169 (79%) diagnostic, 47 (21%) of interventional catheterization. In 45 (26%) patients was to estimate PAH (girls 25). In 15 (33%) patients we have fixed (irreversible) PAH. From that group in 9 patients (60%) PAH was associated with Down syndrome. 30 (66%) patients after cardiac catheterization were with non-fixed (reversible) PAH is not operated on four children (13%). At this point, 15 patients with PAH is evaluating and treating.

Conclusion: Assessment of PAH is imperative for operative correction and inoperable patients require specific treatment prostanoids, endothelin receptor antagonists and inhibitors of phosphodiesterase type-5, alone or in combination.

Key words: pulmonary arterial hypertension, congenital heart anomalies, treatment.