

**Acute vasodilator testing with 100% oxygen can predict adverse outcomes in children with idiopathic and heritable pulmonary arterial hypertension**

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**Objectives**

Acute pulmonary vasodilator testing (AVT) appears to be important for children with idiopathic pulmonary arterial hypertension (IPAH) to identify those who have a favorable outcome. Although several different vasodilators including intravenous epoprostenol and inhaled nitric oxide have been used, the pulmonary vasoreactivity assessed by 100% oxygen has not been well established. The aim of this study was to the prognosis value during AVT with 100% of oxygen in children with IPAH and heritable PAH (HPAH).

**Methods**

A retrospective study was designed to evaluate the in 50 children (younger than 19 years of age) with IPAH/HPAH. A positive response to the AVT is defined as a reduction of pulmonary vascular resistance index (PVRI)  $\geq 20\%$ , unchanged or increased cardiac index (CI), and decrease or no change in the ratio of pulmonary vascular resistance to systemic vascular resistance (Rp/Rs ratio). The adverse events included hospitalization due to heart failure, lung transplantation, and cardiac mortality.

**Results**

The median age was 15 years with 22 males and 28 females. Twenty-two children (44%) were acute vasodilator responders. All children with HPAH were non responders. Although responders had lower brain natriuretic peptide levels (20.4, 4.5-203 pg/ml vs 54.8, 7.6-244.9 pg/ml,  $p < 0.05$ ), hemodynamics at baseline including mean pulmonary arterial pressure, PVRI, CI, and Rp/Rs ratio were not significant difference between responders and non-responders. During follow-up period (median 20 months), 27 (54%) children had an adverse event. Cumulative event-free survival rate in responders was significantly higher than those in non-responders (2-year event-free survival rate; 87% vs 33%, log-rank test,  $p < 0.05$ ).

**Conclusions**

Although hemodynamic data at baseline was not significant difference between responders and non-responders, non-responders may have worse outcomes compared with responders. AVT with 100% oxygen is safe, convenient, and useful for predicting adverse outcomes in children with IPAH and HPAH.