MP3-2

Pulmonary Vasodilator Therapy in Borderline Pulmonary Arterial Hypertension due to Congenital Heart Disease: Treat and Repair or Repair and Treat?

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Introduction: Pulmonary arterial hypertension (PAH) associated with congenital heart disease (CHD) is an important issue both in the adult and pediatric population that may influence outcome and even preclude repair. Pulmonary vasodilators (PV) in borderline cases, before or after surgery, may improve PAH and possibly change the operability criteria for these patients.

Methods: This study reports our experience using PV in 2 groups of CHD patients with borderline PAH: 1) patients with initially prohibiting PAH, where PV improved PAH enough to permit intervention (treat and repair), 2) patients initially deemed operable but developing significant postoperative PAH (repair and treat).

Results: The 1st group included 12 patients aged 13.0±14.2 (0.9-43) years, 4 with single ventricle (SV) physiology. Pulmonary vascular resistance index (PVRI) was 6.4±2.6 (4.4-10.1) Wood units (WU) in the nonSV and 2.5±0.6 WU in the SV patients, not permitting further intervention. PV therapy for 2.0±1.3 years improved ($p<0.01$) systolic and mean pulmonary pressure, PVRI and PVRI/SVRI and permitted repair in the nonSV patients, Fontan in 2 and fenestration occlusion in 2 SV patients. PV were discontinued in 4 patients 0.9±0.3 years after intervention without problems for 5.6±1.4 years subsequent follow up.
The 2nd group included 12 patients aged 13.1±21.1 (0.4-72.7) years, 6 with SV physiology. PV were instituted postoperatively before discharge in 3 nonSV patients due to increasing RV estimated pressures and 6 SV patients due to increasing central venous pressures and pleural effusions. Therapy was discontinued after 2.6±1.9 years after invasively measured improvement in 5 patients without problems for 4.2±3.2 years subsequent follow up. PV were instituted 2 months to 2.8 years postoperatively in 3 patients with septal defects due to invasively measured increased pulmonary pressures and resistances and patients remain on PV during 2.0-8.0 years.

Conclusions: PV in borderline PAH due to CHD may help to manipulate pulmonary vascular resistance pre and postoperatively, permit repair or intervention in otherwise prohibiting resistances and possibly shift the operability criteria in these patients. Further research with larger populations and longer follow up is needed before recommendations for pre and/or postoperative PV therapy can be issued and followed.