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


Dept of Paediatric Cardiology & Adult Congenital Heart Disease, Onassis Cardiac Surgery Center, Athens, Greece

**Introduction:** Pulmonary arterial hypertension (PAH) due to 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.

**Purpose:** This study reports our experience using PV in 2 groups of CHD patients with borderline PAH.

**Patient groups**

1. **Treat and repair group**
   - Patients with initially prohibiting PAH, where PV improved PAH enough to permit intervention

2. **Repair and treat group**
   - Patients initially deemed operable but developing significant postoperative PAH

**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
  - possibly shift the operability criteria in borderline patients

- This is an observational study and further research with larger populations and longer follow up is needed before changes in the current recommendations may be contemplated

**Conflict of interest:** none declared

---

**Results Treat and repair group:**

- 12 patients aged 13.0±14.2 (0.9-43) years
- 4 with single ventricle (SV) physiology
- Pulmonary vascular resistance index (PVRI) not permitting further intervention
  - 6.4±2.6 (4.4-10.1) WU in the nonSV and 2.5±0.6 WU in the SV patients
- PV therapy for 2.0±1.3 years improved (p<0.01) haemodynamics (see diagrams) and permitted intervention:
  - repair in the nonSV patients
  - Fontan in 2 and fenestration occlusion in 2 SV patients
- PV discontinued in 4 pts 0.9±0.3 yrs after intervention without problems for 5.6±1.4 years subsequent follow up

**Results Repair and treat group:**

- 12 patients aged 13.3±21.1 (0.4-72.7) years
- 6 with SV physiology
- PV instituted postoperatively before discharge in:
  - 3 nonSV patients due to increasing RV estimated pressures by ECHO
  - 6 SV patients due to increasing central venous pressures and pleural effusions
- PV discontinued after 2.6±1.9 years in 5/9 pts 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
  - 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
  - possibly shift the operability criteria in borderline patients

- This is an observational study and further research with larger populations and longer follow up is needed before changes in the current recommendations may be contemplated

**Conflict of interest:** none declared