

Follow-up results of transcatheter pulmonary valvotomy in patients with pulmonary atresia and intact ventricular septum

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Objectives. We investigate the long term outcomes of patients with pulmonary atresia and intact ventricular septum (PA-IVS) following transcatheter pulmonary valvotomy.

Methods. Between 1995 and 2010, 55 neonates with PA-IVS and tricuspid valve Z-score ≤ -3.5 (ranging from -3.5 to 0.5 , mean -1.1 ± 0.9) underwent attempted transcatheter decompression of right ventricle. A radiofrequency guidewire or coronary guidewire was used for perforation of pulmonary valve. Balloon dilation was subsequently performed.

Results. Of the 55 patients, perforation of atretic pulmonary valve was attempted with a radiofrequency guidewire (PA 120) in 49 and with a guidewire in 6. The procedure was successful in 49 patients: 4 with a guidewire and 45 with a radiofrequency guidewire, but failed in 6. Three of the 6 failure cases were complicated with pericardial effusion. Of the 49 patients, the mean right ventricular systolic pressure decreased from 115 ± 22 to 54 ± 12 mmHg following valvuloplasty. ($P < 0.01$) A stent was implanted to maintain patency of ductus in 4. Nine underwent a right ventricular outflow tract (RVOT) patch with or without a shunt and 2 underwent a shunt because of persisted cyanosis requiring PGE1 and/or development of infundibular stenosis. Ligation of ductus was performed in 4. There were 4 early mortalities: 1 sepsis & 3 heart failure. There was two late mortalities: severe right heart failure in 1 and renal failure in another. The mean tricuspid valve Z score was significantly lower in patients with RVOT patch than those without. (-1.99 ± 0.34 vs. -0.84 ± 0.85 , $P < 0.05$) In the most recent follow-up, 41 patients achieved biventricular circulation and 2 had 1.5-ventricle circulation.

Conclusions. Transcatheter pulmonary valvotomy can be a definitive treatment in selected patients with PA-IVS. Those with a significantly hypoplastic right ventricle may require RVOT patch despite a successful pulmonary valvotomy.