REFERRAL RATE FOR PULMONARY VALVE REPLACEMENT IN ADULTS WITH PULMONARY STENOSIS WHO HAVE UNDERGONE CHILDHOOD VALVOTOMY: A SINGLE ADULT CONGENITAL HEART CENTER’S EXPERIENCE

ACC Moderated Poster Contributions
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Background: Pulmonary valve stenosis (PS) represents 6-10% of all congenital heart disease. Surgical valvotomy in children with significant PS is effective in relieving valvar obstruction but results in chronic pulmonary regurgitation. Although the Second Natural History Study from 1993 showed only a 4% incidence of reoperation in patients with PS, little is known about the long-term sequela of chronic pulmonary regurgitation and need for pulmonary valve replacement (PVR) later in life.

Methods: Sixty-four adults who underwent surgical valvotomy for severe PS were referred to our Adult Congenital Heart Center between 2001 and 2011. Nine patients were excluded with either sub- or supra-valvar PS or prior PVR. We reviewed the medical records of 55 subjects (20 males), age 38.3 +/- 9.1 years, who had prior pulmonary valvotomy at a mean age of 3.6 +/- 3.9 years. Subjects referred for PVR were compared to those followed with a non-operative strategy.

Results: Among the 55 subjects, 21 (38%) were referred for PVR in favor of clinical observation. Average time from initial consultation to referral for PVR was 0.79 +/- 0.91 years. Compared to the clinical observation group, those referred for PVR had more moderate-to-severe or severe pulmonary valve regurgitation (81% vs. 21%; p = <0.001), reduced right ventricular (RV) function by MRI (EF 44.9% vs. 52.1%; p = 0.016), significant tricuspid regurgitation by echocardiography (moderate tricuspid regurgitation or higher; 38% vs. 12%; p = 0.04), and higher NYHA functional class (class II or greater; 67% vs. 29%; p = 0.0068 ). There was a trend towards larger RV end diastolic volume indices by MRI (169 cc/m^2 vs. 150 cc/m^2; p = 0.42) and lower peak pulmonary stenosis gradient by echocardiography (14.3 mm Hg vs 21.4 mm Hg; p = 0.11) in the surgery group.

Conclusions: Although surgical valvotomy for PS carries an excellent prognosis, a significant proportion were referred for PVR soon after initial evaluation. The high rate of referral for PVR during the fourth decade of life highlights the need for ongoing surveillance of patients who underwent childhood treatment for PS.