Echocardiographic follow-up in congenital valvular aortic stenosis during childhood

D.J. Ten Harkel 1; M. Berkhout 1; M. Witsenburg 2; W. Hop 2; W.A. Helbing 2
1Erasmus MC-Sophia, Pediatrics Dept., Rotterdam, Netherlands; 2Erasmus MC, Epidemiology Dept., Rotterdam, Netherlands

Introduction: In the present era patients with congenital valvular aortic stenosis are diagnosed and are being followed primarily by echocardiography. Heart catheterization is usually reserved for intervention by balloon valvuloplasty. Optimal timing to intervene remains, however, unclear. Since the rate of progression of aortic stenosis as assessed by echocardiography, and possible risk factors are largely unknown, we performed a retrospective review of our patients with congenital aortic valvular stenosis.

Methods: Retrospective review of all echocardiograms and clinical variables of patients with isolated congenital valvular aortic stenosis were obtained. Echocardiograms consisted of left ventricular mass and peak systolic aortic flow. Progression of the peak systolic aortic flow over time was established. Events were defined as intervention (surgical or balloon valvuloplasty) or death. Event-free survival was significantly longer in the older patients and those in 147 (60%). Interventions were performed in 73 patients (30%). The intervention-free survival was significantly longer in the patients with isolated congenital aortic valvular stenosis.

Results: The population consisted of 245 patients: 30 neonates (<1 month of age), 56 infants (<1 year of age) and 159 patients with an age above 1 year. Severe stenosis (>4 m/sec) was present in 42 patients (17%), moderate stenosis (2-4 m/sec) in 56 patients (23%), and mild stenosis (<2 m/sec) in 147 (60%). Interventions were performed in 73 patients (30%). The intervention-free survival was significantly longer in the older patients and those with only mild stenosis. After the first intervention 50% of the patients needed a second intervention within 10 years of follow-up. Five-year overall survival was significantly influenced by age at diagnosis. Neonates had a 27% mortality, while this figure was less than 2% in patients beyond the neonatal age. Sudden unexpected death did not occur in the present study. In 204 patients at least 2 echocardiograms before any intervention were available to estimate progression. During a follow-up period of 9-0.5±2 years there was an annual increase in peak systolic aortic flow of 0.04±0.006 m/sec. Left ventricular mass increased from 70±27 g/m2 to 79±27 g/m2.

Conclusions: During a 10-year follow-up period there was only limited progression in peak systolic aortic flow. Sudden unexpected cardiac death was not present, and the prognosis was excellent despite a significant mortality during neonatal age. Most patients needed reinterventions after a first intervention.

894 Effect of endovascular stenting of aortic coarctation on biventricular function in adults

Y. Lam 1; L.C. Kum 1; W. Li 2; M.Y. Henein 2; M. Mullen 2
1The Prince of Wales Hospital, Medicine and Therapeutics Dept., Hong Kong Sar, China, People’s Republic of; 2Royal Brompton Hospital, Echocardiography Dept., London, United Kingdom

Objective: We sought to investigate the effect of endovascular stenting of aortic coarctation on biventricular function.

Methods: We prospectively studied 21 patients (age 30±10 years) before and 14±2 months after coarctation stenting from year 2003-2005. Measurements of biventricular function and blood pressures were made. The post-stenting results were compared with pre-stenting values (group 1), with 22 age and sex-matched post-surgical repair patients (group 2) and 30 normal controls (group 3).

Results: The peak systolic gradient across coarctation site fell from 55±15 mm Hg to 18±8 mm Hg after stenting (p<0.001). LV mass decreased (257.6±117.9 g to 212.2±70.9 g; p<0.05), LV long axis function improved (LV lateral and septal systolic velocities: LSm, 6.5±1.4 cm/s to 7.9±1.7 cm/s; Sm, 5.8±1.2 cm/s to 7.3±1.6 cm/s; TDI septal early diastolic velocities: SSm, 7.6±1.5 cm/s to 7.8±1.9 cm/s; ep E/Em ratio: 14.8±5.3 to 11.8±3.9; p<0.05 in all) and blood pressure improved after stenting (p<0.05). No significant difference in LVEF conventional LV diastolic measurements (E, A, E/A ratio, IVRT and DT) or LV global indices measured by either MRI or I-123 Tc was found. Post-stenting patients had preserved RV long axis systolic function compared to postsurgical repair patients (TSm 12.0±1.4 cm/s vs 10.7±2.1 cm/s, p<0.05). Both post-stenting and postsurgical repair patients had poorer long axis function compared to normal controls (p<0.01 for all).

Conclusions: Endovascular stenting of aortic coarctation resulted in better blood pressure control, regression of LV mass and improvement in LV long axis function that may provide insight into long-term outcome of the stented patients. We support aortic stenting in anatomically suitable defects given additional potential benefit on LV function preservation. The clinical significance of subclinical myocardial dysfunction in patients with stented or re-paired coarctation warrants further studies.

895 Restrictive right ventricular physiology complicating pulmonary vascular stenosis

Y. Lam 1; L.C. Kum 1; C.P. Chan 1; M.Y. Henein 2; W. Li 2
1Prince of Wales Hospital, Medicine and Therapeutics Dept., Hong Kong Sar, China, People’s Republic of; 2Royal Brompton Hospital, Echocardiography Dept., London, United Kingdom

Objective: To assess diastolic restrictive right ventricular (RV) physiology (antegrade pulmonary artery flow in late diastole) in patients with moderate to severe isolated pulmonary valvular stenosis (PS).

Background: The clinical relevance of restrictive RV physiology in PS patients remains understudied.

Eur J Echocardiography Abstracts Supplement, December 2006