Valvular Heart Disease

SURGICAL INTERVENTION IN MUCOPOLYSACCHARIDOSES-RELATED VALVULAR HEART DISEASE

Moderated Poster Contributions
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Background: The mucopolysaccharidoses (MPS) are inherited lysosomal storage disorders that result in progressive systemic deposition of glycosaminoglycans. Cardiac involvement is common and usually involves the left-sided valves. We reviewed our experience with valve surgery in these patients (pt).

Methods: The records of pt with MPS undergoing valve surgery between 1970 and 2012 were reviewed.

Results: Ten pt aged 21-43 yrs (4 men), 3 pt reported previously, underwent 13 valve operations. Median age at first surgery was 36 (+7) yrs. Median BSA was 1.51 (+ 0.4) square meters. Maroteaux-Lamy was present in 7 pt, Morquio in 3 pt. The most common preoperative finding was severe aortic stenosis in 62% (n=8) or severe mitral stenosis in 46% (n=6). Severe mitral regurgitation was present in two pt, severe aortic regurgitation and subaortic membrane in one pt each. Aortic and mitral valve replacement (AVR & MVR) was performed in 46% (n=6), isolated AVR in 38% (n=5) and isolated tricuspid valve (TV) replacement in one pt. Median AVR size was 19 (+3) mm and MVR size was 22 (+2) mm. Aortic root enlargement (n=5), left ventricular septal myectomy (n=1) or both (n=1) was necessary to relieve left ventricular outflow tract (LVOT) obstruction. Difficult endotracheal intubation was noted in 69% (n=9), with video laryngoscopy and/or fiberoptic intubation used in 54% (n=7). There was one early postoperative death in a pt that presented with Class IV heart failure and underwent redo AVR, MVR and TV repair. There were three late deaths, two from respiratory failure. Other systemic features included joint disease (80%), corneal clouding (60%), sleep-disordered breathing (50%), spinal stenosis (40%) and subglottic stenosis (20%).

Conclusions: Adult pt with surgical valvular disease due to MPS most commonly present with aortic and mitral valve involvement. Small prostheses are often necessary due to small annulus size and either septal myectomy or aortic root enlargement is frequently required to relieve LVOT obstruction. A multidisciplinary approach is necessary to optimize perioperative management. Timing of valve surgery should include assessment of systemic features that may limit mobility and mask cardiac symptoms.