Surgery for Congenital Heart Disease

Surgical treatment of congenital mitral valve disease: Midterm results of a repair-oriented policy

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Objective: Management of congenital mitral valve disease is challenging because of a wide morphologic spectrum, frequent associated lesions, and small patient size. We evaluated the results of a repair-oriented policy.

Methods: All consecutive patients with congenital mitral valve disease who underwent surgery between 1996 and 2006 were studied retrospectively. Patients with atrioventricular canal, atrioventricular discordance, or ischemic regurgitation were excluded.

Results: During this period, 71 children (median age 2.9 years, range 3 days–20.8 years) underwent surgery. All but 1 underwent primary mitral valve repair. Twenty-two (30%) were younger than 12 months. Associated cardiac lesions were present in 45 children (63%) and were addressed concurrently in 35; previous cardiac procedures had been performed in 17 patients (24%). Mitral incompetence was predominant in 60 (85%) and stenosis in 11 (15%). During a median follow-up of 47.8 months (range 2–120 months), 14 patients underwent 17 mitral reinterventions: 14 repairs and 3 replacements. After 60 months, overall survival was 94% ± 2.8%; freedoms from reoperation and prosthesis implantation were 76% ± 5.6% and 94% ± 3.6%, respectively. There were 4 deaths, and all survivors remain in New York Heart Association class I or II with moderate (6 patients) or less mitral dysfunction.

Conclusion: Surgical repair of the congenital mitral valve can be successfully performed with low mortality, satisfactory valvular function at midterm follow-up, and acceptable reoperation rate while obviating risks associated with valvular prostheses. Suboptimal primary repair was significant predictor for reoperation but re-repair was often successful.

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In the past decade, the surgical approach to congenital mitral valve disease has significantly evolved as successive midterm and long-term series have been reported.[1-3] Pediatric patients can derive the same benefits from mitral valve repair as adults with regard to preservation of valvular tissue, subvalvular apparatus, and ventricular geometry, leading to optimal valve and ventricular function. Furthermore, avoidance of mechanical prostheses is especially desirable in young children, in whom annular growth should be fostered and who may have little physical space for the prosthesis in the heart.

After pediatric mitral valve replacement, mismatch between native annulus and mitral prosthesis has been shown to be a risk factor for both early and late death.[4-6] The probability of mitral valve prosthesis re-replacement was demonstrated to be

Abbreviations and Acronyms
PAP = pulmonary arterial pressure
PTFE = polytetrafluoroethylene

inversely related to the absolute size of the prosthesis initially implanted.[7] Finally, the cumulative risk generated by a lifelong commitment to anticoagulation should be avoided whenever possible.

Diagnostic tools are evolving rapidly and allow superior anatomic diagnosis and monitoring of the surgical repair. The range of surgical techniques modified from adult surgery into pediatric practice or specially developed for pediatric patients is large and allows tailoring of the surgical techniques to anatomic requirements.

Congenital mitral valve disease is rare and frequently associated with other cardiac malformations. Because it is usually complex, intervention is ideally postponed to allow time for annular growth and tissue maturity.[8] This is usually considered to be safe, because depressed systolic ventricular function has been shown to recover after successful mitral valve surgery in pediatric patients.[9,10] Severe congestive cardiac failure refractory to maximal medical therapy, however, can result in surgery being undertaken in the first months of life.

In 1996, our unit implemented a strategy whereby mitral valve replacement if necessary is planned when the mitral valve annulus diameter allows it to be done with low early or long-term risk. Mitral valve repair in this context may be considered a palliative procedure designed to allow time for growth. This study reviews our 10-year experience in children undergoing this repair-oriented mitral valve strategy.

Materials and Methods

From January 1996 to March 2006, a total of 71 patients living in Australia underwent surgery at our institution for congenital mitral valve disease. Data were obtained from institutional databases, supplemented by medical records from referring cardiologists or general practitioners, from January 2006 to March 2006. This study was approved by our institutional human ethics committee. Median age at operation was 2.9 years (range 3 days–20.8 years); 22 patients (30%) were younger than 12 months. Median weight at operation was 15.0 kg (range 3.0–99.4 kg). Six patients were dependent on mechanical ventilation before the operation. Primary mitral valve repair was possible in all but 1 case. Seventeen patients had undergone 19 nonmitral cardiac interventions before mitral surgery. Associated cardiac lesions were present in 45 children (63%) and addressed concurrently in 35 when not previously treated.

Patients with partial or complete atrioventricular canal, atrioventricular discordance, or ischemic mitral regurgitation from anomalous origin of the left coronary artery from the pulmonary artery were excluded. Patients with idiopathic dilated cardiomyopathy and functional mitral regurgitation were also excluded. Children with cleft mitral valve were included, as were 3 patients undergoing staged univentricular pathways with normally sized left ventricle, hypoplastic right ventricle, and congenital mitral valve dysplasia. Genetic, chromosomal, or systemic syndromes were present in 18 patients (25%); these included Marfan syndrome, Shone syndrome, Barlow's disease, William syndrome, and Down syndrome.

Timing of Surgery

Indications for surgery varied according to the etiology and anatomy, the age of the patient, the size of the mitral valve annulus, and clinical status. Neonates and infants with severe mitral valve disease were only considered for operation if they had severe symptoms. In patients with an annulus larger than the smaller valve prostheses available (20 to 21 mm with cuff), no symptoms were required if the valve could be repaired simply without annuloplasty (cleft mitral valve); for more complex valves, symptoms were usually present at the time of surgery. Surgical indications for patients with predominant mitral stenosis were dictated by symptoms only. No specific threshold figure for either pulmonary arterial pressure (PAP) or transmitral gradient triggered a surgical indication if few or no symptoms were present.

Preoperative Evaluation

Preoperative valve function was assessed by transthoracic echocardiography according to the American Society of Echocardiography guidelines.[11] Mitral incompetence was severe in 37 patients, moderate in 19, and mild in 3, with associated severe stenosis in 1, moderate stenosis in 1, and mild stenosis in 4. In 1 patient, the cleft mitral leaflet was discovered during ventricular septal defect repair and closed concurrently. Mitral stenosis was severe (mean gradient $>$15 mm Hg) in 7 patients and moderate (mean gradient 10–15 mm Hg) in 4, with coexistent moderate mitral incompetence in 2 and mild incompetence in 2. All patients with predominant mitral stenosis had pulmonary hypertension; mean peak systolic PAP was 64 mm Hg (range 45–100 mm Hg).

Intraoperative echocardiography was used to assess mitral valve function before and after repair. In this era, transesophageal echocardiography was used in 62 cases (87%), with epicardial echocardiography used in the remainder. No patients underwent diagnostic catheter study.

Mitral Anatomic and Functional Classification

We classify the mitral valves according to three criteria: hemodynamic, functional, and anatomic. This is also in accordance with a standardized classification.[12] Hemodynamically, the valves may be considered to be predominantly regurgitant or stenotic. Mitral incompetence was predominant in 60 valves (85%), and stenosis predominated in 11 (15%). The functional classification was according to the Carpentier classification,[13] with normal (type I), enhanced (type II), and restricted leaflet (type III) motion. From the anatomic point of view, we divided the congenital mitral valve anomalies into those with nondysplastic leaflets and those with dysplastic leaflets. Nondysplastic leaflet anatomy can occur with annular dilation, with or without elongation of the chordae or the papillary muscle. Such anomalies are usually found with significant volume loading of the left ventricle (large ventricular septal defect or large patent ductus arteriosus). In such cases, the papillary muscle may have a beige, ischemic appearance. A