Supraannular Mitral Valve Replacement in A Child with Congenital Mitral Stenosis

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Introduction

Surgical management of congenital malformation of the mitral valve in the paediatric age group remains a therapeutic challenge. Reconstructive techniques in infants and children have been evolved more slowly than in adults due to large variability of mitral disease and the uncertain effect that growth will have on the mitral apparatus, and replacement is reserved for patients with severe valvular pathologies not amenable to repair. Valve repair is indicated whenever feasible and should be considered before occurrence of pulmonary hypertension (1). The choice of the ideal valve substitute remains controversial. Experience with mitral valve replacement in the paediatric age group is limited, particularly in infants and children within the first few years of life. This selected group of patients poses unique problems associated with limited valve selection, obstructive hemodynamics of small prostheses, rapid patient growth, and difficulty with anticoagulation (1,2).

Case Report

A 4-year-old girl was followed-up for two years at our institution because of a moderate congenital mitral stenosis. She was hospitalised several times for recurrent episodes of pneumonia. Auscultation of the heart revealed a mild apical diastolic rumble with a loud P2. Pulmonary auscultation exposed some minimal rales in the posterobasal portion of each hemithorax. Laboratory findings were in normal ranges. Cardiac X-ray revealed moderate cardiac enlargement with a prominent left atrial chamber. There was a normal sinus rhythm and slightly rightward axis on the electrocardiogram. Transthoracic echocardiographic (TTE) examination showed; a large left atrium (3.9 cm diameter), minimal tricuspid valve insufficiency, good cardiac contractions, narrow mitral annulus (15 mm diameter), restricted mitral opening with a turbulent jet through the left ventricle at the end of each diastole, 40 mm Hg diastolic gradient between left atrium and left ventricle, 6 mm wide mitral orifice, thick and shortened papillary muscles, and more than 50 mm Hg diastolic pulmonary artery pressure. With these findings the patient was taken to surgical intervention.

Following median sternotomy and aortobicaval cannulation, cardiopulmonary bypass was assessed. Systemic hypothermia (28 ˚C) was used and myocardial protection was provided with the intermittent antegrade cold blood cardioplegia and topical hypothermia. The mitral valve was approached via interatrial septum. No associated cardiovascular or extracardiac abnormalities were detected. The prominent presentation was a typical mitral stenosis with short chordae tendineae, obliteration of interchordal spaces and reduction of interpapillary distance. Commissurotomy was performed along the anterolateral and posteromedial commissures. Intervalvular fusions were partially freed by gentle blunt dissection of the fibrous tissue. Intraoperative valve function was assessed by transosephageal echocardiography (TEE), which revealed residual gradient of 10 mm Hg between left atrium and left ventricle. In addition, a 9 mm mitral orifice, together with a mild-moderate mitral insufficiency was also found.

Patient quickly recovered and was extubated on the 7th postoperative hour with a stable hemodynamic status. However, postoperative TTE examination showed, moderate-severe mitral insufficiency, 27 mm Hg peak gradient between left atrium and left ventricle, 65-70 mm Hg systolic pulmonary artery pressure, and a very large left atrium. Patient gradually deteriorated despite inotropic support and on the 15th postoperative day the patient was intuba-

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ted and she was taken to the operating room and a supra-annular mitral valve replacement with #23 St.Jude (St Jude Medical, Inc.) prosthetic valve was performed using standard everting, horizontal, pledgetted mattress sutures. Patient did very well after the operation and was back home 10 days after the reintervention (Fig 1). The patient was followed-up by echocardiographic examination periodically in every six months. Last control TTE on the 18th month after discharge revealed; good cardiac contractions, no pericardial effusion, right ventricular pressure of 30 mm Hg, functioning prosthetic valve at the mitral position with no paravalvular leakage, and 10 mm Hg gradient between left atrium and left ventricle (Fig 2). Her functional status on the last examination was NYHA class I.

Discussion

The spectrum of congenital mitral valve abnormalities ranges from repairable cleft leaflets to restrictive and challenging lesions of isolated mitral stenosis. Most of the patients with isolated congenital mitral stenosis have severe symptoms in early childhood (3,4). Therefore these patients need early medical or surgical management depending on the clinical course of the disease. When symptoms are mild or even moderate, operation is delayed in the hope, that when it becomes necessary and if valve replacement is required, an adult-sized device can be used (3). A recent option to surgical management is balloon dilatation, which is appropriate even for infant patients having mild mitral stenosis with unsuccessful medical management (4,5). Because of the significant short and long-term problems with prosthetic valves in children, attention should be paid to mitral repair techniques for these valves. However, reoperation after chest closure and transfer to the ICU after failed mitral valve repair according to gradually deteriorating hemodynamic findings have been reported in children, despite intraoperative TEE examination (6). Although further refinement of valvuloplasty methods decreases the need for valve replacement, for patients with severe congenital deformity, particularly in the mitral position, there will still be a small need for infant valve replacement. Especially, cases with mitral stenosis remain extremely problematic in younger children compared to mitral insufficiency (7).

The choice of prosthetic valve type remains controversial. Bioprosthetic valves were predominantly used to avoid anticoagulation in young children. However, high probability of early calcific degeneration and subsequent need for early replacement of the bioprosthetic valves have been demonstrated (2,8). Mechanical prostheses appear superior to bioprostheses particularly in children despite the requirement of anticoagulation with warfarin derivatives, which subjects active, growing children to the risks of bleeding (2,5,9). The results of series demonstrate that mitral valve replacement is associated with a substantial operative and late mortality in infants and young children within the first 5 years of life. The serious underlying condition of these patients, however, justifies this approach when valve-conserving procedures are unsuccessful (4,5,9).

Children requiring mitral valve replacement, whose native annulus is too small to accept the smallest prosthesis available, present a difficult management problem. An alternative choice is replacement of the mitral valve with a prosthesis positioned above the native annulus entirely within the left atrium to pro-

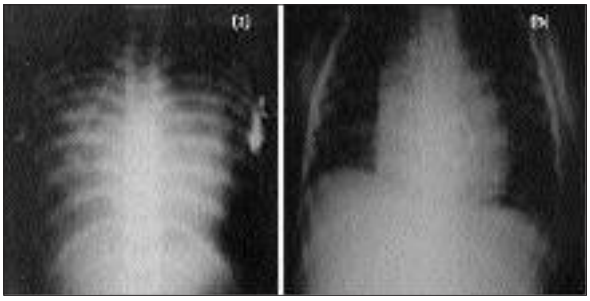

Figure 1.a) Chest X-Ray of the patient before the second operation, b) Chest X-Ray of the patient after the mitral valve replacement.

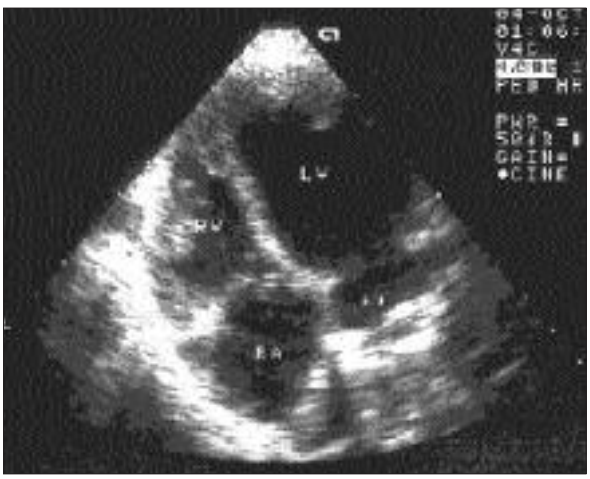

Figure 2. The echocardiographic examination shows the supraannular placed mitral valve prosthesis.

vide maximal prosthetic valve area. It is also reported that an aortic prosthesis can be successfully replaced in reverse position into the supraannular site of the left atrium (10). This supraannular positioning of the synthetic prostheses appears to be associated with satisfactory intermediate-term results and freedom from reintervention (4,10).

In patients with congenital mitral valve disease, reconstructive surgery is the primary goal. When necessary, valve replacement can be performed safely and provide extended palliation for many patients. The operative mortality and hemodynamic results of valve replacement are initially satisfactory; however, the risk of thromboembolism and endocarditis, the requirement for long-term anticoagulation, and the need for multiple valve replacements are of concern with the growing child (4,5,7,9).

Improvements in surgical techniques have reduced the operative mortality of mitral valve surgery even in infant patients, although ongoing morbidity and mortality continue to limit the usefulness of surgical management. Supraannular mitral valve replacement is a preferable approach to replace an appropriate prosthesis in children with severe congenital mitral stenosis and small mitral annulus.

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