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Surgical results of mitral valve repair for congenital mitral valve stenosis in paediatric patients

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Abstract

OBJECTIVES: Mitral valve (MV) repairs have been performed in paediatric patients with congenital MV stenosis. However, congenital MV stenosis lesions are a heterogeneous group of lesions, and their repair remains challenging.

METHODS: From March 1999 to September 2014, MV repair was performed in 22 patients with congenital MV stenosis. The median age was 10.3 months (ranging from 22 days to 9.1 years), and the mean body weight was 7.9 ± 4.0 kg at the time of the operation. Multiple-level left-side heart obstructions were present in 9 (45%) patients.

RESULTS: The main aetiology of the mitral stenosis was a supravulvar mitral ring in 8 patients, valvular stenosis in 4 patients, a parachute deformity of the papillary muscles in 4 patients and other abnormal papillary muscles in 6 patients. The mean MV pressure gradient improved from 10.4 ± 3.9 mmHg to 3.4 ± 1.7 mmHg after MV repair (n = 18, P < 0.0001). The mean follow-up duration was 6.7 ± 5.4 years. One patient died postoperatively due to septic shock. Four patients required a second operation (2 patients for mitral stenosis, 1 patient for left ventricular outflow tract obstruction and mitral stenosis and 1 patient for mitral regurgitation). Among them, 2 patients died: 1 patient died due to cardiopulmonary bypass weaning failure and another patient died due to multiple cerebral infarcts. At the last follow-up, the mean MV pressure gradient was 4.5 ± 3.1 mmHg for all patients who did not have reoperation, and moderate or greater mitral insufficiency was detected in 3 patients. At 10 years, the survival rate was 85.9 ± 7.6%, and the freedom from reoperation rate was 77.5 ± 10.1%. In the log-rank test, MV repair in the neonate was associated with mortality (P = 0.010), and presentation of mitral insufficiency was associated with reoperation (P = 0.003).

CONCLUSIONS: MV repair in paediatric patients with congenital mitral stenosis showed acceptable results. The follow-up echocardiogram also revealed satisfactory results. Close follow-up is necessary to detect the development of postoperative mitral stenosis or regurgitation.

Keywords: Congenital heart disease • Mitral stenosis • Valve repair

INTRODUCTION

Congenital mitral valve (MV) stenosis (CMS) is rare, and its surgical management is challenging because of the wide spectrum of pathology and the high incidence of other co-existing left-sided heart obstructions, which affect clinical severity [1, 2]. In more common forms, the anatomic defect encompasses the valve leaflets and the subvalvular apparatus. In less common forms, there is accessory tissue that forms at or above the valve annulus, causing the obstruction or tethering of the valve annulus and preventing growth [3].

Surgical treatment of CMS has become a major therapeutic choice, although the methods of surgical treatment are limited compared with those for acquired mitral stenosis. Surgical options include MV repair and MV replacement with a prosthetic valve. However, in a young population, the use of prosthetic valves is less desirable, because the mismatch between the native annulus and the mitral prosthesis has been shown to be a risk

factor for both early and late mortality; the valve may also require repeated replacement as the child grows [4]. When possible, MV repair is desirable and should be attempted, because it conserves the subvalvular apparatus and ventricular geometry, thereby preserving left ventricular function. Preserved left ventricular function leads to long-term survival benefits [5].

There are a few studies on the efficacy of MV repair for CMS [1, 6-9]. The aim of this study was to analyse our experience with MV repair in children with CMS.

MATERIALS AND METHODS

Patients

From March 1999 to September 2014, a total of 22 patients with CMS underwent surgical intervention for congenital mitral stenosis

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The materials + methods part of our research is the part I'm most unfamiliar with. Since Traci has been working on this mostly

explains why research is relevant

Table 1: Patient characteristics (n = 22)

| Characteristics | Value |
|-------------------------------------|-----------------------------|
| Demographics | |
| Male:female, n | 9:13 |
| Age at operation, months (median) | 10.3 (22 days to 9.1 years) |
| Body weight at operation, kg (mean) | 7.9 + 4.0 |
| Neonates | 4 |
| Infants | 8 |
| Children | 10 |
| Associated lesions, n | |
| Aortic stenosis | 3 |
| Coarctation of the aorta | 8 |
| Ventricular septal defect | 13 |
| Atrial septal defect | 6 |
| Patent ductus arteriosus | 7 |
| Pulmonary stenosis | 1 |
| Tricuspid regurgitation | 2 |

at Seoul National University Children's Hospital (Table 1). Patients presenting with atrioventricular septal defects or functional single ventricles were excluded.

Indications for surgical treatment were not standardized and varied according to the pathology of the MV lesion, patient age and clinical status. In general, patients were considered candidates for surgical treatment upon presentation with standard indications, such as severe symptoms (e.g. heart failure and failure to thrive) or evidence of significant pulmonary hypertension. MV repair was performed concurrently with the repair of associated cardiac anomalies at the surgeon's discretion with the preferred technique. The retrospective database and chart review was approved by the Institutional Review Board at Seoul National University Children's Hospital, which waived patient consent because of the retrospective nature of the data analysis.

Preoperative evaluation and operation

The quantitative variables that were analysed included the MV annular dimension and z-score as well as the MV inflow Doppler mean gradients (Table 2). Mitral stenosis was quantified by the measurement of MV inflow Doppler mean gradients (mmHg) and was graded as mild (<5 mmHg), moderate (5–10 mmHg) or severe (>10 mmHg). A more than moderate mitral stenosis was considered to be significant and was observed in all 22 patients: the mean value was 10.4 ± 3.9 mmHg (moderate in 13 patients and severe in 9 patients). Peak tricuspid regurgitation velocity, signs of interventricular septal flattening and dilatation of the inferior vena cava or right atrium were used to assess the probability of pulmonary hypertension. There were 12 patients who had evidence of significant pulmonary hypertension from preoperative echocardiographic evaluation (peak tricuspid regurgitation velocity >3.4 m/s in 6 patients, 2.9 m/s < peak tricuspid regurgitation velocity <3.4 m/s with other echocardiographic signs in 6 patients) [10].

MV anatomic diagnoses were derived from the preoperative transthoracic echocardiography examination and direct intraoperative findings. Although most patients showed complex structural abnormalities in each of the valvular components (leaflets, chordae, papillary muscles and the supra-annular apparatus), the classification method described by Carpentier *et al.* was used [11].

Table 2: Mitral valve anatomy (n = 22)

| Anatomy | Value | Reoperation |
|--|-----------------|-------------|
| Mitral valve | | |
| Annulus diameter, mm | 11.6 + 4.4 | |
| Z-value | -1.6 + 2.0 | |
| | (-4.71 to 1.35) | |
| Mean mitral inflow pressure gradient, mmHg | 10.4 + 3.9 | |
| Insufficiency moderate or greater | 3 | |
| Type of congenital mitral stenosis | n | |
| Type A (normal papillary muscle) | 12 | |
| Supra-annular ring | 8 | 0 |
| Leaflet fusion (intra leaflet ring) | 4 | 2 |
| Type B (abnormal papillary muscle) | 10 | |
| Parachute deformity | 4 | 1 |
| Papillary muscle abnormality | 6 | 1 |

After the establishment of a standard bicaval cardiopulmonary bypass with moderate hypothermic and cardioplegic arrest, the MV was approached through the atrial septum in 18 patients and through a left atriotomy in 4 patients. In all cases, MV function was estimated by transoesophageal echocardiography during weaning from the cardiopulmonary bypass and just after the separation from bypass.

Statistical analysis

Preoperative and postoperative data were collected from the patients' medical records. The characteristics of the study population were expressed as frequencies, medians with ranges or means with standard deviations, as appropriate. For the comparison of the preoperative and postoperative MV inflow pressure gradient, the paired t-test analysis was used. Overall survival and freedom from MV reoperation were analysed with the Kaplan-Meier survival analysis with a log-rank test to make comparisons between factors. One patient, who had failed MV repair and underwent MV replacement during the same operation, was not involved in the analysis for freedom from reoperation rate and risk factor of reoperation. The SPSS version 21.0 software (SPSS, Inc., Chicago, IL, USA) was used for all statistical analyses, and a P-value <0.05 was considered statistically significant.

RESULTS

Patient characteristics

The patient demographics are summarized in Table 1. Twelve patients (55%) were younger than 12 months of age (4 neonates). Multiple left heart obstructions, including Shone's complex and its variants [12], were associated with CMS in 9 patients (41%). Mitral insufficiency (MI) was detected in 6 patients, of whom 3 cases were moderate or greater. Of all patients, 5 (23%) had undergone prior surgical interventions for co-existent obstructive lesions on the left ventricular outflow tract or had undergone repair of other congenital heart defects.

According to Carpentier *et al.*'s classification [11], Type A CMS was identified in 12 (55%) patients. In these 12 patients, the associated findings included 8 patients with a supra-annular ring and

we will include similar information. since this is a retrospective research on our materials + methods mostly in examining patient pool.

something we also excluded. we can see through this section of the research that

made by our cardiothoracic surgery team (statistician). this is something I have yet to become aware of or have seen in our research

analyze statistical results and tie them to our overall purpose and goal with our research

Table 3: Operative procedures

| | |
|--|----|
| Procedures for mitral valve, n | |
| Repair | 21 |
| Resection of supra-valvular mitral ring | 8 |
| Papillary muscle splitting | 7 |
| Chordal splitting/resection | 6 |
| Commissurotomy | 5 |
| Leaflet slicing | 1 |
| Replacement, unsuccessful repair | 1 |
| Concomitant procedure, n | |
| Coarctoplasty | 4 |
| Ventricular septal defect closure | 11 |
| Atrial septal defect closure | 5 |
| Patent ductus arteriosus division | 2 |
| Right ventricular outflow tract widening | 1 |
| Aortic valve repair | 3 |
| Subaortic stenosis relief | 2 |
| Tricuspid valve repair | 1 |

isolating the different congenital MV malformation and repair,

4 patients with fused leaflets. Additionally, Type B CMS was identified in 10 (45%) patients. In these 10 patients, the associated findings included 4 patients with parachute deformity variants and 6 patients with abnormal papillary muscles (hammock defect in 3 patients, fused papillary muscles in 2 patients and abnormal accessory muscle to posterior leaflet in 1 patient) (Table 2).

Surgical techniques and postoperative management

isolating repair type

The MV repair methods are presented in Table 3. A supra-valvular mitral ring was resected in 8 patients, of whom 4 did not need any other procedures. A single or dominant papillary muscle was split in 7 patients. Fused or shortened chordae were mobilized or resected in 6 patients. Commissurotomy was performed in 5 patients. There was 1 patient in whom the MV repair failed, and MV replacement was performed during the same operation.

The concomitant procedures included the following: ventricular septal defect closure in 11 patients, atrial septal defect closure in 4 patients, coarctoplasty in 4 patients, aortic valvuloplasty (aortic valvotomy, raphe release and leaflet slicing) in 3 patients, patent ductus arteriosus division in 2 patients, right ventricular outflow tract widening in 1 patient, subaortic fibrotic band wide resection in 1 patient and left ventricular outflow tract myectomy in 1 patient.

We used sedatives and milrinone for all patients after surgery and slowly weaned them off the ventilator. There were 4 patients who required inhaled nitric oxide therapy for pulmonary hypertension. In addition, 2 of them took sildenafil for 3 months after discharge.

most important!
Outcome and mortality

The mean length of follow-up was 6.7 ± 5.4 years. There was 1 hospital death in a 2.9-kg neonatal patient with a parachute MV, a single papillary muscle, severe pulmonary hypertension and preoperative renal failure. The patient underwent MV repair and atrial septal defect closure with fenestration. The renal failure did not improve, and the patient died 3 months after the operation from postoperative septic shock that was suspiciously related to a peritoneal catheter infection.

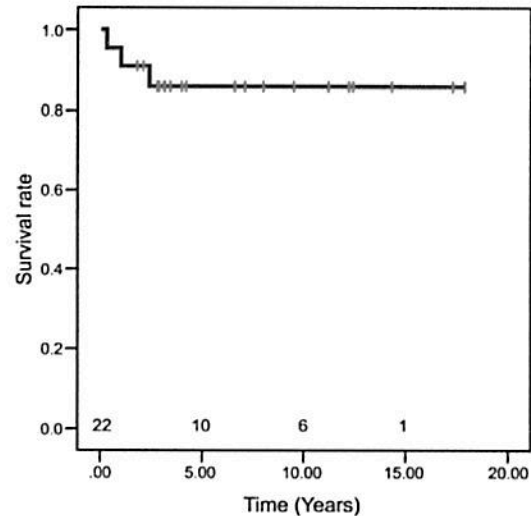


Figure 1: Kaplan-Meier graph presenting freedom from death.

review of patient death

Two late deaths occurred during the follow-up period. A 1-month-old boy who underwent commissurotomy for mitral stenosis developed progressive mitral stenosis and underwent MV replacement 1 year after the first operation. The patient died of multiple cerebral infarcts and acute respiratory distress syndrome after the repeat operation. A 9-year-old girl initially underwent MV repair (chordal fenestration and leaflet slicing), aortic valve repair and left ventricular myectomy for mitral stenosis and left ventricular outflow tract obstruction. Twenty-nine months after the operation, the patient underwent MV replacement and aortic valve replacement with the Konno procedure for mitral stenosis and left ventricular outflow tract obstruction. The patient could not be weaned off the cardiopulmonary bypass due to myocardial dysfunction and died 4 days after this operation due to multiple-organ failure despite extracorporeal membrane oxygenation support. The overall survival rate was $85.9 \pm 7.6\%$ at 10 years (Fig. 1). Age (neonate versus non-neonate) at the time of the MV repair ($P=0.010$) was significantly associated with mortality according to the log-rank test. However, anatomic lesions (supra-valvular ring versus complex valvular or subvalvular anomalies) ($P=0.194$), MI ($P=0.134$) and multiple left heart obstructions ($P=0.694$) were not associated with mortality according to the log-rank test (Fig. 2).

There were 4 reoperations. A 2-month-old girl who had undergone coarctoplasty as a neonate later underwent mitral papillary muscle splitting for CMS and aortic valvotomy for congenital aortic stenosis. Three years after the operation, the patient underwent a repeat papillary muscle splitting for progressive mitral stenosis. An 18-month-old boy who had undergone coarctoplasty as a neonate weighing 2.8 kg underwent MV repair for mitral stenosis and severe MV regurgitation. Seven months after the operation, the patient underwent MV replacement for progressive mitral regurgitation. Another 2 reoperations resulted in mortality as described above. All 3 MV replacements were performed with mechanical prostheses; among those, 2 patients had multiple left heart obstructive lesions as described above, and all 3 patients exhibited a restrictive small mitral valvar opening with leaflet thickening and short chords as well as the presentation of MI at the initial MV repair. At 10 years, the freedom from reoperation

common w/ pediatric MV repair

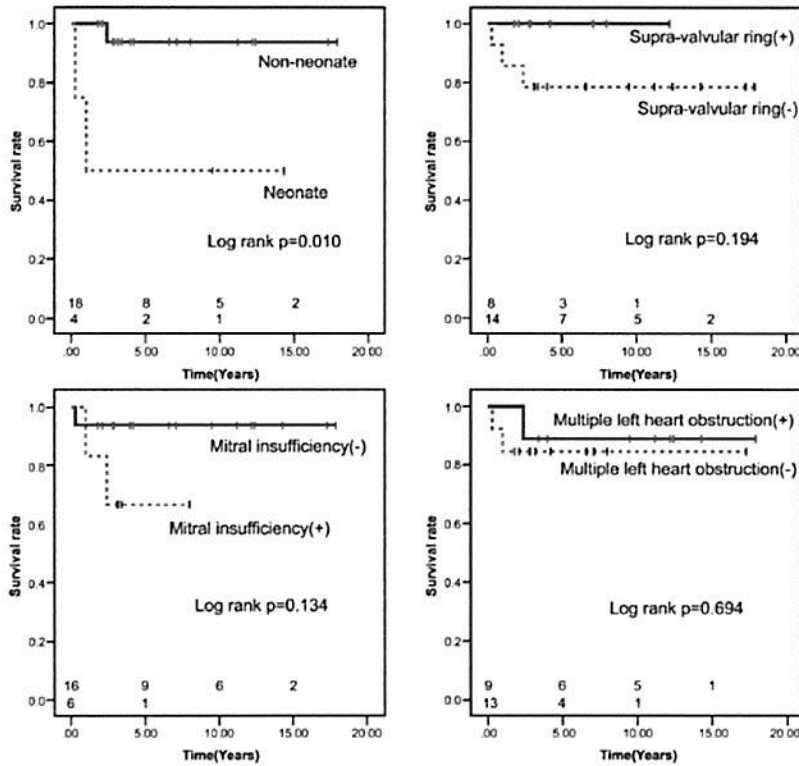


Figure 2: Risk factor analysis for mortality.

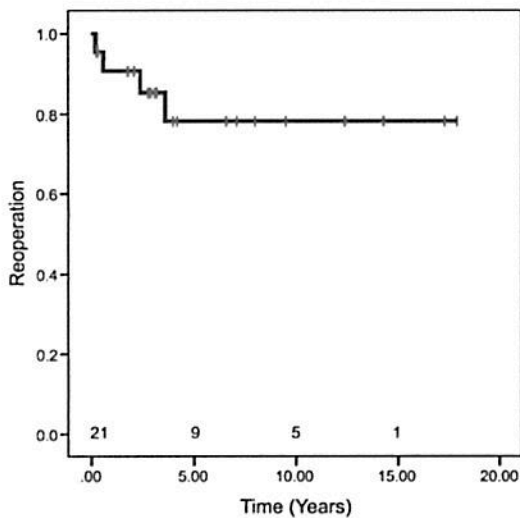


Figure 3: Kaplan-Meier graph presenting freedom from reoperation.

rate was $77.5 \pm 10.1\%$ (Fig. 3). MI ($P=0.003$) was significantly associated with reoperation according to the log-rank test. However, age (neonate versus non-neonate) at the time of the MV repair ($P=0.558$), anatomic lesions (supravulvular ring versus complex valvular or subvalvular anomalies) ($P=0.119$) and

multiple left heart obstructions ($P=0.307$) were not associated with reoperation according to the log-rank test (Fig. 4).

Postoperative echocardiography at discharge showed that the mean mitral inflow pressure gradient significantly improved from 10.4 ± 3.9 to 3.4 ± 1.7 mmHg ($n=18$, $P=0.001$). At the last follow-up, the mean mitral inflow pressure gradient was 4.5 ± 3.1 mmHg for all patients who did not have reoperation for MV (Fig. 5), and moderate or greater MI was detected in 3 patients.

DISCUSSION

ties back to introduction
 Congenital mitral stenosis is a rare and morphologically heterogeneous lesion that affects both the leaflets and the subvalvular apparatus of the MV. Many classifications have been proposed for CMS. Ruckman and Van Praagh [13] proposed a simple classification based on pathological findings in cadavers. This classification, while useful in comparing pathologic groups, did not clearly reflect the mechanisms responsible for the obstruction. More recently, Carpentier et al. [11] described a functional classification based on the location of the major lesion. We classified our patients according to this more recent classification.

The surgical methods for MV repair for CMS remain limited and challenging. Surgical options include MV repair and MV replacement with mechanical prostheses. Furthermore, an expandable cardiac valve implant for infants and children with CMS has recently been introduced [14]. MV replacement in paediatric patients

↓ I remember reading about this classification on one of my research assessments for my original work, may be something worth looking at again to better understand patient classification that may or may not be used in our research.

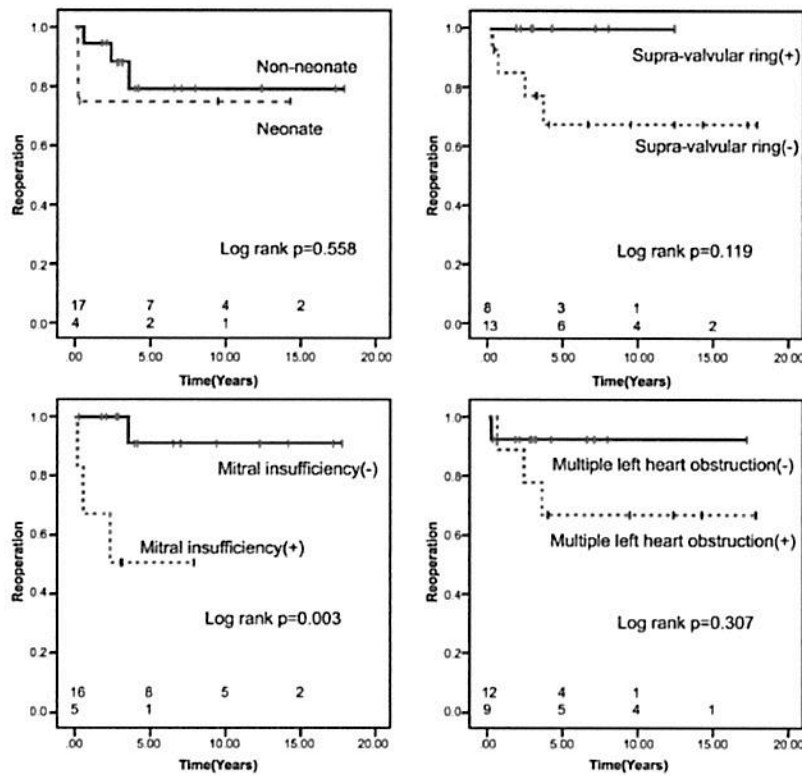


Figure 4: Risk factor analysis for reoperation.

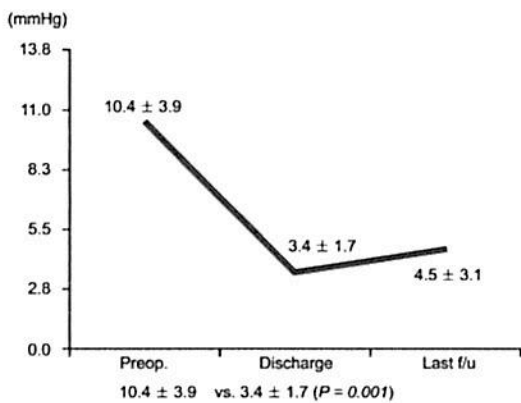


Figure 5: Serial change of mean mitral inflow pressure. preop.: preoperative; f/u: follow-up.

seems like the discussion part of our research will be very over my head since I have yet to learn surgical techniques

limited availability of adequately sized prostheses for small children, the need for anticoagulation therapy and the poor long-term results with a reported 10-year survival rate of 33-74% [4, 15-18]. Therefore, repair of the MV is the preferred approach. A number of approaches for the visualization of the MV have been described, including a vertical transatrial septal incision, a transverse atrial septal incision and a left atrial incision at the interatrial groove with or without division of the superior vena cava. In the 18 patients who underwent MV repair via the

trans-septal approach, we typically opened the boundary of the fossa ovalis with or without an extended incision into the septum secundum. Combining this approach with strategically placed traction sutures, the valve could be pulled into an optimal angle, and it usually ensured adequate visualization of the entire valve and subvalvular apparatus. A standard left atriotomy through the interatrial groove was performed in the other 4 patients without the division of the superior vena cava. Standard left atriotomy was usually useful in the patients who had an enlarged left atrium. A variety of repair techniques were employed based on the individual patient's valve pathology. Our surgical strategy for patients with CMS involved a variety of standard techniques that were tailored to the pathology of the individual patients, and we showed that the repair could be performed in this population with good rates of early survival and freedom from the need for replacement.

Multiple left-side obstructive lesions with CMS generally occur as a component of Shone's complex and remain a challenge. One complicating factor is that the degree of MS can be underestimated due to the co-existence of left ventricular outflow tract obstruction, which may mask the need for surgical intervention on the MV. Additionally, because of the presence of multilevel left ventricular inflow and outflow tract obstructive lesions, decision making regarding the timing of intervention, one-stage repair versus multiple-stage repair and single-ventricle repair versus biventricular repair is crucial, as failed valve repair can lead to pulmonary hypertension [1, 19]. In this study, the 4 patients who underwent reoperation after MV repair

CONGENITAL