Long-term results after mitral valve repair in children

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Abstract

Objective: We analysed the long-term results of mitral valve (MV) repair in children. Methods: We reviewed clinical records of 139 children (<18 years) who underwent MV repair between 1988 and 2007. Patients with atrioventricular septal defect, single ventricle or atrioventricular discordance were excluded. Median age was 2.3 years (2 months to 17.6 years), and 47 children (34%) were infants. Mitral regurgitation (MR) was predominant in 125 patients (90%), and 91 (73%) of these showed MR grade ≥3. Mitral stenosis (MS) was predominant in 14 patients (10%), and median mean pressure gradient across the MV was 9.0 mmHg (0—20 mmHg). Associated cardiac lesions were present in 111 patients (80%) and were addressed concurrently in 105 patients. Various surgical techniques were used according to the functional and pathologic findings of MV.

Results: There was no early death. Median follow-up was 8 years (2 months to 20 years, 78% complete). Twenty-six patients required 29 MV re-operations, and 11 of these required MV replacements. At 15 years, freedom from MV re-operation and MV replacement was 77% and 90%, respectively. Diagnosis of MS and MV status on discharge (MR grade ≥3 or MS gradient ≥10 mmHg) were significant risk factors for re-operation. There were three late deaths, and the overall survival was 97% at 15 years. Among 122 survivors with MR, 102 patients (84%) underwent echocardiography during follow-up. The degree of MR decreased significantly and only five patients showed MR grade 3. Among 14 survivors with MS, eight patients (57%) underwent echocardiography during follow-up. The degree of MS decreased significantly and median MS gradient was 2.8 mmHg (0—10 mmHg). All survivors remain in the NYHA class I or II. Conclusions: MV repair in children showed excellent survival, acceptable re-operation rate and satisfactory valve function at long-term follow-up. Residual MV dysfunction was a significant risk factor for re-operation, but re-repair was successful in more than half of the patients who underwent re-repair.

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1. Introduction

Mitral valve (MV) disease in children is rare and its surgical management is challenging because of the wide spectrum of pathology and a high incidence of co-existing cardiac anomalies. In dealing with the MV disease of children, surgeons have to choose between MV repair and MV replacement. When possible, MV repair is desirable and should be attempted because it conserves the subvalvar apparatus and ventricular geometry, preserving left ventricular function. Furthermore, MV replacement in paediatric patients poses significant problems because of the limited availability of adequate-sized prostheses, the need for anticoagulation and poor long-term survival [1—4]. Several groups have reported excellent results with MV repair in children [5—10]. The aim of this study was to analyse our 20-year experience of MV repair in children.

2. Materials and methods

2.1. Patients

Between January 1988 and December 2007, 139 children (aged <18 years) underwent MV repair for either congenital or acquired MV disease. Patients with atrioventricular septal defect, single ventricle or atrioventricular discordance were excluded. A retrospective study was performed, and all data were collected by reviewing medical records. The Institutional Review Board of our institution approved this study. Median age at operation was 2.3 years (2 months to 17.6 years), and 47 children (34%) were infants (aged <1 year). Median body weight was 10.9 kg (range: 3.7—73.0 kg). Genetic syndromes were
present in six patients (4.3%), and these included Marfan syndrome (three), Williams syndrome (two) and Down’s syndrome (one).

2.2. Pathology and severity of MV lesions

MV function was assessed by transthoracic echocardiography (TTE). Severity of mitral regurgitation (MR) was estimated by semi-quantitative grading (grades I—IV) according to the maximum length and width of the abnormal jet relative to the left atrium. Because several pathological findings co-existed in some patients, the predominant one was used to classify the lesion according to the Carpentier’s functional classification [11] (Table 1). Most of the patients in whom annulus dilatation was a predominant pathology for MR had associated ventricular septal defects (VSDs) or patent ductus arteriosus (PDA), and the annulus dilatation was secondary to volume-loaded left ventricle. Haemodynamically, MR was predominant in 125 patients (90%), and mitral stenosis (MS) was predominant in 14 patients (10%). Severity of MR was grade IV in 47 patients, grade III in 44, grade II in 26 and grade I in eight. MS was severe (mean gradient >10 mmHg) in four patients, moderate (mean gradient 5–10 mmHg) in eight and mild (mean gradient <5 mmHg) in two.

2.3. Associated cardiac lesions

Associated cardiac lesions were present in 111 (80%) patients (Table 2). Previous operations for associated cardiac lesions were performed in 12 (8.6%) patients. Concomitant repair of associated lesions was performed in 105 (76%) patients.

2.4. Indications for surgery

Indications for surgery varied according to the pathology of the MV lesions, patient’s age and clinical status. Because of the fragile nature of the MV tissue, neonates and young infants with severe MV disease underwent surgery only if they had severe symptoms (e.g., heart failure and failure to thrive). Otherwise, surgery was delayed to about 2—3 years of age. In older children, severe symptoms, increasing left ventricular dimension on serial echocardiography or evidence of rising pulmonary arterial pressure were considered as indications for surgery. For patients with MR due to annulus dilatation secondary to volume-loading lesions such as VSD, it is our policy to repair the MV only when MR was equal to or greater than grade 3. In patients with mild MR (34 patients, grade ≤2), MV repair was performed concurrently with repair of associated cardiac anomalies at the surgeon’s own discretion. Eight of these patients had cleft MV, which could be repaired simply.

2.5. Surgical techniques

MV repair was performed through a median sternotomy under cardiopulmonary bypass and moderate systemic hypothermia. Antegrade intermittent cold blood or crystalloid cardioplegia was used for myocardial protection. Access to the MV was gained either by a left atriotomy (81 patients) or trans-septally (58 patients). The annulus, leaflets and subvalvar apparatus were carefully evaluated with the aid of saline injection test to determine the precise anatomy of the lesion and to choose optimal method of repair. Various repair techniques were used according to the functional and pathologic findings of MV (Table 3).

Annulus dilatation producing central MR was repaired by various reduction annuloplasty techniques. In the earlier part of our experience, bilateral or unilateral Wooler–Kay type annuloplasty [12] was commonly used. In recent years, we used artificial ring annuloplasty for older children and suture plication (since 1995) or posterior annuloplasty using strip (since 1998) for younger children. Strip materials used for annuloplasty were autologous pericardium in 10 patients, Gore-Tex (W.L. Gore & Associate, Inc., Flagstaff, AZ, USA) in five and others in three. In some patients, the strip was interrupted at mid-portion for growth of the posterior
annulus. For very young patients with small annuli, we preferred posterior annuloplasty using suture plication. MV cleft was corrected by a direct suture technique, either completely or partially, according to the minimal acceptable age-dependent MV diameter to avoid iatrogenic MS. MR caused by leaflet prolapse was repaired by formation of artificial chordae using Gore-Tex sutures, shortening of the elongated chordae or chordae transfer technique. Since 2000, we favored the use of artificial chordae because of its simplicity. Edge-to-edge repair (Alfieri technique) was used in five small children (2—15 months). For patients with MS, papillary muscle splitting was most commonly used. Resection of supravalvar ring was performed in four patients. Since 1997, intra-operative trans-oesophageal echocardiography (TEE) was routinely performed to assess the adequacy of MV repair.

2.6. Follow-up

All patients underwent TTE before discharge from the hospital and were followed-up by our paediatric cardiologists. TTE was performed during follow-up when a change in clinical status was noted or at the attending cardiologist’s own discretion. Follow-up completeness was 78% and the median follow-up duration was 8 years (2 months to 20 years).

2.7. Statistical analysis

Continuous data were expressed as median (range). Follow-up completeness was measured by calculating modified Clark’s index as previously described [13]. Analysis of overall survival and freedom from MV re-operation or MV replacement were performed with the Kaplan—Meier method. Risk factors for MV re-operation were determined with Cox proportional hazards analysis. SPSS (SPSS Inc., Chicago, IL, USA) was used for all statistical analysis, and a p-value <0.05 was considered statistically significant.

3. Results

3.1. Survival

There was no early death. Three late deaths occurred during follow-up. A 15-year-old girl, who initially underwent Wooler—Kay annuloplasty for severe MR (annulus dilatation, anterior leaflet prolapse), underwent mechanical MV replacement due to residual MR. She died of cerebral haemorrhage associated with anticoagulation 16 years after the MV replacement. A 3-year-old girl who underwent Wooler—Kay annuloplasty for MR died of dilated cardiomyopathy caused by viral myocarditis 11 years after the operation. An 11-year-old girl with coarctation of the aorta, VSD and severe MR underwent patch coarctoplasty, closure of the VSD and strip annuloplasty of the MV. One year after the operation, she underwent proximal descending aorta replacement due to aneurysm of the coarctoplasty site. She died of bacterial endocarditis 1 year after the second operation. The overall survival rate was 97.1 ± 2.1% at 15 years (Fig. 1).

3.2. Re-operation

Intra-operative revisions of MV repair, guided by TEE, were performed in nine (6.5%) patients. Excluding the patients who had received intra-operative revisions, 26 patients required 29 MV re-operations during follow-up, and 11 of these required 12 MV replacements. Ten out of the 57 patients (18%) who initially underwent Wooler—Kay type annuloplasty for MR underwent re-operation for progressive MR. Four of these patients underwent re-do Wooler—Kay type annuloplasty, two underwent posterior annuloplasty using suture plication and one underwent strip annuloplasty. Three older children underwent artificial ring annuloplasty. Two out of the 18 patients (11%) who initially underwent strip annuloplasty underwent re-operation (MV replacement and re-do strip annuloplasty) for progressive MR. One patient who underwent Duran ring annuloplasty (25 mm) for MR at 2 years of age developed progressive MS due to pannus formation, and she underwent removal of the ring and Wooler—Kay type annuloplasty 7 years after the initial operation. One out of

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**Table 3**

Techniques used for repair of MV.

<table>
<thead>
<tr>
<th>Repair technique</th>
<th>Patients</th>
<th>%</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Mitral regurgitation</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Wooler—Kay annuloplasty</td>
<td>57</td>
<td>45.6</td>
</tr>
<tr>
<td>Cleft repair</td>
<td>22</td>
<td>17.6</td>
</tr>
<tr>
<td>Strip annuloplasty</td>
<td>18</td>
<td>14.4</td>
</tr>
<tr>
<td>Ring annuloplasty</td>
<td>14</td>
<td>11.2</td>
</tr>
<tr>
<td>Artificial chordae</td>
<td>12</td>
<td>9.6</td>
</tr>
<tr>
<td>Chordae shortening</td>
<td>10</td>
<td>8.0</td>
</tr>
<tr>
<td>Posterior annulus plication</td>
<td>9</td>
<td>7.2</td>
</tr>
<tr>
<td>Papillary muscle splitting</td>
<td>7</td>
<td>5.6</td>
</tr>
<tr>
<td>Secondary chordae resection</td>
<td>6</td>
<td>4.8</td>
</tr>
<tr>
<td>Edge-to-edge repair</td>
<td>5</td>
<td>4.0</td>
</tr>
<tr>
<td>Commissure obliteration</td>
<td>5</td>
<td>4.0</td>
</tr>
<tr>
<td>Chordae transfer</td>
<td>4</td>
<td>3.2</td>
</tr>
<tr>
<td><strong>Mitral stenosis</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Papillary muscle splitting</td>
<td>11</td>
<td>78.6</td>
</tr>
<tr>
<td>Commissurotomy</td>
<td>4</td>
<td>28.6</td>
</tr>
<tr>
<td>Resection of supravalvar ring</td>
<td>4</td>
<td>28.6</td>
</tr>
<tr>
<td>Secondary chordae resection</td>
<td>2</td>
<td>14.3</td>
</tr>
<tr>
<td>Leaflet slicing</td>
<td>1</td>
<td>7.1</td>
</tr>
<tr>
<td>Fenestration of chordae</td>
<td>1</td>
<td>7.1</td>
</tr>
</tbody>
</table>

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Fig. 1. Kaplan—Meier curves showing overall survival. Numbers above the x-axis represent patients at risk.
the five patients (20%) who initially underwent edge-to-edge repair underwent artificial chordae formation for progressive MR. A 2-year-old patient who underwent resection of supravalvar ring developed progressive MS due to leaflet thickening and underwent leaflet slicing 1 year after the initial operation. A 2-month-old patient with MS who underwent papillary muscle splitting and commissurotomy developed progressive MS and underwent re-do papillary muscle splitting and commissurotomy 1 year after the initial operation. A 9-month-old patient who underwent papillary muscle splitting and commissurotomy developed progressive MR and underwent artificial chordae formation and artificial ring annuloplasty 11 years after the initial operation. At 15 years, freedom from re-operation (excluding intra-operative revisions) and MV replacement was 76.9/69.4% and 89.5/88.3%, respectively (Figs. 2 and 3). Diagnosis of MS and MV status on discharge (MR grade 3 or MS mean gradient 10 mmHg) were significant risk factors for re-operation (Table 4).

### 3.3. Follow-up

Among the 122 survivors with MR, 102 patients (84%) underwent echocardiography during follow-up. Significant improvement in degree of MR was noted. Degree of MR was none in 42 patients, grade 1 in 36, grade 2 in 19 and grade 3 in five. All patients who underwent edge-to-edge repair showed mild MR and no MS. Among 14 survivors with MS, eight patients (57%) underwent echocardiography during follow-up. The degree of MS decreased significantly and median MS gradient was 2.8 mmHg (0—10 mmHg). All survivors remain in the NYHA class I or II.

### 4. Discussion

Surgical management of MV disease in children is challenging because of the delicate and fragile nature of MV tissue in infants, wide spectrum of pathology and a high incidence of co-existing cardiac anomalies. Surgical options include MV repair and MV replacement with mechanical prostheses. MV replacement in paediatric patients poses significant problems because of the limited availability of adequate-sized prostheses for small children, the need for anticoagulation and the poor long-term result with reported 10-year survivals of 33—74% [1—4]. Oversized mechanical prostheses in small children resulting in prosthes—patient mismatch were associated with poor survival [1]. Supra-annular MV replacement was associated with worse survival than annular MV replacement and was associated with increased risk of complete heart block when the prosthesis was subsequently replaced [4].

When feasible, MV repair is desirable and should be attempted because it conserves the subvalvar apparatus and ventricular geometry, preserving left ventricular function. Preserved left ventricular function leads to survival benefits in the long term [14]. Several groups have reported satisfactory results with MV repair in children with mid- to long-term survival of 77—94% [5—10]. Our results of MV repair in children were also excellent, with an overall survival of 97% at 15 years.

Annulus dilatation and leaflet prolapse were most frequently present in patients with MR (93 of 125, 74%). Annulus dilatation was repaired by various annuloplasty techniques. In the earlier part of our experience, Wooler—Kay type annuloplasty was commonly used. Ohno et al. reported the long-term results of commissure plication annuloplasty for congenital MR with freedom from re-
operation of 86% at 13 years [15]. Sugita et al. also reported the long-term results of this type of annuloplasty with freedom from re-operation of 92% at 15 years and stated that the results were excellent with abnormality of the posterior leaflet and its apparatus or normal leaflet motion [16]. In recent years, we favoured artificial ring annuloplasty for older children and posterior annuloplasty using strip or suture plication for younger children. Oppido et al. used similar annuloplasty technique with polytetrafluoroethylene (PTFE) band for small children [10]. Prêtre et al. reported suture plication posterior annuloplasty technique using resorbable sutures for small children with good short-term results [17]. We hope that the absorbable annuloplasty technique may be useful in managing children with small annuli. The issue of annular growth is important when performing annuloplasty in small children. Although we had not enough echocardiographic data to evaluate the annular growth, we did not observe any significant problems associated with annular non-growth in patients who had undergone various partial annuloplasty techniques.

Leaflet prolapse was repaired by shortening of the elongated chordae in the earlier part of our experience. Recently, we favoured the use of artificial chordae because of its simplicity and favourable mid-term outcomes in children [18–20]. Although the use of artificial chordae for MV prolapse is an established repair technique in adult patients, there has been a concern about the fate of artificial chordae used in growing paediatric patients. Minami et al. showed, by follow-up echocardiographic measurements, that a type of ‘biologic adaptation’ occurred in patients who underwent chordal replacement with PTFE, and explained this as a compensatory extensive growth of the mitral leaflet and papillary muscle [19]. This was supported by satisfactory valve function at mean follow-up of 5 years (MR was mild or less than mild in 92% of the patients).

Repair of MV cleft was quite straightforward with good results. Although indications for repair of isolated MV cleft with less than mild MR are controversial [21], we believe that it should be repaired early following diagnosis because this kind of repair has virtually no risk, can prevent secondary change of the leaflet tissue making repair difficult and can provide satisfactory results. Interestingly, edge-to-edge repair (Alfieri technique) was used in five small children with good results. They all had severe annular dilatation and anterior leaflet prolapse with some component of leaflet retraction. We hope that this technique may be useful in managing children with severely dilated annulus and poor leaflet quality that is difficult to repair with conventional techniques.

MS in children is rare and frequently associated with other obstructive lesions of the left heart structures. Surgical repair of congenital MS has been associated with greater postoperative mortality and morbidity [7,22–24]. Repair techniques for MS are rather limited compared with those for MR. In our study, papillary muscle splitting was most commonly used.

Freedom from re-operation and MV replacement was 77% and 90% at 15 years, respectively, and these are comparable to those previously reported [5–10]. Twenty-six patients required 29 MV re-operations, and 15 of these underwent re-repair. Frequently, re-repair was possible after initial Wooler–Kay type annuloplasty, by using re-do Wooler–Kay type annuloplasty, plication posterior annuloplasty or artificial rings in older children. Diagnosis of MS and MV status on discharge (MR grade ≥ 3 or MS gradient ≥ 10 mmHg) were significant risk factors for re-operation. Since 1997, intra-operative TEE was routinely performed to assess the adequacy of MV repair, and nine patients underwent intraoperative revisions with good results. Honjo et al. reported that intra-operative TEE considerably underestimated the degree of residual ativoventricular dysfunction compared with postoperative TTE [25]. We believe that surgeons should have low threshold for revision of repair on the basis of intraoperative TEE findings to improve operative results, because residual MV dysfunction was a significant risk factor for re-operation in our study.

In conclusion, MV repair in children showed excellent survival, acceptable re-operation rate and satisfactory valve function at long-term follow-up. Diagnosis of MS and residual MV dysfunction were significant risk factors for MV re-operation, but re-repair was successful in more than half of the patients who underwent re-operation.

5. Limitations of the study

This study is limited by its retrospective design and inclusion of patients managed over a 20-year period. Furthermore, it is important to acknowledge that the follow-up was incomplete (78%).

References


