Mitral Valve Plasty for Idiopathic Rupture of Mitral Valve Posterior Chordae in Infants

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Introduction: Idiopathic mitral valve chordal rupture is rare among infants. Once it has occurred, acute heart failure progresses, and emergency surgical repair is necessary in most cases. Our surgical experience with idiopathic mitral valve chordal rupture is reported.

Patients and Methods: From September 2008 to May 2012, four infants (3 males, 1 female; median age 5.5 months) underwent mitral valve plasty for severe mitral valve regurgitation due to prolapse of posterior mitral valve leaflet. Patient history, surgical procedure, operation time, mortality, postoperative echocardiography data (mitral valve regurgitation grade: 0-trivial, mild, moderate, severe, transmitral flow: TMF) and pathology were examined.

Results: Three cases required emergency surgery; 1 case, elective surgery. Intraoperative findings showed chordal rupture of the P2 segment in 3 cases and P1 + P3 segments in 1 case. Quadrangular resection with annular plication was performed for 1 case. Quadrangular resection with annular plication and the Kay procedure were performed for 3 cases. Mitral valve regurgitation improved from severe to trivial-mild in all cases. Pathological examination showed a myxomatous degenerative change in the mitral valve.

Conclusion: Mitral valve plasty was performed for idiopathic mitral valve chordal rupture in infants. The surgical procedures were the same as for adult cases and achieved satisfactory results.

Keywords: acute heart failure, mitral valve plasty, mitral valve regurgitation

Introduction

Recently, idiopathic mitral valve chordal rupture in infants has become widely known.1,2 Once mitral valve chordal rupture has occurred, it leads to dramatic heart failure, and appropriate treatment is needed. Since severe mitral valve regurgitation is usually refractory to medical treatment, surgical treatment is required in most cases. Mitral valve plasty is a challenging procedure because of the wide spectrum of morphologic abnormalities. Our successful experience with mitral valve plasty for idiopathic mitral valve chordal rupture is reported.

Patients and Methods

Patients

From September 2008 to May 2012, four infants (3 males, 1 female) diagnosed with severe mitral valve regurgitation were referred to our hospital. Echocardiography showed that the posterior mitral valve chordae were ruptured. The patients’ median age was 5.5 months...
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Operative procedure
After full median sternotomy, cardiopulmonary bypass (CPB) was established with direct cannulation to the ascending aorta, superior vena cava, and inferior vena cava. After cross clamping of the ascending aorta, cardioplegic solution was injected anterogradely and retrogradely. The mitral valve was exposed by conventional left atriotomy. Mitral valve configuration and the prolapse site were checked with the water test. In cases 1 to 3, Anterior and posterior commissures were compressed by Kay suture3) using 4–0 monofilament suture with an autopericardial pledget. The prolapsed posterior valve was resected in a quadrangular shape, and the mitral annulus was partially plicated by 4–0 monofilament suture with autopericardium. The resected valve was repaired with 6–0 monofilament sutures. After confirming control of mitral valve regurgitation, we weaned patients from CPB.

Evaluation
Patient history, surgical procedure, operation time, mortality, postoperative echocardiography data (mitral valve regurgitation grade: 0-trivial, mild, moderate, severe, transmitral flow: TMF) and pathology were examined.

Results
Case 1 had acute purulent lymphadenitis 10 days before the mitral valve chordal rupture. The other cases did not have a specific patient history. In cases 1–3, the first symptoms were tachypnea and poor sucking. They were seen in the hospital and diagnosed with severe mitral valve regurgitation due to posterior chordal rupture. They were referred to our hospital and emergency surgeries were performed. In case 4, though the patient suddenly developed tachypnea and poor sucking, the symptoms resolved the following day. After several days, the patient visited a hospital for a vaccination, and a cardiac murmur was noticed. Echocardiography revealed severe mitral valve regurgitation with posterior chordal rupture. Since the patient could tolerate volume overload caused by the mitral valve regurgitation, he underwent elective surgery. The segments of the mitral valve chordal rupture were P1 + P3 in case 1 and P2 in cases 2 to 4. The surgical procedures involved quadrangular resection with annular plication and Kay suture in cases 1–3, and quadrangular resection with annular plication in case 4. The average operation time was 209.5 minutes (190–240 minutes), the average aorta clamp time was 66.7 minutes (47–91 minutes) and the average cardiopulmonary bypass time was 98.2 minutes (47–91 minutes) (Table 2). The median postoperative follow-up period was 13.8 months (3.4–47.9 months). All cases are alive. Postoperative echocardiography showed that mitral valve regurgitation in the acute period was trivial in 3 cases and moderate in 1 case. During the midterm period, mitral valve regurgitation was trivial in 3 cases and mild in 1 case (Table 3). There was no mitral valve stenosis. No re-operations were needed. On pathological examination, only an acute culture check was performed in case 1, with no pathological slices. The pathological diagnosis of cases 2 to 4 were myxomatous degenerative changes in the mitral valve (Fig. 1).

Discussion
The epidemiology of and surgical procedures for idiopathic mitral valve chordal rupture in infants were reviewed.

Epidemiology
Mitral valve chordal rupture usually occurs at 2–6 months of age. It causes severe mitral valve regurgitation and leads to dramatic heart failure. The causes of mitral valve chordal rupture are primary (spontaneous)
Table 2

<table>
<thead>
<tr>
<th>Case</th>
<th>Procedure</th>
<th>Operative time (min)</th>
<th>CPB time (min)</th>
<th>Aorta clamp time (min)</th>
<th>Ventilator (POD)</th>
<th>Discharge (POD)</th>
<th>Complications</th>
<th>Postoperative status</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>QR with annular plication, Kay</td>
<td>240</td>
<td>129</td>
<td>91</td>
<td>1</td>
<td>20</td>
<td>none</td>
<td>alive</td>
</tr>
<tr>
<td>2</td>
<td>QR with annular plication, Kay</td>
<td>193</td>
<td>90</td>
<td>68</td>
<td>4</td>
<td>21</td>
<td>none</td>
<td>alive</td>
</tr>
<tr>
<td>3</td>
<td>QR with annular plication, Kay</td>
<td>215</td>
<td>100</td>
<td>61</td>
<td>1</td>
<td>20</td>
<td>none</td>
<td>alive</td>
</tr>
<tr>
<td>4</td>
<td>QR with annular plication</td>
<td>190</td>
<td>74</td>
<td>47</td>
<td>1</td>
<td>19</td>
<td>left pneumothorax</td>
<td>alive</td>
</tr>
</tbody>
</table>

QR: quadrangular resection; CPB: cardiopulmonary bypass; POD: postoperative day

and secondary, for example, infective carditis, myocardial infarction, Kawasaki disease, and trauma. In the present cases, there was no infective tissue in the mitral valve. Postoperative pathological examination revealed no vegetations. There were no appreciable past histories, such as anomalous origin of the coronary arteries, Kawasaki disease, myocardiitis, and trauma. On pathology, myxomatous degenerative change seem to be related to chordal rupture. Mitral valve tissues resected from cases 2 to 4 (Fig. 1-a, 1-b, and 1-c) were stained with Elastica-Masson stain. Normal mitral valve tissue from a neonate who died from neuroblastoma was also stained as a control (Fig. 1-d). The valve was thickened 2–4 times in tissues of ruptured chordae (Fig. 1-a, 1-b, and 1-c) compared to control tissue (Fig. 1-d). On histology, decreased elastic fibers, increased collagen fibers, and deposition of extracellular mucin were found in cases 2 to 4. These findings are consistent with myxomatous degenerative change.

Clinically, most cases presented with poor sucking and tachypnea as initial manifestations. In case 4, the infant could tolerate volume overload caused by severe mitral valve regurgitation, and elective mitral valve plasty was performed. On the other hand, acute heart failure progressed dramatically and required emergency surgery in the other cases. It is very difficult to control acute heart failure caused by chordal rupture, so most cases have to undergo mitral valve repair immediately. If there are symptoms of acute failure caused by mitral valve chordal rupture, we should not hesitate to undertake emergency surgery.

Surgical Procedure

Mitral valve replacement or mitral valve plasty is usually performed for severe mitral valve regurgitation. Mitral valve plasty is the first choice because mitral valve replacement has some problems, for example, size restriction of the prosthetic valve, relative mismatch between patient and prosthetic valve after growth of the patient, and trouble with anticoagulant drug therapy.

Previous papers have shown several procedures for mitral valve plasty. Kay suture, Alfieri stitch, artificial-chordae, chordae shortening and quadrangular resection with annular plication are performed for prolapse of the posterior mitral valve leaflet. Mitral valve plasty procedures are, therefore, selected on a case-by-case basis. In adults, quadrangular resection with annular plication is the gold standard for prolapse of the posterior mitral valve leaflet. It is a simple technique, and we are proficient in it. The same procedure was, therefore, performed in those presenting infants. One must consider two important points about mitral valve plasty in infants. First, the left atrium is small, and the surgical procedure is difficult. To get a good surgical view, we freed the superior vena cava and dissected Waterston’s groove as much as possible. As for another technique, by retracting the Kay sutures, the valve was moved to the left atrium side, and a good surgical view was obtained. During case 4 elective surgery, the left atrium was larger than in the other 3 cases. Actually, the left atrium/ascending aorta ratio was 2.81; the surgical view was better than in the other 3 cases. Second, future growth of mitral valve must be kept in mind. Therefore, we avoided mitral valve annular plasty using an annular ring. Although we performed annular compression sutures for the resected valve segment, those were loose.

Postoperative echocardiography showed that all cases did not have symptomatic mitral valve stenosis (peak TMF was below 2 m/s). However, in cases 1 and 2, color Doppler showed slight acceleration flow. Case 4 had a good view with a dilated left atrium, and Kay suture was not performed. In case 4, postoperative echocardiography...
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showed a lower peak TMF than in the other 3 cases. Although the Kay suture is one of the better techniques, indications for it should be considered.

Honjo, et al.\textsuperscript{11)} reported that mitral valve annular growth was appropriate after mitral valve plasty without prosthetic materials. In the present study, mitral valve plasty was performed without prosthetic materials, and mitral stenosis may decrease in the future.

We think that the basic principle of mitral valve plasty is common to adults and infants. The standard mitral valve plasty procedure corresponding to each type of mitral valve regurgitation is, therefore, applicable to infants. (for example, artificial-chordae for anterior mitral valve prolapse\textsuperscript{13)} and quadrangular resection with annular plication for posterior mitral valve prolapse).

Conclusion

Good results were achieved in the treatment of idiopathic mitral valve posterior chordal rupture in infants.
Future growth must be taken into account with any surgical procedure.

**Disclosure Statement**

The authors declare no conflicts of interest associated with this report.

**References**