Surgical Correction of Congenital Mitral Insufficiency*

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Congenital mitral insufficiency is most frequently associated with other intracardiac lesions, usually of the endocardial cushion defect type. Other forms of congenital mitral insufficiency are being reported with increasing frequency. Valvuloplasty procedures on these mitral valves have had a high success rate; however, in some instances replacement is necessary.

Most frequently, congenital mitral insufficiency is associated with other congenital heart defects, mainly of the endocardial cushion defect type. Clinical cases of isolated mitral insufficiency, although unusual,1,2 are being described with increasing frequency, and their clinical and pathologic features are being well documented.3–5 Surgical correction of these uncommon types of isolated mitral incompetence are also being reported with increasing frequency.6–8

This report reviews our experience with 67 cases of congenital mitral insufficiency operated upon in our department in the past ten years.

Clinical Material and Technique of Repair

Table 1 shows the types of patients with congenital mitral insufficiency operated upon by our unit.

Our surgical technique for the correction of the mitral insufficiency which accompanies endocardial cushion defects has been previously reported.9–18 We operate upon these defects using extracorporeal circulation with mild hypothermia. After opening the right atrium, the incompetent mitral valve is first corrected. A suture is placed in the thickened nodular area of the leaflets at the tip of the cleft, and the cleft is closed toward the base with interrupted silk sutures (Fig 1). A pericardial patch is used to close the ostium primum atrial defect. The conduction mechanism is avoided by placing sutures parallel to it through the base of the septal leaflet of the tricuspid valve as shown in Figure 2.

Three patients with isolated cleft of the anterior leaflet of the mitral valve are noted in Table 1. Surgical correction in those cases was similar to that used in cleft mitral valves associated with other defects of the endocardial cushion type.

One of the two patients with corrected transposition was a 15-year-old boy with two clefs of the left atrioventricular valve. These clefs were closed at operation using our routine technique. In addition, annuloplasties of both the left and right atrioventricular valves were performed. The second patient, a 52-year-old woman, had a tricuspid left atrioventricular valve, ruptured chordae tendineae, and a dilated annulus. A suture plication of the valve converting it into a biicuspid valve and annuloplasty were performed.

The patient with mitral insufficiency accompanying an anomalous left coronary artery was a ten-year-old girl. The left coronary artery was ligated at its origin from the pulmonary artery. Mitral insufficiency and left heart failure persisted, and four months later the patient was reoperated

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was repaired by a plication of the posterior leaflet and the cleft in the anterior leaflet of the tricuspid valve was sutured.

**RESULTS**

Hospital and late mortality in our series are shown in Table 1.

The causes of death in our series of endocardial cushion defects have been previously reported. It is our belief that the mitral valve should always be repaired. In two instances, early in our series, mitral insufficiency due to a cleft anterior leaflet was not recognized, and this led to early postoperative pulmonary edema and death of these two patients.

The follow-up in 55 patients with mitral insufficiency associated with endocardial cushion defects is shown in Table 2. Late results have been very gratifying. There have not been any instances of mitral stenosis following the repair in the entire series. Only four patients required reoperation to correct residual mitral insufficiency. In three of these four cases a harsh loud systolic murmur was heard immediately following operation, indicating an inadequate repair. One adult developed late mitral insufficiency. He had a large ostium primum atrial defect with dilatation of both tricuspid and mitral annuluses which were not corrected at the initial operation. With the passage of time, the insufficiency increased and he required double valve replacement. The only other valve replacement in the entire series was performed in another adult with similar valve pathology as the case described, dilatation of the mitral annulus and stretched and elongated chordae tendineae.

One of the three patients with isolated cleft of the anterior leaflet of the mitral valve had been previously diagnosed as having endocardial fibroelastosis. He is now completely asymptomatic, has no cardiac murmur and a normal heart size. The other two are greatly improved.

The 15-year-old boy with corrected transposition developed severe insufficiency and heart failure following operative repair. This was due to a lack of tissue of the "mitral" valve, and he required reoperation. Mitral valve replacement with a Starr-Edwards prosthesis was performed. Unfortunately, the patient died on the seventh postoperative day from bronchopneumonia and pulmonary edema. The 52-year-old woman did well postoperatively, but died suddenly at home 45 days after surgery. The cause of death is not known as an autopsy was not performed.

The patient with mitral insufficiency accompanying an anomalous left coronary artery is leading a normal life, free of symptoms, four years after sur-Thus, the results of our series would indicate that mitral valve repair is a safe and gratifying procedure.

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**Table 1—Congenital Mitral Insufficiency**

<table>
<thead>
<tr>
<th>Mortality</th>
<th>No.</th>
<th>Early</th>
<th>Late</th>
</tr>
</thead>
<tbody>
<tr>
<td>Mitral insufficiency as part of endocardial cushion defects:</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Partial A-V canal</td>
<td>38</td>
<td>8</td>
<td>3</td>
</tr>
<tr>
<td>Complete A-V canal</td>
<td>23</td>
<td>4</td>
<td>0</td>
</tr>
<tr>
<td>Isolated cleft anterior leaflet of mitral valve</td>
<td>3</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>Mitral insufficiency and corrected transposition of the great vessels</td>
<td>2</td>
<td>1</td>
<td>1</td>
</tr>
<tr>
<td>Mitral insufficiency and anomalous left coronary artery</td>
<td>1</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td></td>
<td></td>
<td>67</td>
<td>13</td>
</tr>
</tbody>
</table>

**Table 2—Endocardial Cushion Defects: Follow-up Findings in 55 Patients (6 Months to 8 Years)**

<table>
<thead>
<tr>
<th>Mortality</th>
<th>No.</th>
<th>Percent</th>
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<tr>
<td>Improvement in symptomatology</td>
<td>51</td>
<td>93</td>
</tr>
<tr>
<td>Decrease in cardiac size</td>
<td>46</td>
<td>85</td>
</tr>
<tr>
<td>Decrease in pulmonary hypertension</td>
<td>51</td>
<td>93</td>
</tr>
<tr>
<td>Soft systolic murmur present</td>
<td>39</td>
<td>71</td>
</tr>
<tr>
<td>No murmurs present</td>
<td>16</td>
<td>29</td>
</tr>
</tbody>
</table>

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Surgery. The heart is normal in size and there is no murmur present.

Discussion

Competency of a normal mitral valve depends on the anatomic and functional normality not only of the leaflets but also of the annulus, chordae tendineae and papillary muscles, as well as the left atrium and left ventricle. The congenital defect responsible for insufficiency can be localized in one or several of these structures. Based on these anatomic structures, we have classified the different types of congenital mitral insufficiency which can be found clinically (Table 3).

The anomaly of the mitral valve associated with endocardial cushion defects usually consists of inadequate substance of the anterior leaflet, resulting in a complete or partial cleft. It may accompany the other defects which integrate the complex of the complete atrioventricularis communis, be a part of any of the less complicated partial forms, or associated with some bizarre, anatomic derangements included as intermediary types. In certain instances, anomalous chordae tendineae connect the valve to the ventricular septum and to anomalous papillary muscles.14,15

In small children, under two years of age, we prefer not to do open heart surgery because the friability of the leaflets prevents holding the sutures well. If surgery is necessary in these cases, it may be better to do a pulmonary artery banding, although this at times may increase the severity of the insufficiency.16 The severity of the mitral insufficiency is not always increased, however, in the presence of a cleft mitral valve. We have banded two patients with endocardial cushion defects with cleft mitral valves with significant improvement (Fig 3 and 4). In one of these, the presence of a cleft mitral valve was confirmed at subsequent open heart repair; in the other the diagnosis of complete A-V canal was made on clinical grounds, cardiac catheterization and cineangiography. The latter shows a typical gooseneck appearance of the left ventricular outflow tract of endocardial cushion defect with associated deformed mitral valve (Fig 5).

Table 3—A Classification of Congenital Mitral Insufficiency

1. Associated with other congenital heart anomalies
   a. Endocardial cushion defects
   b. Corrected transposition of the great vessels
   c. Chronic ischemia and fibrosis due to aberrant coronary arteries
   d. Miscellaneous (endocardial fibroelastosis, patent ductus arteriosus, aneurysmal dilation of left atrium or left ventricle, ventricular septal defects, coarctation of the aorta)
2. Isolated
   a. Isolated cleft leaflets
   b. Perforated leaflets
   c. Abnormal or accessory chordae tendineae-anomalous chordal insertion
   d. Isolated dilated annulus
   e. Double orifice of the mitral valve
   f. Ebstein anomaly of the mitral valve
   g. Anomalies of the papillary muscles

Figure 3. Preoperative chest x-ray film of infant, aged 2 1/2 months, with endocardial cushion defect with cleft mitral valve.

Figure 4. Post-banding chest x-ray film of same patient shown in Figure 3, at age five years. There is persistent moderate cardiomegaly with essentially normal pulmonary vascularity.
Several authors have advocated the use of prosthetic valves in children with complete A-V canal associated with severe mitral incompetence.\textsuperscript{17,18} We have avoided doing this as we think in the long run that this is not a satisfactory answer. A repair should always be attempted first. Only when the valve tissue is inadequate or a grossly incompetent mitral valve remains after the repair should the valve be replaced. The possibility of utilizing an inverted aortic homograft or heterograft should be considered in these instances.

In corrected transposition of the great vessels, the left atroioventricular valve has the form and structure of the tricuspid valve of normal hearts. In other instances, a grossly incompetent "mitral" valve\textsuperscript{19,20} has been related to short chordae, Ebstein's malformation of the valve\textsuperscript{21} or other defects of the cusps.\textsuperscript{22}

Mitral insufficiency has also been reported due to an anomalous left coronary artery arising from the pulmonary artery.\textsuperscript{23-26} These patients can present the clinical picture of endocardial fibroelastosis. Severe myocardial damage from chronic ischemia and the resultant cardiac dilatation can lead to a dilated mitral annulus which causes the insufficiency. The diagnosis of this entity is important because of the possibility of surgical correction of the anomalous coronary and the dilated annulus, as well as the reversibility of left heart failure, and the prevention of further myocardial damage.

Some degree of thickening of the leaflets of the mitral valve can be present in cases of endocardial fibroelastosis. The chordae can also become involved, limiting even more the mobility of the valve.\textsuperscript{27} Calcification is rare in childhood but when present is usually superimposed on a previously damaged mitral valve.\textsuperscript{28} Certain cases of mitral insufficiency accompanying patent ductus arteriosus have been related to the presence of endocardial sclerosis affecting the valve leaflets.

Aneurysmal or saccular dilatation of the left atrium accompanied by mitral insufficiency has also been reported.\textsuperscript{1,26} In these cases, as with those of aneurysmal dilatation of the left ventricle, the underlying pathology responsible for the insufficiency is an isolated or unexplained dilatation of the mitral annulus.

The isolated cleft of the anterior leaflet of the mitral valve as a separate entity was first described by Edwards.\textsuperscript{4} In a similar way, anomalous insertion of the chordae tendineae, especially those connecting with the posterior leaflet, can also exist as an isolated and chief defect. In certain instances, it is the lack of a proper chordal attachment that produces mitral incompetence.\textsuperscript{29} and in rare occasions abnormal chordae exert a restraining influence on the leaflet preventing the proper closure of an accessory commissure.\textsuperscript{3} The same etiology of accessory commissures with accessory chordae producing a leak through the valve can be incriminated in some instances of double mitral valve with mitral incompetence.\textsuperscript{30}

A dilated mitral annulus usually accompanies other anomalies of the leaflets or chordae, but it may appear as the only mitral pathology. More knowledge is needed in relation to the specific function that the papillary muscles play in the closure of the mitral valve. Recent studies have shown that selective papillary muscle infarction does not necessarily result in mitral insufficiency.\textsuperscript{51} It is possible, however, that congenital muscular anomalies can result in papillary muscular dysfunction responsible for some obscure cases of congenital mitral incompetence.

In isolated mitral insufficiency, reparative procedures usually give good results.\textsuperscript{6} Suturing of the cleft or plication of the anterior leaflet have been very rewarding in our hands; the plication of the posterior or mural leaflet, however, does not always lead to such successful results in this group of patients,\textsuperscript{82} and a replacement is necessary in many instances.

\textbf{References}


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