Coarctation of the Aorta with Congenital Mitral Regurgitation

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SUMMARY

Among 861 infants and children with coarctation of the aorta examined between 1950 and 1973, inclusive, 18 (2.1%) also had congenital mitral regurgitation (MR).

Resection of the coarctation of the aorta was performed in 16 of these 18 patients (89%). There were three operative deaths early in the series, giving a total mortality rate of 18% since 1950. Over the past decade, mortality in this group has been reduced to zero. Mitral valve replacement was performed following resection of the coarctation in 2/13 patients (13%), both successfully. The follow-up of the 13 postoperative patients and the two who have not undergone surgery has ranged from 5 to 18 years, with a median of nine years. At the last examination, an appreciable pressure gradient persisted between the right arm and the legs in 4/15 patients (27%). Weight remained below the third percentile in none. The clinical picture of congestive heart failure was present in none, but significant cardiomegaly was found radiologically in 11/15 (73%) and the electrocardiogram remained abnormal in 8/15 (53%). Over the period of follow-up, the MR became clinically worse in 4/15 patients (27%), remained unchanged in 7/15 (47%), and improved following resection of the coarctation in 4/15 patients (27%).

The pathologic anatomy of congenital MR associated with coarctation of the aorta was reviewed and classified. Two unique cases were presented — rupture of chordae tendineae and perforation of the posterior leaflet, both apparently congenital. When mitral surgery becomes necessary, an understanding of the pathologic anatomy of congenital MR may well make it possible to avoid mitral valve replacement in selected cases.

Additional Indexing Words:
Aorta
Xenograft
Coarctation of the aorta
Mitral valve replacement
Congenital mitral regurgitation

**Coarctation of the Aorta** is frequently associated with patent ductus arteriosus, bicuspid aortic valve, and ventricular septal defect. However, the association of coarctation with congenital mitral regurgitation (MR) is much less well known. This report presents our experience with this challenging combination of anomalies.

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**Material and Methods**

This study is based on 18 patients with coarctation of the aorta and congenital MR: 11 males and seven females (males/females = 1.6/1).

The diagnosis of coarctation of the aorta was confirmed by surgery in 15 patients, by cardiac catheterization in two and by necropsy in one. The diagnosis of MR was established by angiocardiography in 12 patients, by necropsy in three, and by clinical and hemodynamic criteria in three. The latter three patients were catheterized more than 10 years ago, before satisfactory angiocardiography was available to us, and the diagnosis of MR was based on the typical apical pansystolic murmur with axillary radiation, left atrial enlargement on fluoroscopy, and elevated pulmonary wedge or left atrial pressures. Patients with evidence of prior rheumatic fever or bacterial endocarditis were excluded, as were patients with common atrioventricular canal.

This was designed as a study of coarctation of the aorta in which the associated MR, though congenital, was not due to endocardial cushion defect in which MR is to be expected.

**Results**

From 1950 to 1973, inclusive, 861 infants and children with coarctation of the aorta were seen at the
Children’s Hospital Medical Center in Boston. Of these, 18 had associated congenital MR (2.1%). The coarctation was postductal in 13, juxta ductal in five, and preductal in none.

Additional cardiovascular anomalies were present in seven of these 18 cases (39%): 1) patent ductus arteriosus, with exclusively left-to-right shunt, in four; 2) aortic stenosis in two, valvar in one and subvalvar in the other; and 3) ventricular septal defect that was small and closed spontaneously in one.

Initial Visit to a Cardiologist

The salient clinical findings at the initial cardiac evaluation are summarized in table 1. The age when first seen ranged from three weeks to 10 years, the median being five months. The weight ranged from below the 3rd to the 75th percentile, the median being the third percentile.

The characteristic murmur of MR was heard on the first visit in all but two patients (cases 4 and 13, table 1). In one (case 4), the typical murmur was heard two months after the first visit, at six months of age, while in the other (case 13) the typical murmur was not detected until three years after the initial visit, when the child had reached 3½ years of age.

An apical mid-diastolic rumble, without presystolic accentuation, was heard in 11 of these 18 patients (61%). Significant mitral stenosis was excluded, subsequently, in all by disappearance of the murmur, by catheterization or by necropsy. This apical mid-diastolic rumble without presystolic accentuation was considered to indicate increased blood flow across the mitral valve secondary to the MR, patent ductus arteriosus, or ventricular septal defect.

Congestive heart failure with tachypnea, tachycardia, hepatomegaly and cardiomegaly was present in 13 of these 18 patients (72%). All 10 infants who presented initially under one year of age were in cardiac failure, whereas only three of the eight patients first seen beyond this age were in failure (37.5%).

Electrocardiograms were available in all 18 patients and showed pure right ventricular hypertrophy in four (22%), pure left ventricular hypertrophy in five (28%), combined ventricular hypertrophy in six (33%), and appeared normal for age in three (17%). Under one year of age none had a normal electrocardiogram, and over one year of age none had pure right ventricular hypertrophy.

### Table 1

<table>
<thead>
<tr>
<th>Case</th>
<th>Sex</th>
<th>Date</th>
<th>Age (yrs)</th>
<th>Weight %ile</th>
<th>BP RA</th>
<th>Log</th>
<th>Murmurs Syst</th>
<th>CHF</th>
<th>ECG RVH LVH</th>
<th>X-ray CE LAE</th>
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<td>N.T.</td>
<td>F</td>
<td>12/22/53</td>
<td>3/12</td>
<td>10</td>
<td>122F</td>
<td>70F</td>
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<td>+</td>
<td>+</td>
</tr>
<tr>
<td>2.</td>
<td>A.K.</td>
<td>F</td>
<td>6/3/54</td>
<td>3/12</td>
<td>10</td>
<td>124/70</td>
<td>60F</td>
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<td></td>
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</tr>
<tr>
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<td>D.G.</td>
<td>M</td>
<td>8/24/54</td>
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<td>10</td>
<td>125/80</td>
<td>75F</td>
<td>+</td>
<td>0</td>
<td>+</td>
</tr>
<tr>
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<td>T.W.</td>
<td>M</td>
<td>10/22/57</td>
<td>4/12</td>
<td>&lt; 3</td>
<td>150/80</td>
<td>85F</td>
<td>0</td>
<td>+</td>
<td>+</td>
</tr>
<tr>
<td>5.</td>
<td>J.H.</td>
<td>F</td>
<td>1/27/58</td>
<td>7.5/12</td>
<td>10</td>
<td>140/90</td>
<td>110/90</td>
<td>+</td>
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<td>0</td>
</tr>
<tr>
<td>6.</td>
<td>C.M.</td>
<td>F</td>
<td>2/17/58</td>
<td>5/12</td>
<td>&lt; 3</td>
<td>170/100</td>
<td>65F</td>
<td>+</td>
<td>0</td>
<td>+</td>
</tr>
<tr>
<td>7.</td>
<td>L.B.</td>
<td>M</td>
<td>11/24/58</td>
<td>2</td>
<td>&lt; 3</td>
<td>150/100</td>
<td>95/60</td>
<td>+</td>
<td>0</td>
<td>0</td>
</tr>
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<td>R.T.</td>
<td>M</td>
<td>5/25/59</td>
<td>8/12</td>
<td>&lt; 3</td>
<td>160/60</td>
<td>85F</td>
<td>+</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>9.</td>
<td>K.C.</td>
<td>M</td>
<td>11/28/61</td>
<td>2/12</td>
<td>25</td>
<td>190/90</td>
<td>60F</td>
<td>+</td>
<td>0</td>
<td>+</td>
</tr>
<tr>
<td>10.</td>
<td>J.M.</td>
<td>M</td>
<td>7/19/62</td>
<td>10</td>
<td>25</td>
<td>140/100</td>
<td>106/96</td>
<td>+</td>
<td>0</td>
<td>+</td>
</tr>
<tr>
<td>11.</td>
<td>R.D.</td>
<td>M</td>
<td>11/27/62</td>
<td>2 3/12</td>
<td>&lt; 3</td>
<td>118/88</td>
<td>74/40</td>
<td>+</td>
<td>0</td>
<td>+</td>
</tr>
<tr>
<td>12.</td>
<td>J.B.</td>
<td>M</td>
<td>6/17/63</td>
<td>9</td>
<td>75</td>
<td>160/104</td>
<td>94F</td>
<td>+</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>13.</td>
<td>T.R.</td>
<td>M</td>
<td>8/9/63</td>
<td>3/12</td>
<td>&lt; 3</td>
<td>135/90</td>
<td>90F</td>
<td>0</td>
<td>0</td>
<td>+</td>
</tr>
<tr>
<td>14.</td>
<td>S.B.</td>
<td>F</td>
<td>9/12/64</td>
<td>3/2/52*</td>
<td>&lt; 3</td>
<td>160/90</td>
<td>60F</td>
<td>+</td>
<td>+</td>
<td>+</td>
</tr>
<tr>
<td>15.</td>
<td>L.W.</td>
<td>M</td>
<td>11/7/66</td>
<td>3 5/12</td>
<td>50</td>
<td>177/88</td>
<td>84F</td>
<td>+</td>
<td>+</td>
<td>0</td>
</tr>
<tr>
<td>16.</td>
<td>E.S.</td>
<td>F</td>
<td>2/1/67</td>
<td>1 11/12</td>
<td>&lt; 3</td>
<td>160/90</td>
<td>80</td>
<td>+</td>
<td>+</td>
<td>+</td>
</tr>
<tr>
<td>17.</td>
<td>K.Cr.</td>
<td>F</td>
<td>9/9/67</td>
<td>2/12</td>
<td>&lt; 3</td>
<td>110F</td>
<td>60F</td>
<td>+</td>
<td>+</td>
<td>+</td>
</tr>
<tr>
<td>18.</td>
<td>N.B.*</td>
<td>M</td>
<td>12/30/68</td>
<td>2/12</td>
<td>10</td>
<td>120F</td>
<td>40F</td>
<td>+</td>
<td>0</td>
<td>+</td>
</tr>
</tbody>
</table>

Abbreviations: F = flush blood pressure; + = present; 0 = absent; -- = not evaluated; BP = blood pressure; CE = cardiac enlargement; CHF = congestive heart failure; Diast = diastolic; LAE = left atrial enlargement; LVH = left ventricular hypertrophy; %ile = percentile; RA = right arm; RVH = right ventricular hypertrophy; Syst = systolic.

*3/52 = 3 weeks.
**Initial cardiac evaluation was performed at the Babies’ Hospital, New York.

Chest X-rays revealed cardiomegaly in all. Fluoroscopy showed left atrial enlargement in 10 of 13 patients (77%).

Catheterization Findings
The preoperative hemodynamic data in 16 patients are summarized in table 2. Pressures across the coarctation were measured in 10 cases. The systolic gradient was less than 50 mm Hg in four (cases 2, 3, 5 and 7), between 51 and 80 mm Hg in five (cases 6, 8, 11, 15 and 16), and greater than 80 mm Hg in one (case 9).

Left atrial or pulmonary capillary wedge mean pressure was measured in 14 patients and exceeded 10 mm Hg in 12 (86%). The "v" wave equalled or exceeded 15 mm Hg in 10 of 13 cases with satisfactory tracings (77%).

The pulmonary arterial mean pressures were normal or only mildly elevated (less than 25 mm Hg) in seven of the 16 patients (44%), moderately elevated (between 25 and 44 mm Hg) in eight (50%), and markedly elevated (above 45 mm Hg) in only one (6%).

Left ventricular end-diastolic pressure was elevated (above 10 mm Hg) in six of the seven patients in whom it was measured (86%). In these six cases, the pulmonary arterial wedge pressure was also elevated.

Angiocardiograms were available in 10 patients. Selective left ventriculography was performed in seven and showed significant MR in all (fig. 1). Pulmonary arterial injection in three patients showed reopacification of the left atrium during left ventricular systole, suggesting at least moderate MR.

Surgery
Resection of the coarctation was performed in 16 of these 18 patients (89%), from one month to 12 years of age, the median being two years of age. Two have had no surgery. One (case 5) has diffuse hypoplasia of the aortic arch without discrete coarctation, and the parents of the other (case 10) have repeatedly refused surgery. The indication for surgery was congestive heart failure in eight patients and the procedure was elective in eight. The majority (11 of 16) had surgery within one year of their initial visit, eight of these 11 being in congestive failure.

Death occurred intraoperatively or immediately postoperatively in three of these 16 patients early in the series (cases 3, 4 and 12, tables 1 and 2). Thus, the mortality rate for the total series since 1950 was 18%. However, over the past decade, the operative mortality in this group has been zero.

Mitral valve replacement was performed following coarctation resection in two of the 13 surviving patients, both successfully: in case 17 at 4½ years using a #5 Kay-Shiley disc valve, and in case 18 at 4½ years using a 25 mm Hancock glyceraldehyde preserved porcine xenograft, model 332.

Follow-Up
The 13 operative survivors and the two who have not had surgery have been followed for four to 18 years, the median follow-up duration being nine years. These 15 patients now range from five to 23 years of age, with a median of 11 years.

The clinical findings at the last visit are summarized in table 3. At that time none of the 15 patients were below the third percentile for weight, whereas eight of the 15 were below the third percentile initially (53%). An appreciable pressure gradient (> 25 mm Hg) persisted between the right arm and the legs in four (27%). The 13 patients who have not had mitral valve replacement all have a typical apical pansystolic murmur. The apical mid-diastolic rumble without presystolic accentuation persisted in seven of the 15 patients (47%). The clinical picture of congestive heart failure, present initially in 12 of these 15 patients (80%), was absent. However, cardiomegaly persisted radiologically in 11 (73%). The electrocardiogram, initially abnormal in 12 (80%), remained abnormal in eight (53%).

What Has Been the Natural History of the Congenital Mitral Regurgitation?
It has been progressive in four of the 15 children (27%): patients 10, 15, 17 and 18 (table 3). Patient 10,

![Figure 1](image-url)

*Figure 1*
Selective left ventricular angiocardiography, case 15, via retrograde aortic catheter, postero-anterior projection. Note the coarctation of the aorta (C) and the marked reflux of contrast from the left ventricle (LV) into the enlarged left atrium (LA) and left atrial appendage (LAA). Ao = aorta.
### Table 2

**Pre-Operative Catheterization Data—Pressures mm Hg**

<table>
<thead>
<tr>
<th>Case</th>
<th>Date</th>
<th>Age (yrs)</th>
<th>Asc Ao</th>
<th>Desc Ao</th>
<th>LA</th>
<th>PC</th>
<th>PA</th>
<th>LV</th>
<th>Comments</th>
</tr>
</thead>
<tbody>
<tr>
<td>1.</td>
<td>10/28/58</td>
<td>5 6/12</td>
<td>155/70</td>
<td>95/65</td>
<td>a = 12</td>
<td>v = 15</td>
<td>30/10</td>
<td>135/15</td>
<td>MR on LV cine, LV puncture</td>
</tr>
<tr>
<td>2.</td>
<td>10/11/57</td>
<td>3 8/12</td>
<td>130/70</td>
<td>95/65</td>
<td>a = 22</td>
<td>v = 24</td>
<td>60/25</td>
<td>100/19</td>
<td>LV puncture, General anesthesia</td>
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<td>3.</td>
<td>1/5/56</td>
<td>2 9/12</td>
<td>+120/66</td>
<td>+90/70</td>
<td>v = 16</td>
<td>34/10</td>
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</tr>
<tr>
<td>4.</td>
<td>1/15/58</td>
<td>7/12</td>
<td>+120/66</td>
<td>+90/70</td>
<td>v = 30</td>
<td>60/25</td>
<td></td>
<td></td>
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</tr>
<tr>
<td>5.</td>
<td>4/24/61</td>
<td>10 8/12</td>
<td>138/70</td>
<td>110/60</td>
<td>a = 11</td>
<td>v = 11</td>
<td>20/10</td>
<td>160/12</td>
<td>MR on LV cine</td>
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<tr>
<td>6.</td>
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<td>5/12</td>
<td>+125/55</td>
<td>+57/42</td>
<td>v = 22</td>
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<td>60/10</td>
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<td>7.</td>
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<td>3 6/12</td>
<td>140/85</td>
<td>104/80</td>
<td>a = 11</td>
<td>v = 24</td>
<td>45/15</td>
<td>220/25</td>
<td>MR on LV cine</td>
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<tr>
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<td>5/27/59</td>
<td>8/12</td>
<td>*150/74</td>
<td>*90/56</td>
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<td>58/34</td>
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<td>+210/84</td>
<td>+83/38</td>
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<td>105/70</td>
<td></td>
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<td>v = 12</td>
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<td>v = 18</td>
<td>70/25</td>
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<td>*122/92</td>
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<td>*45/40</td>
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<td>v = 23</td>
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<td>105/40</td>
<td></td>
<td></td>
<td>v = 30</td>
<td>45/15</td>
<td></td>
<td>MR on LV cine</td>
</tr>
</tbody>
</table>

Abbreviations: + = brachial artery; ++ = femoral artery; → = withdrawal; * = simultaneous; Asc Ao = ascending aorta; Desc Ao = descending aorta; LA = left atrium; LV = left ventricle; MR = mitral regurgitation; PA = pulmonary artery; PC = pulmonary capillary.
who also has aortic stenosis, has had bacterial endocarditis three times during the follow-up period. He now has moderate mitral and aortic regurgitation, and is regarded as a candidate for double valve replacement when the parents agree. In patient 15, despite some

**Table 3**

<table>
<thead>
<tr>
<th>Case</th>
<th>Year</th>
<th>Age (yrs)</th>
<th>DUR of F-U (yrs)</th>
<th>Weight.downcase()</th>
<th>BP</th>
<th>Chf</th>
<th>Mmururs</th>
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<th>X-ray</th>
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<td>17</td>
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<td>+</td>
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<td>15</td>
<td>10</td>
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<td>110/70</td>
<td>+</td>
<td>+</td>
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<td>+</td>
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<td>+</td>
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<td>9</td>
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Abbreviations: + = present; 0 = absent; LAE = left atrial enlargement; -- = not evaluated; LVH = left ventricular hypertrophy; CE = cardiac enlargement; RA = right arm; CHF = congestive heart failure; RVH = right ventricular hypertrophy; Diast = diastolic; Syst = systolic; DUR of F-U = duration of follow-up.

*Deep S in V1 and V4 suggesting RVH probably represents late posterior and rightward forces occasionally seen with left ventricular hypertrophy.*

Cases 3, 4, and 12 died at surgery.

**Figure 2**

Heart specimen of case 3. **a)** The left atrium (LA) is markedly hypertrophied and enlarged. The anterior leaflet of the mitral valve is asymmetrical, triangular, with a thickened and rolled free margin indicating mitral regurgitation (MR). The anterior mitral leaflet inserts directly into the large postero-median papillary muscle of the left ventricle (LV). The antero-lateral papillary muscle is not visible from this left atrial view. The chordae tendineae at the postero-median commissure are few, short and thick. **b)** The left ventricular view reveals that the antero-lateral papillary muscle group (APM) is very small and high, whereas the postero-median papillary muscle group (PPM) is much larger and lower, resulting in an asymmetrical insertion of the anterior mitral leaflet. The chordae tendineae attaching the anterior mitral leaflet to the papillary muscles are very short, and there are almost no interchordal spaces. Endocardial fibroelastosis (EFE) is diffuse in extent, relatively mild in degree, but is most obvious in the region of the hypoplastic APM. The aortic valve (AoV) is unremarkable. The rolling of the free margin of the anterior mitral leaflet indicating MR is well seen.

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apparent progression of the MR, cardiac enlargement is still only moderate. Patients 17 and 18 have had mitral valve replacement, as indicated previously. Patient 18 was operated on recently and is progressing well. Patient 17 was operated on a year ago and there has been a striking reduction in heart size.

Mitral regurgitation remains clinically unchanged in seven of these 15 patients (47%). Although mild-to-moderate cardiomegaly persists, these patients are presently well compensated without medication. In adult life, some of these patients may need mitral valve surgery.

Mitral regurgitation has improved following resection of the coarctation in four of these 15 patients (27%): cases 1, 6, 14 and 16 (table 3). Cardiac enlargement has decreased and the electrocardiographic abnormalities have become less pronounced. Mitral regurgitation now appears clinically mild and mitral valve surgery probably will not be necessary.

Pathology

Three formalin fixed heart specimens (cases 3, 4 and 12) and one surgically excised mitral valve (case 18) were studied.

Case 3 (A56-16) had surgical excision of an adult type of coarctation of the aorta at 2 1/2 years of age. The aortic valve was bicuspid due to rudimentary development of the left coronary-noncoronary com-
misure, but aortic stenosis was not present.

Congenital MR was related to short, thick chordae tendineae (fig. 2). The anterior mitral leaflet inserted directly into the postero-medial papillary muscle group without intervening chordae tendineae (fig. 2a and b). The chordae tendineae that were present, for example at the postero-medial commissure (fig. 2a) were few, short and thick when compared with normal chordae tendineae (fig. 3). The antero-lateral papillary muscle group was markedly hypoplastic, whereas the postero-medial papillary muscle group was considerably hypertrophied (fig. 2b). Endocardial fibroelastosis of the left ventricle was diffuse in extent, but mild to moderate in degree (fig. 2b).

From the left atrial aspect — the surgeon’s view — the asymmetry of the anterior mitral leaflet was striking (fig. 2a), being approximately triangular. Normally, the anterior mitral leaflet is approximately semicircular (fig. 3). The anterior mitral leaflet appeared asymmetrical because the leaflet adjacent to the antero-lateral commissure was much shallower than that adjacent to the postero-medial commissure (fig. 2a). This, in turn, appeared to be due to the abnormal height and smallness of the hypoplastic

![Figure 3](image_url)

**Figure 3**

Normal mitral valve apparatus. In a) note that the anterior mitral leaflet is approximately semicircular. Both the upper (antero-lateral) and the lower (postero-medial) papillary muscle groups are equally well seen from the left atrial aspect. The chordae tendineae are quite long and thin, and interchordal spaces are numerous. In b) the antero-lateral papillary muscle group (APM) has four components or “heads.” Note that one papillary muscle head and its chordae anchor the commissure. LA = left atrium; LAA = left atrial appendage; MV = mitral valve; PPM = postero-medial papillary muscle group; PV = pulmonary veins; Sept I = septum primum; Sept II = septum secundum; VS = ventricular septum. b) is reproduced with permission from Van Praagh et al., Am J Cardiol 13: 510, 1964.
antero-lateral papillary muscle group (fig. 2b). This high, small antero-lateral papillary muscle group was not visible from the left atrial aspect (fig. 2a). Normally, both papillary muscle groups are seen equally well from the left atrium (fig. 3). The free margin of this asymmetrical anterior mitral leaflet was thickened and rolled, the left atrium was hypertrophied and enlarged, and jet lesions of the left atrial endocardium were found — all corroborating that MR had been present.

Case 4 (A58-23) had surgical excision of a postductal coarctation of the aorta at eight months of age. A bicuspid aortic valve was present due to rudimentary development of the right coronary-noncoronary commissure.

Mitral regurgitation was due to rupture of chordae tendineae. Left atrial hypertrophy and enlargement, and a prominent jet lesion of the left atrial endocardium corroborated MR. From the left atrial aspect, the anterior mitral leaflet was asymmetrical, being shallower toward the antero-lateral rather than the postero-medial commissure. From the left ventricular aspect (fig. 4a), it was seen that two large chordae had ruptured between the anterior mitral leaflet and the antero-lateral papillary muscle group where remnants of the ruptured chordae were found. The findings suggested that these chordae had curled back and that their ruptured ends had fused smoothly with the ventricular surface of the anterior mitral leaflet (fig. 4b). A probe could be inserted between these curled-back chordae and the anterior mitral leaflet. There was no gross or histologic evidence of bacterial endocarditis or other acquired pathology of the mitral valve.

Since the presumably ruptured ends of the chordae had fused smoothly with the anterior mitral leaflet (fig. 4b, arrows), it is clear that chordal rupture did not occur soon before death. It is thought that this chordal rupture occurred prenatally, not only because of the smooth fusion of the chordae to the ventricular surface of the anterior mitral leaflet, but also because this seven-month-old boy was first seen by a cardiologist at only four months of age when he was in congestive heart failure and his weight was below the third percentile (table 1). This case is considered to demonstrate a previously unknown anatomic type of congenital MR.

Case 12 (A63-134), at nine years of age, had surgical excision of a postductal coarctation of the aorta in which there was no lumen whatever. Postductal aortic arch atresia had produced functional interruption of the aortic arch and was associated with large circumventing collaterals.

**Figure 4**

Heart specimen, case 4. a) From the left ventricular aspect, the free margin of the asymmetrical anterior mitral leaflet is seen to be thickened and rolled, indicating mitral regurgitation. Two chordae tendineae, that should be attached to the antero-lateral papillary muscle group (APM), appeared to be curled back on the ventricular surface of the anterior mitral leaflet. b) Closer inspection confirms this impression. The previously ruptured ends of the chordae tendineae appear to have fused smoothly (arrows) with the anterior leaflet of the mitral valve (AL of MV). A probe can be passed between the curled-back chordae and the anterior mitral leaflet. Ao = aorta; LV = left ventricle; MR = mitral regurgitation; PPM = postero-medial papillary muscle group.
Mitral regurgitation was indicated by left atrial hypertrophy and enlargement with prominent endocardial jet lesions. Mitral regurgitation occurred through a gap at the antero-lateral commissure (fig. 5). This gap was due to absence of the chordae tendineae and the papillary muscle head that usually anchor the commissure. Normally, each papillary muscle group is composed of several different components or heads. For example, note that the antero-lateral papillary muscle group in figure 3b has four heads: one large one to the large anterior leaflet; one smaller one to the commissure; and two smaller heads to the smaller posterior leaflet. In case 12 (fig. 5), the commissural chordae tendineae and the papillary muscle head from which the chordae arise were absent, resulting in a commissural gap. There was no gross or histologic evidence of infarction, atrophy or fibrosis of the papillary musculature.

It is noteworthy that a gap is different from a cleft. A cleft is a fissure within the leaflet substance of the definitive valve, as in an ostium primum type of atrial septal defect. In case 12, however, the problem was not a fissure within the leaflet substance, but an unanchored, gaping commissure (fig. 5).

Case 18 (S73-1567) was a boy who underwent resection of a postductal coarctation of the aorta at two months and had mitral valve replacement at 4½ years using a 25 mm Hancock glyceraldehyde preserved porcine xenograft, model 332. The patient’s excised mitral valve (fig. 6) had a large (9 × 5 mm) perforation in the approximate center of the posterior leaflet. The margins of the perforation were smooth and glistening, 2 to 4 mm thick, without gross or histologic evidence of bacterial endocarditis, and not associated with aberrant chordae tendineae (fig. 6b). The anterior mitral leaflet, chordae tendineae, and papillary muscle heads were normally formed. The papillary musculature was grossly and histologically unremarkable. The orifice of the mitral valve appeared competent (fig. 6b), and the regurgitation seemed due entirely to the perforation.

This patient was not regarded as having a double mitral orifice 15, 16) because the anterior and posterior mitral leaflets were not fused together abnormally, and 2) because there were no aberrant chordae tendineae related to the ventricular surface of this abnormal opening (fig. 6b). This perforation was thought to be congenital, not only because the patient presented so young (in congestive heart failure at two months of age, table 1), but also because gross and histologic examination of the margin of this perforation revealed no evidence of acquired pathology. Multiple histologic sections, including special stains for bacteria and tubercle bacilli, were negative.

Discussion

Left-sided cardiovascular anomalies tend to occur together. For example, coarctation of the aorta frequently is associated with bicuspid aortic valve, as in 46% of cases in a recent anatomic study by Becker et al.4 Similarly, pre ductal coarctation of the aorta is associated occasionally with subaortic stenosis, parachute mitral valve, and supramitral stenosing ring, as was first reported by Shone et al.17 Moreover, the association between coarctation of the aorta and congenital MR may be regarded as another example of this tendency for left-sided cardiovascular malformations to be multiple.

However, the association between coarctation of the aorta and congenital MR is infrequent. Excluding common atroventricular canal (in which MR is to be expected, particularly with the ostium primum type of atrial septal defect), coarctation of the aorta was associated with congenital MR in only 18/861 infants and children in our experience (2.1%), and in only 23/407 infants and children in the study of Easthope et al. (5.7%).18

The coarctation of the aorta may be postductal, juxtaductal or preductal. In our series, the coarctation was postductal or juxtaductal (adult type) in all 18
Excised mitral valve of Case 18. a) Atrial view. For orientation, note the antero-lateral papillary muscle group (ALP), the postero-medial papillary muscle group (PMP), the anterior leaflet (Ant), the posterior leaflet (Post), and the normal appearing mitral orifice (MO) in the open position. The millimeter scale parallels what would be the frontal plane of the body. There is a large perforation (Perf) in the middle of the posterior leaflet through which the mitral regurgitation occurred. b) Ventricular view. For orientation, the mitral valve is shown as it would appear if viewed from the left ventricular apex. Note the mitral leaflets, Ant and Post, and the papillary muscles, ALP and PMP. The millimeter rule approximately parallels the supero-inferior plane (the Y axis). The mitral orifice (MO) is shown in the closed position and it appears entirely competent. Perf of posterior leaflet is relatively large (9 × 5 mm), and there are no vegetations or aberrant chordae tendineae about its margins.

cases, whereas in 13 of the cases of Easthope et al. the coarctation was preductal (infantile type) in 11, and juxtaligamental in two patients.

The pathologic anatomy of congenital MR reported in association with coarctation of the aorta is summarized and classified in table 4. We agree with Edwards and his colleagues that congenital MR may be classified in terms of which component of the mitral valve is anomalous — leaflets, chordae tendineae, or endomyocardium (table 4). However, it should also be appreciated that abnormalities of more than one component of the mitral apparatus may well

<table>
<thead>
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<td><strong>Pathologic Anatomy of Congenital Mitral Regurgitation Associated with Coarctation of the Aorta</strong></td>
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| Leaflet abnormalities | case 4,11 case 113  
Cleft anterior leaflet  
Perforation of posterior leaflet  
Thick myxomatous leaflets |
| Chordal abnormalities | case 4,11 case 113  
Short chordae  
Long chordae  
Ruptured chordae  
Deficient chordae |
| Endocardial and myocardial abnormalities | our case 3 (fig. 3), case 6,14 case 1118  
Endocardial fibroelastosis  
Dilated annulus  
Dilated chordae |

*By guest on April 20, 2017*
be present, as in case 3 (fig. 2). Therefore, table 4 should be regarded as an effort to classify congenital MR in patients with coarctation of the aorta in terms of the mitral anomaly that predominates.

The MR in cases 4 and 18 (figs. 4 and 6, respectively) is regarded as very probably congenital. These two cases are noteworthy because they are considered to be previously unknown anatomic types of congenital MR.

An understanding of the pathologic anatomy of congenital MR is relevant to its optimal surgical management. If one is dealing with short chordae tendineae — an immature mitral valve with few interchordal spaces and with leaflets that insert more or less directly into the papillary muscles (fig. 2) — then mitral valve replacement appears unavoidable. However, if one is dealing with a commissural gap due to chordal and papillary deficiency (fig. 5), then commissural plication may well suffice to render the mitral valve competent. Similarly, if one recognizes that regurgitation is due to a leaflet perforation (fig. 6), it may well be possible to patch the perforation, thereby avoiding mitral valve replacement.

Moreover, in our experience, resection of the coarctation usually produced considerable clinical improvement and the MR did not progress during childhood in the majority (11/15 patients, 73%). What proportion of these patients will ultimately require mitral valve surgery is presently unknown.

Acknowledgment

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References

Coarctation of the Aorta with Congenital Mitral Regurgitation
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