Successful Replacement of "Parachute" Mitral Valve in a Child

By Santiago Prado, M.D., Morris Levy, M.D., and R. L. Varco, M.D.

The deformity called "parachute mitral valve" represents a form of congenital mitral stenosis in which the chordae tendineae converge to insert into one papillary muscle. This type of malformation is rare and has been described previously only in association with other anomalies; with corrected transposition of the great vessels,
as a component of a developmental complex
with other obstructive lesions of the left side of the heart, supravalvular ring of the left atrium, subaortic stenosis and coarctation of the aorta; and in association with a patent ductus arteriosus, coarctation of the aorta, and an anomalous coronary artery.

It has been pointed out that "mitral commissurotomy" does not relieve the obstruction in cases of "parachute deformity" of the mitral valve. It appears pertinent therefore to report a case of isolated congenital mitral stenosis of the so-called "parachute type," in which successful replacement of the valve was accomplished with evident clinical improvement.

Case Report

R. R. (U.H. no. 969566). This 3½-year-old boy was born of a normal pregnancy and delivery, but at 24 hours of age began to have episodes of cyanosis and respiratory distress. At approximately 1½ months of age he developed a chronically recurrent diarrhea. At 6 months of age he was found to have lactose in his urine and his diarrhea subsided with a lactose-free, galactose-free diet.

He continued to have episodes of respiratory distress, which were interpreted as pneumonia or bronchitis. In addition, he exhibited easy fatigability and poor growth and development. In view of his peculiar frail habitus with sunken eyes, his chromosome pattern was investigated and a normal karyotype was found. A murmur had been heard since infancy, but was initially interpreted as nonsignificant. Subsequently, it became more prominent and viewed as the systolic murmur of a ventricular septal defect. Roentgenograms of the thorax showed a normal cardiac shadow during the first 2 years of life. Subsequently, minimal cardiomegaly appeared, with definite prominence of the left atrium and increased pulmonary vascular markings, which were attributed to a left-to-right shunt.

The electrocardiogram was initially within normal limits, but later showed wide notched P waves indicative of left atrial enlargement.

At 3 years of age, he continued to show limited exercise tolerance and had difficulty sleeping without elevation of his head by several pillows. Respiratory symptoms persisted, with hypersecretion of mucous, dyspnea, and occasional cyanosis. Fine rales were present on auscultation of the chest. There was a prominent murmur, which was now identified as diastolic in time, associated with an accentuated first sound (fig. 1). The electrocardiogram (fig. 2) showed evidence of left atrial enlargement, and the vectorcardiogram suggested right ventricular hypertrophy. Roentgenograms of the chest showed a slightly enlarged heart, an enlarged left atrium, and increased pulmonary vascularity (fig. 3).

Right heart catheterization (table 1) showed slightly increased pressures in the right atrium, right ventricle, and pulmonary artery, with definitely increased right and left pulmonary artery wedge pressures. The left side appeared to be fully saturated and with normal pressures.

Selective angiograms were obtained by injecting the right and the left ventricles. The pulmonary veins appeared dilated, and the left atrium was enlarged. The left ventricle had an unusual configuration, with a filling defect between the apex and the outflow tract (fig. 4). There was minimal insufficiency of the mitral valve. The coronary artery pattern was also unusual, the right coronary arising from the left. No pressure gradient was found between the two portions of the left ventricle. In view of the clinical findings and experience with a similar earlier case, mitral stenosis due to the anomalous papillary muscle of a parachute mitral valve was diagnosed.
"PARACHUTE" MITRAL VALVE

Figure 1
Preoperative tracing. $S_1$, first sound; $S_2$, second sound; $T$, tricuspid area; $M$, mitral area; $DM$, diastolic murmur. The diastolic murmur (rumble) is accentuated in presystole. $S_1$ is accentuated. Note the wide and notched P wave in the electrocardiogram (lead II). Paper velocity, 100 mm. per second.

Operation
The chest was opened by a right lateral thoracotomy. The lungs appeared edematous and fibrotic. The left atrium was large and tense. The mitral valve had the appearance of a fibrotic membrane with a funnel-shaped orifice and thick chordae. There appeared to be only one papillary muscle, and the left ventricular endocardium had the whitish appearance seen with endocardial fibroelastosis. It appeared mandatory to replace the mitral valve. The thickened chordae and a portion of the papillary muscle were excised, along with the anterior leaflet of the mitral valve, and a Starr-Edwards valve with an orifice area of 1.53 cm.$^2$ was inserted. Total pump time was 1 hour and 34 minutes. The lowest degree of esophageal temperature was 17 C. and the lowest rectal 27.5.

The immediate postoperative period was stormy, requiring a tracheostomy and the use of a Bennett respirator. On the third day he suffered several convulsions, but the cause was undetermined. The patient had been digitalized prior to surgery. Ten days after surgery he was placed on Warfarin sodium (coumadin). Dosage of the drug varied around 4 mg., depending upon the prothrombin time. Continuous maintenance on anticoagulants has been considered advisable.\footnote{5}

Since surgery, improvement has been evidenced by increased exercise tolerance and absence of orthopnea. The diastolic murmur is no longer present, and there is instead the auscultatory findings associated with placement of the Starr-Edwards valve.\footnote{5} The lungs are most of the time free from rales. He has had no symptoms that can be attributed to pulmonary hypertension or heart failure. However, 4 months postoperatively he had another convulsion and was therefore placed on anticonvulsive therapy.

Roentgenograms have shown some decrease in size of the left atrium but there has been no change in the over-all size of the heart shadow, and the lungs still show increased vascularity. The electrocardiogram shows only questionable left atrial enlargement.

Discussion
This appears to be the first case of successful replacement of the mitral valve in a patient with congenital mitral stenosis of the "parachute" type, and one of the few performed at this age in any type of mitral stenosis.\footnote{6-8}

Inasmuch as the orifice area of the Starr-Edwards valve is 1.53 cm.$^2$ (the effective orifice area is probably only about 80 per cent of this\footnote{9}), it is hoped that it will allow an adequate blood flow at least through the childhood years, though ultimate replacement with a larger valve may well be necessary.

Summary
A Starr-Edwards mitral valve of orifice area 1.53 cm.$^2$ was successfully inserted to replace
Figure 2

Preoperative electrocardiogram. Note the wide and notched P waves in lead II and the deep negative deflection of the P waves in V1. AORS in frontal plane is approximately +90°. There are no signs of right ventricular hypertrophy.

Figure 3

Posteroanterior roentgenogram, preoperative. Note slight enlargement of the heart, left atrial enlargement, and increased vascularity.

Figure 4

Selective left ventriculogram, preoperative. Unusual appearance of the left ventricle is seen, with mitral insufficiency manifested by the regurgitation of contrast medium into the left atrium.
"PARACHUTE" MITRAL VALVE

Table 1

Right and Left Heart Catheterization

<table>
<thead>
<tr>
<th>Site</th>
<th>Oxygen saturation, Cuvette</th>
<th>Pressure</th>
</tr>
</thead>
<tbody>
<tr>
<td>SVC</td>
<td>68</td>
<td></td>
</tr>
<tr>
<td>RA (high)</td>
<td>73</td>
<td>a = 13, v = 8, m = 7</td>
</tr>
<tr>
<td>RA (mid)</td>
<td>71</td>
<td></td>
</tr>
<tr>
<td>RA (low)</td>
<td>75</td>
<td></td>
</tr>
<tr>
<td>IVC</td>
<td>78</td>
<td></td>
</tr>
<tr>
<td>RV (low)</td>
<td>57</td>
<td>30/0-5</td>
</tr>
<tr>
<td>RV (high)</td>
<td>64</td>
<td></td>
</tr>
<tr>
<td>MPA</td>
<td>69</td>
<td>30/15</td>
</tr>
<tr>
<td>RPA</td>
<td>69</td>
<td></td>
</tr>
<tr>
<td>RPA wedge</td>
<td>99</td>
<td>M = 25</td>
</tr>
<tr>
<td>LPA wedge</td>
<td>99</td>
<td>M = 25</td>
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<tr>
<td>Aorta</td>
<td>100</td>
<td>100/70</td>
</tr>
<tr>
<td>LV</td>
<td>100</td>
<td>100/0-10</td>
</tr>
</tbody>
</table>

SVC, superior vena cava; RA, right atrium; IVC, inferior vena cava; MPA, main pulmonary artery; LPA, left pulmonary artery; RPA, right pulmonary artery; LV, left ventricle.

Note: (1) High pulmonary artery wedge pressures on both sides; (2) there was no difference in pressures between the two chambers of the left ventricle or from these to the aorta; (3) no evidence of a left-to-right shunt.

a "parachute" mitral valve producing clinically significant mitral stenosis.

References


Truth

Where lies the land to which the ships would go? Far, far ahead, is all her seamen know.—ARTHUR HUGH CLOUGH. 1819-1861.
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