Postoperative Evaluation of Mitral Valve Function in Ostium Primum Defect with Cleft Mitral Valve (Partial Form of Atrioventricular Canal)

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SUMMARY
Postoperative cardiovascular studies were carried out in eight patients with ostium primum defect and cleft mitral valve. Apical systolic murmurs were present in all patients, but in seven of them no mitral regurgitation was detected by pulmonary wedge pressure and left ventricular angiography. Surgical repair of the cleft anterior leaflet without resection of subjacent chordae tendineae, as performed in these patients, usually results in a competent mitral valve. Postoperative angiograms showed persistence of the characteristic preoperative deformity of the medial border of the left ventricular outflow tract especially in systole. As illustrated in one of these patients, subaortic obstruction occasionally complicates abnormalities of the atrioventricular valves.

Additional Indexing Words:
Endocardial cushion defect  Subaortic obstruction  Left ventricular angiography

That apical systolic murmurs after repair of ostium primum atrial defects with cleft mitral valve indicate residual or surgically induced regurgitation has been suggested. Rashell and associates reported that most of their group of 53 patients had apical systolic murmurs following surgical correction; residual incompetence of the mitral valve was presumed. In contrast, the hemodynamic studies carried out by Braunwald and Morrow in 18 patients, at intervals of 4 months to 5½ years after surgery for incomplete persistent atrioventricular canal, identified only one with significant residual mitral regurgitation. In the series of 28 patients with incomplete, or partial, common atrioventricular canal studied by Lillehei and co-workers, mitral regurgitation was suspected in 12, in only three of whom was it substantiated at postoperative right heart catheterization.

This report presents the results of an investigation which was undertaken to clarify the postoperative hemodynamic status and the anatomy in this lesion utilizing right and left heart catheterization and left ventricular angiography. Each of the eight patients studied had repair on an ostium primum defect and cleft mitral valve; systolic murmurs at the apex were a constant postoperative finding.

Methods
Cardiovascular studies were performed from 1 to 3 years after operation. Patients less than 12 years of age were premedicated with a mixture of meperidine, promethazine, and chlorpromazine. For right heart catheterization hydrogen-sensing catheter technics were employed for detection of left-to-right shunts; quantitation of the shunt was determined by oxygen saturation measurements. Right heart and pulmonary capillary pressures were determined at rest and following exercise; in six patients, cardiac output, determined by dye-dilution technic using indocyanine
green, was measured at rest and after 4 min of exercise.

Retrograde left heart catheterization was carried out with simultaneous measurements at rest of (1) left ventricular and pulmonary capillary pressures and (2) left ventricular and right ventricular pressures.

Left ventricular angiograms were successfully completed in seven patients after retrograde catheterization. Biplane frontal and lateral projections were made at a rate of 6 per second during and following the rapid injection of diatrizoate (75% Hypaque) into the left ventricle. Each film exposure was recorded in relation to a simultaneous electrocardiogram.

**Group Studied**

Eight patients of the group of 19 with ostium primum defects who had surgery between November 1961 and June 1965 were selected for study. They ranged in age from 3 to 22 years.

**Preoperative Assessment**

The diagnosis of ostium primum or endocardial cushion defect was made clinically in each patient. Electrocardiograms showed a typical superior QRS axis with counterclockwise rotation in the frontal plane. Cardiomegaly was seen on all x-rays. The only patient with disabling symptoms was the youngest (case 1) who had congestive heart failure and was receiving digitalis at the time of operation.

Auscultation revealed a variety of systolic murmurs which were usually maximal along the left sternal border. Apical systolic murmurs, grade II to III/VI, radiating into the axilla were conspicuous in four patients; in each, rumbling mid-diastolic murmurs were audible between the apex and xiphoid.

Preoperative cardiac catheterization was carried out on each patient at this institution (table 1). A left-to-right shunt at the atrial level was documented in all; pulmonary to systemic flow ratios ranged from 2:1 to 4:1. The pulmonary arterial systolic pressure was abnormally elevated (> 30 mm Hg) in three patients. The mean left atrial pressure, measured by passing the catheter through the interatrial

### Table 1

<table>
<thead>
<tr>
<th>Patient</th>
<th>Diagnosis</th>
<th>Age at operation (yr)</th>
<th>Study (yr PO)</th>
<th>Q_P/S</th>
<th>Pressure (mm Hg)</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>PA</td>
</tr>
<tr>
<td>1. L.K.</td>
<td>Common atrium mitral cleft</td>
<td>3</td>
<td>2</td>
<td>1:1 (3.5:1)</td>
<td>17/9 (33/10)</td>
</tr>
<tr>
<td>2. R.S.</td>
<td>Ostium primum mitral cleft</td>
<td>13</td>
<td>2</td>
<td>1:1 (3.0:1)</td>
<td>15/5 (25/8)</td>
</tr>
<tr>
<td>3. J.G.</td>
<td>Ostium primum mitral cleft</td>
<td>5½</td>
<td>3</td>
<td>1:1 (2.0:1)</td>
<td>20/8 (44/22)</td>
</tr>
<tr>
<td>4. R.B.</td>
<td>Ostium primum mitral cleft</td>
<td>22</td>
<td>2</td>
<td>1:1 (4.0:1)</td>
<td>30/10 (20/5)</td>
</tr>
<tr>
<td>5. S.T.</td>
<td>Ostium primum mitral cleft, tricuspid cleft</td>
<td>6</td>
<td>3</td>
<td>1:1 (2.2:1)</td>
<td>25/10 (25/8)</td>
</tr>
<tr>
<td>6. T.N.</td>
<td>Ostium primum mitral cleft, tricuspid cleft</td>
<td>14</td>
<td>3</td>
<td>1:1 (3.0:1)</td>
<td>27/5 (15/8)</td>
</tr>
<tr>
<td>7. D.P.</td>
<td>Ostium primum mitral cleft, tricuspid cleft deficiency</td>
<td>8</td>
<td>1½</td>
<td>1:1 (2.0:1)</td>
<td>24/6 (20/7)</td>
</tr>
<tr>
<td>8. N.M.</td>
<td>Ostium primum mitral cleft, sub-aortic obstruction</td>
<td>11</td>
<td>1</td>
<td>1:1:1 (2.0:1)</td>
<td>40/25 (74/36)</td>
</tr>
</tbody>
</table>

* Preoperative figures in parentheses.

Abbreviations: Q_P/S = pulmonary to systemic flow ratio; PA = pulmonary artery; PC = mean pulmonary capillary pressure; LA = mean left atrial pressure; RA = mean right atrial pressure; LV = left ventricular pressure; PO = postoperative.
MITRAL VALVE FUNCTION

communication, was normal (<12 mm Hg) in all patients.

Biplane angiocardiograms were made on five patients with selective injection into the left ventricle via the venous route and mitral valve. Regurgitation along the catheter was seen in each case during ventricular systole, with the contrast material immediately crossing the atrial septal defect and opacifying portions of right atrium and right ventricle. It was not possible to determine how much of this mitral regurgitation was artificially induced by the catheter.

Operative Findings

The spectrum of ostium primum defect and cleft mitral valve lesions was noted as follows (Table 1): common atrium with anomalous left superior vena cava, absent coronary sinus and cleft mitral valve in one patient (case 1); ostium primum defect and cleft mitral valve in three patients (cases 2 to 4); ostium primum defect, cleft mitral valve, and deficiency of the tricuspid valve or cleft in its septal leaflet in three (cases 5 to 7); and ostium primum defect, cleft mitral valve, and interventricular communication (probe patency) lying beneath the valve ring at the level of the cleft in one patient (case 8).

Mitrail incompetence was determined by palpation of the regurgitant jet before repair of the mitral valve in six patients and was described as "slight" or "mild" (Table 2).

Surgical Technic

After the establishment of cardiopulmonary bypass, repair of the mitral valve deformity consisted of suturing the cleft in the anterior leaflet regardless of the ability of the surgeon to detect mitral regurgitation at the operating table. The subjacent chordae tendineae were not resected. When a cleft was evident in the septal leaflet of the tricuspid valve, it was closed. The ostium primum defect in the atrial septum was reconstituted with a Teflon prosthesis.

In the patient with a common atrium, a tunnel was created by utilizing a crimped Teflon prosthesis which redirected the coronary sinus and left superior vena caval flow into the area of the right atrium. The mitral cleft and ostium primum defect were repaired in the usual manner.

Postoperative Clinical Assessment

All patients were clinically well and asymptomatic; none were receiving digitalis or other medications.

Heart size on chest x-rays had decreased in all patients and was normal or near normal except in the patient with residual subaortic obstruction (case 8). Left atrial enlargement was not visible.

Apical systolic murmurs, grade II to IV/VI, were present in all patients. They first became apparent in four patients following surgery, and in the other four, they remained of the same intensity or became louder (Table 2). The murmur was pansystolic in three of the eight patients. Other auscultatory findings included short, left basilar ejection systolic murmurs; soft, rough, systolic and mid-diastolic murmurs at the lower left sternal border; and in one patient (case 8) a harsh ejection systolic murmur at the right base became more conspicuous.

Results of Postoperative Cardiovascular Studies (Figs. 1 to 7)

Intracardiac and Pulmonary Pressures and Flows (Table 1)

In seven patients (cases 1 to 7) right and left heart pressures were normal, and none had

Table 2

<table>
<thead>
<tr>
<th>Patient</th>
<th>Apical systolic murmur*</th>
<th>Postoperative LV angiogram</th>
</tr>
</thead>
<tbody>
<tr>
<td>MR at operation</td>
<td>Preoperative</td>
<td>Postoperative</td>
</tr>
<tr>
<td>Total</td>
<td>6/8</td>
<td>4/8</td>
</tr>
</tbody>
</table>

*Intensity graded I to VI; (P) = pansystolic; MR = mitral regurgitation.
Patient 6. Typical postoperative frontal left ventriculogram of a patient with ostium primum defect and a complete cleft in the anterior leaflet of the mitral valve. (A) In systole. (B) In diastole. In systole note the mildly irregular contour of the right medial border of the left ventricle and the transverse linear defect (arrow) which is related to the thickened, coapted edges of the cleft anterior leaflet, demarcating its superior and inferior segments. In ventricular systole, no mitral regurgitation is evident on the frontal or lateral films. In diastole, the left ventricular outflow tract appears to be of normal width.

Figure 2

Patient 5. Preoperative and postoperative angiocardograms in ventricular systole. (A) Before operation. Note the deformed upper medial border of the left ventricle and some regurgitation of contrast material along the catheter which has been passed from the right atrium through the ostium primum defect and mitral valve. (B) After operation. Persistent irregularity of the medial border of the left ventricle is seen. There is no evidence of mitral regurgitation.
an intracardiac shunt. In the six patients in whom cardiac output was determined (cases 2 to 7), it was normal at rest and rose in a normal fashion during exercise without significant change in intracardiac or pulmonary vascular pressures. The simultaneously recorded left ventricular and pulmonary capillary pressure curves were of normal contour and failed to show any evidence of mitral regurgitation or obstruction.

Hemodynamic abnormalities were detected on postoperative right and left heart catheterization in one patient (case 8). A systolic pressure gradient of 75 mm Hg was recorded postoperatively on withdrawal of the catheter from the left ventricle to the aorta, a finding compatible with subaortic obstruction (fig. 6). The preoperative gradient of 26 mm Hg between the left ventricular and brachial systolic pressures was not appreciated at the time. On postoperative study of the right heart, a small residual left-to-right shunt (Q P:S = 1.1:1) was detected at the atrial level. Pulmonary artery pressure was elevated but significantly decreased from preoperative level.

**Figure 3**

*Patient 5. Preoperative and postoperative angiocardiograms in ventricular diastole. (A) Before operation. The typical narrow left ventricular outflow tract, produced by the open, superior segment of the cleft anterior leaflet of the mitral valve, is seen (arrow); the catheter has slipped out of the left ventricle. (B) After operation. The outflow tract appears to be wider as the non-opaque blood entering the left ventricle is now directed more caudally with closure of the mitral cleft.*

**Left Ventricular Angiocardiograms**

The frontal left ventricular angiocardiograms in each patient showed persistence of the deformity of the outflow tract in systole. The characteristic irregular contour of the medial border of the left ventricle results from the abnormal attachments of the cleft anterior leaflet of the mitral valve to the upper portion of the muscular ventricular septum. This left ventricular deformity in systole is shown in three patients in figures 1A, 2, and 4. Postoperative angiocardiograms in diastole showed some variation: In one patient (fig. 5B) residual narrowing of the horizontally positioned outflow tract is evident, while in two other patients, a relatively wider, more normal appearance of the outflow tract can be seen (figs. 1B and 3B).

Mitril regurgitation could not be detected postoperatively in frontal and lateral ventriculograms in the group of patients (cases 1 to 7) who had normal hemodynamics. In the only patient (case 8) who had mitral incompetence, which was slight, localized subaortic stenosis was clearly demonstrated (figs. 6 and 7).
Figure 4

Patient 3. Preoperative and postoperative angiograms in ventricular systole. The preoperative film (A) with the catheter passing into the left ventricle through the ostium primum defect shows the characteristic irregularity of the medial wall of the left ventricular outflow tract. Opacification of the right heart is secondary to mitral regurgitation and left-to-right shunt through the ostium primum defect. The postoperative selective retrograde left ventriculogram (B) shows persistent deformity of the medial wall of the left ventricular outflow tract. There is no mitral regurgitation.

Discussion

Apical systolic murmurs were consistently noted postoperatively in this small series of patients with ostium primum defect and cleft mitral valve and were usually more intense than those recorded preoperatively. In the seven patients who had normal mean pulmonary capillary pressure tracings and no evidence of regurgitation of contrast material, the murmur could not be attributed to mitral incompetence. The origin of the apical systolic murmurs and their relationship to the repaired anterior mitral leaflet and intrinsic deformity of the left ventricular outflow tract are not clear; this may be elucidated further by left ventricular cineangiography and phonocardiography.

The observation that there was no instance of major mitral regurgitation after operation may simply reflect that this group of patients with ostium primum defect had only mild, or no valvular incompetence preoperatively. It seems to indicate however, that the surgical technic of repairing the cleft of the anterior leaflet of the mitral valve without resection of the subjacent or accessory chordae tendineae usually results in competent valve function, as previously reported by Braunwald and Morrow.

The postoperative frontal left ventricular angiograms in all patients showed in systole a persistent deformity of the medial border of the left ventricle, with a concave or flat, serrated contour and a nonopaque indentation formed by the coapted margins of the cleft anterior mitral leaflet (figs. 1A, 2B, and 4B). No impressive change had occurred compared to the characteristic preoperative configuration of the left ventricle, originally described by Baron and associates in endocardial cushion defects. Rastelli and associates also noted persistence of this appearance on postoperative C. V. Vol. XL, July 1969
left ventriculograms in two cases following suture of the mitral cleft.

In diastole, however, the postoperative left ventricular angiograms frequently showed a change in the contour of the outflow tract visualized on preoperative study. That the characteristic "gooseneck" narrowing of the contrast material in the ventricular outflow region produced as the superior segment of the cleft anterior mitral leaflet is pushed upward by the less opaque blood entering the left ventricle was readily appreciated on preoperative angiograms (figs. 3A and 5A). On the postoperative frontal left ventricular angiograms this type of narrowing tended to be less in diastole and in some cases the region returned to a near normal appearance. Somerville and Jefferson have found that lack of mobility of the superior segment of the mitral leaflet on preoperative angiograms was accompanied by considerable anatomic fixation (from accessory valve tissue, chordae, or papillary muscle), difficulties in surgical repair, and a poor postoperative clinical result.

Subaortic obstruction associated with endocardial cushion defects, as previously discussed by Sellers, Lillehei and their associates probably occurred in one of our patients. The adherence of the cleft anterior leaflet of the mitral valve on the upper portion of the deficient ventricular septum often encroaches on the left ventricular outflow tract. Surgical repair of the cleft in some cases, particularly when the base of the leaflet is pulled closer to the ventricular septum, can further narrow the left ventricular outflow tract and produce obstruction. The possibility however that a separate entity of "discrete subaortic stenosis," as reported in one case by Braunwald and Morrow, was the major abnormality in our patient 8 must be considered since the preoperative study indicated

Figure 5

Patient 3. Preoperative and postoperative angiocardiograms in ventricular diastole. The preoperative film (A) made 1/6 sec following figure 4A, shows characteristic "gooseneck" narrowing of left ventricular outflow tract. This is produced by the open, superior segment (arrow) of the cleft anterior mitral valve leaflet which separates the contrast material in the superior portion of the left ventricular outflow tract from the nonopaque blood entering the ventricle in diastole. The postoperative film (B), made 1/6 sec following figure 4B, shows residual diastolic deformity of the left ventricular outflow tract.
Figure 6

Patient 8. Subaortic obstruction. Postoperative withdrawal pressure tracing from the body of the left ventricle to the aorta indicating a systolic gradient of 75 mm Hg across the outflow tract.

Figure 7

Patient 8. Postoperative left ventricular angiograms. (A) Lateral view in systole shows discrete local narrowing in the outflow tract (arrow). A tiny jet of contrast material is seen entering the left atrium from the base of the mitral cleft and a small amount of extravasated contrast material is noted in the posterior wall of the left ventricle. (B) Simultaneous frontal film in systole shows the site of subaortic stenosis (arrow), the characteristic deformity of the medial border of the left ventricle, and slight mitral regurgitation. (C) In ventricular diastole (3/5 sec after B) nonopaque blood enters the left ventricle; note the fixed configuration of the localized narrowed region in the left ventricular outflow tract.

a gradient of 26 mm Hg between the left ventricle and brachial artery. The postoperative study of this patient clearly delineates subaortic stenosis (figs. 6 and 7) and suggests that the left ventricular outflow obstruction was intensified by surgical repair of the mitral valve with
a resulting gradient of 75 mm Hg. One should be alerted to the complicating abnormality of subaortic stenosis in carefully assessing pre-operative left ventricular hemodynamic and angiographic findings; it may not be readily apparent at the time of surgical exploration.

References

In considering the history of ideas, I maintain that the notion of 'mere knowledge' is a high abstraction which we should dismiss from our minds. Knowledge is always accompanied with accessories of emotion and purpose.—ALFRED NORTH WHITEHEAD: Adventures of Ideas. New York, The Macmillan Co., 1933, p. 5.
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