Case Report

CONGENITAL MITRAL INSUFFICIENCY ASSOCIATED WITH VENTRICULAR SEPTAL DEFECT: THE REPORT OF A SUCCESSFULLY CORRECTED CASE

Atoh Masuda, Akira Nonoyama, Sumio Kotani, Masaaki Nakahashi, Tetsuhide Ishii, and Terumasa Kagawa

Ventricular septal defect associated with mitral valve disease is uncommon except in cases of corrected transposition of the great vessels and of persistent common atioventricular canal, and the reported corrections of this anomaly-complex are few.\(^1\)\(^2\)

Reported here is a surgically corrected case of congenital mitral insufficiency and ventricular septal defect associated with a bidirectional shunt in which the right to left shunt was dominant.

Case Report

A four-year-old boy was admitted to the thoracic surgery department of the Kansai Medical School Hospital in November, 1970. He was suffering from easy fatiguability, shortness of breath, and restricted activity associated with retarded physical development. Pregnancy and delivery had been normal, but on his 40th day, cardiac murmur had been detected and his subsequent history revealed frequent respiratory infection. Restricted in activity he could not walk continuously any long distance, although he was no squatter. Shortly before admission, cyanosis with shortness of breath had become apparent even at rest. He had no history of rheumatic fever.

Physical examination revealed a slightly cyanotic boy whose body weight was 15.5 kg, whose heart rate was 120 per minute, and whose blood pressure was 92/70 mmHg. A remarkable precordial bulge was demonstrated. The apical beat was strong, but the thrill was not palpable. At auscultation, a grade 3 of 6 holosystolic murmur in the 4th left sternal border and a grade 5 of 6 holosystolic murmur from apex to left axilla and left upper back were heard, and the pulmonic second sound was accentuated. The liver was not palpable and no edema was demonstrated. Laboratory examination were as follows: RBC 420 x 10\(^4\), WBC 7600, hemoglobin contents 12.6 g/dl, total serum protein 6.0 g/dl, A/G ratio 1.70, RA and CRP negative, ASLO 12 Todds. No abnormalities were evident in the liver- and renalfunction tests.

The chest X-ray showed a grossly enlarged heart (the CTR was 70%), and an increased pulmonary vasculature over both lung fields (Fig.1a). The electrocardiogram (Fig.2a) showed normal sinus rhythm, right axis deviation to 90 degrees, and a pattern of biventricular hypertrophy with left atrial overloading. The vectorcardiographic finding was the same (Fig.2b).

Right heart catheterization indicated, as shown in Table I, a bidirectional shunt with a dominant right to left shunt and marked pulmonary hypertension with a systolic pressure of 102 mmHg and a mean pressure of 90 mmHg. The pulmonary wedge pressure could not be estimated.

Key Words: Congenital Mitral Insufficiency Ventricular Septal Defect Mitral Valve Replacement

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The pulmonary-systemic resistance ratio was 1.3 and the pulmonary-systemic pressure ratio was 0.9. The arterial oxygen saturation had decreased to 88%.

Dye injected into the left ventricle was immediately reflected in the left atrium and a few opacifications were demonstrated in the right ventricle and the pulmonary artery (Fig. 3).

The diagnosis was made of mitral insufficiency associated with ventricular septal defect and severe pulmonary hypertension. The pulmonary hypertension of the present case was suspected to be causally related with both the mitral incompetence and the ventricular septal defect, although the mitral factor was considered dominant. From the judgement that cure would be possible if both lesions could be corrected simultaneously, the patient was operated upon November 30, 1970.

The heart was approached through a median sternotomy and the cardiopulmonary bypass was established using a TempTrac oxygenator under moderate hypothermia. The right ventricle was opened and a large defect of the membranous septum was closed with a Dacron patch. After this, left atriotomy was done. In the mitral valve, both cusps were thick and shortened without fusion of the commissures, and the mobility of cusps was very poor. Since valvuloplasty or annuloplasty was not indicated, the cusps were resected and valve replacement was done. The anterior papillary muscle was very large, and both cusps were inserted directly to the anterior papillary muscle without chordae. The connection between both cusps and the posterior papillary muscle was almost the same, but the posterior cusp was connected to the posterior papillary muscle through thick and short chordae. Fig. 4 shows the resected valve with the papillary muscles. The cusps were hypertrophic and smooth without verrucae and calcification.

The mitral valve was replaced with a Björk-Shiley prosthetic valve of 27 mm in external diameter and, to make sure, the foramen ovale was opened after right atriotomy. The form and function of the tricuspid valve were normal. After the incisions in the right ventricle and both atria were closed, the patient was taken off cardiopulmonary bypass. The perfusion time was 184 minutes. The systolic right ventricular pressure was 80 mmHg immediately after bypass as compared with systemic arterial pressure of 110 mmHg. The right atrial mean pressure was 13.4 mmHg, and the left atrial mean pressure was 6.0 mmHg.

Although the patient was assisted by mechanical respiration for ten days after surgery, his recovery was uneventful. His excellent condition.

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Fig. 2. Preoperative electrocardiogram (a) and vectorcardiogram (b) showing right axis deviation and biventricular hypertrophy with left atrial overloading.

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TABLE 1  RIGHT HEART CATHETERIZATION

<table>
<thead>
<tr>
<th>Site</th>
<th>Before operation</th>
<th>After operation</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Pressure (mmHg)</td>
<td>O₂ Sat. (%)</td>
</tr>
<tr>
<td></td>
<td>(systolic/diastolic)</td>
<td></td>
</tr>
<tr>
<td>PC</td>
<td>102/82 (90)</td>
<td>65.0</td>
</tr>
<tr>
<td>PA</td>
<td>92/20</td>
<td>66.5</td>
</tr>
<tr>
<td>RV</td>
<td>(5.5)</td>
<td>61.5</td>
</tr>
<tr>
<td>RA</td>
<td>(5)</td>
<td>59.5</td>
</tr>
<tr>
<td>SVC</td>
<td>(5.5)</td>
<td>63.0</td>
</tr>
<tr>
<td>IVC</td>
<td>114/70</td>
<td>88.0</td>
</tr>
</tbody>
</table>


Fig.3. Preoperative levocardiogram. Catheter tip was placed in the left ventricle.

at the time of his discharge on the 46th post-operative day has continued up to now, one year after the operation. Anticoagulant therapy has been continued since the first postoperative week.

Right heart catheterization on the 45th day after surgery showed a decrease in pulmonary arterial pressure (Table I), the systolic being 62 mmHg, diastolic being 32 mmHg, mean pressure being 50 mmHg, and the wedge pressure being 32 mmHg.

Chest X-ray one year after surgery showed a decreased cardiac shadow (the CTR was 52%) with an almost normal pulmonary vasculature.

TABLE II   MAIN TYPES OF CONGENITAL MITRAL VALVE DISEASE ASSOCIATED WITH VENTRICULAR SEPTAL DEFECT

<table>
<thead>
<tr>
<th>Author</th>
<th>Valve lesion</th>
<th>Types of anomaly</th>
<th>Operations</th>
</tr>
</thead>
<tbody>
<tr>
<td>Edwards² (1958)</td>
<td>MI</td>
<td>Accessory commissures in the posterior cusp</td>
<td>None</td>
</tr>
<tr>
<td>Neufeld⁶ (1961)</td>
<td>MI</td>
<td>Cleft anterior cusp</td>
<td>None</td>
</tr>
<tr>
<td>Edwards⁵</td>
<td>MI</td>
<td>Ectopic, low insertion of the chordae</td>
<td></td>
</tr>
<tr>
<td>Gilbert⁷ (1964)</td>
<td>MI</td>
<td>Cleft anterior cusp</td>
<td>Closure of VSD</td>
</tr>
<tr>
<td>Hollman¹ (1965)</td>
<td>MS</td>
<td>Supravalvular ring</td>
<td>Closure of VSD, incision of ring</td>
</tr>
<tr>
<td>Flage² (1967)</td>
<td>MI</td>
<td>Lack of chordal support of septal cusp</td>
<td>Closure of VSD, suture of flailing edge of cusp to adjacent chordae</td>
</tr>
<tr>
<td>Messmer⁸ (1970)</td>
<td>MI</td>
<td>Cleft septal cusp</td>
<td>Plastic repair</td>
</tr>
<tr>
<td>Author's case</td>
<td>MI</td>
<td>Anomalous short or absent chordae</td>
<td>Closure of VSD, prothetic valve replacement</td>
</tr>
</tbody>
</table>

(Fig.1b).

Histologically, the cusps revealed a slight cell infiltration and an increase in connective tissue, but no inflammatory change was evident. The mitral valve change seen in the present case was supposed to be a type of congenital anomaly corresponding to the anomalous short or absent chordae described by Talner (1961).³

DISCUSSION

Ventricular septal defect associated with mitral valve disease is uncommon except in cases of

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corrected transposition of the great vessels and of persistent common atrioventricular canal, and the reported corrections of this anomaly-complex are few. Particularly rare is ventricular septal defect associated with congenital mitral valve deformity. The literature on cases of ventricular septal defect accompanied by distinct congenital mitral valve disease as reported by Edwards, Neufeld, Gilbert, Hollman, Flege, Messner have been summarized in Table II. Hollman and his colleagues also reported on seven other cases of mitral valve disease accompanied ventricular septal defect. The eight cases reported by Hollman corresponding to 3.6% of the 221 cases of ventricular septal defect in his series consisted of four mitral stenosis, three mitral insufficiency, and one stenoininsufficiency, but their anatomical details were not described except for one mitral stenosis case due to supravalvular ring and one mitral insufficiency case due to short chordae. Simultaneous surgical repair of both lesions had been performed in the former case only.

The probable mechanisms combining ventricular septal defect with mitral valvular deformation are: 1) casual combination, 2) dilated mitral annulus due to hypertrophy and dilatation of the left ventricle, 3) valvular sclerosis due to endocardial fibroelastosis (secondary type), 4) endocardial cushion defect without atrial septal defect, and 5) congenital anomaly as a complex. The high incidence of bacterial endocarditis in ventricular septal defect has been noted, but the complication of rheumatic endocarditis in ventricular septal defect has not been emphasized.

Three years ago, the authors treated a 18-year old girl with ventricular septal defect and mitral stenoininsufficiency by means of closure of the septal defect and prosthetic valve replacement. In this case, the combining of ventricular septal defect and rheumatic mitral valve disease was suspected to be accidental.

Mitril regurgitation by secondary annular dilatation due to hypertrophy and dilatation of the left ventricle was considerably demonstrated subclinically. The authors recognized two such cases out of 20 ventricular septal defects with pulmonary hypertension performed selective cinelevocardiography. However, the regurgitation occurring in this type usually disappears after closure of the septal defect.

The lesion in the present case could have occurred either by casual combination or combination as a complex; however, no similar case has yet been reported. The mitral valve change in our case corresponded to that of anomalous short or absent chordae described by Talner (1961). The generating mechanism of the present anomaly was suggested by the report of Layman and Edwards (1969) whose cases were due to hypoplasia of cusps and chordae; however, the lesions in their cases were not accompanied by ventricular septal defect.

On the subject of the surgical treatment of mitral insufficiency associated with ventricular septal defect, Hollman emphasized, since the severity of ventricular septal defect would be underestimated that the surgical priority should be given to this defect, because mitral valve disease and ventricular septal defect would interfere with each other hemodynamically and so the left to right shunt would be smaller as compared with ventricular septal defect without complication. On the other hand, the presence of pulmonary venous hypertension due to the mitral valve disease might be confused with pulmonary hypertension due to ventricular septal defect and the operative indication might misapply. Therefore, measurements of left atrial pressure and pulmonary circulation time should be made and selective levocardiography should be performed before surgery, and the character and severity of the mitral valve disease should be appreciated as accurately as possible, and, if necessary, a simultaneous repair of both lesions should be performed.

With regard to the operation against mitral insufficiency in childhood, valvuloplasty or annuloplasty has been advocated by many investigators. However, the results of these operations is not always well. Flege and his colleagues reported that only 6 out of 13 patients having undergone valvulo- or annuloplasty were hemodynamically satisfactory after surgery. The results of valvulo- or annuloplasty by Levy and his colleagues were not favorable as well. The real character of congenital mitral valve malformation is so complicated that it is reasonable that the satisfactory repair is not accomplished by means of the plastic surgery which could only depend upon the findings made during operation.

On the other hand, prosthetic valve replacement can not regard as an ideal treatment because of the questionable durability of the artificial valve, the possibility of narrowing in the orifice area after growth, and the necessity of long postoperative anticoagulant therapy. However, valve replacement in severe cases like the present one is a rescue method. The question of orifice...
narrowing after growth is irrelevant here, because a dilated annulus is not uncommon even in childhood and an adult-sized valve is usually used. In the present case, the adult sized Björk-Shiley valve (27 mm in outside diameter) was easily used. A low profile valve is desirable because it does not reduce left ventricular volume. Although Bloodwell and his colleagues\textsuperscript{11} have used the caged discoid valve, a tilting valve like that of Björk-Shiley\textsuperscript{12} or Wada-Cutter seems to be favorable from hemodynamical point of view.

**SUMMARY**

Presented here is the successfully corrected case of a four-year-old boy with congenital mitral insufficiency and ventricular septal defect associated with pulmonary hypertension. The mechanism combining both lesions and the surgical treatment for mitral insufficiency in childhood are also discussed.

**REFERENCES**