Surgical Indications and Results for Congenital Cardiac Anomalies associated with Left to Right Shunt and Severe Pulmonary Hypertension

AKIRA SHOHTSU, SHIGEYUKI TAKEUCHI, AND TADASHI INOUE

This paper was designed to assess the surgical indications and results for ventricular septal defect with the ratio of pulmonary to systemic artery pressure (Pp/Ps) greater than 0.8, patent ductus arteriosus with Pp/Ps greater than 0.8 and atrial septal defect with pulmonary artery systolic pressure greater than 60 mmHg.

Ventricular Septal Defect

It is our principle to perform elective closure of ventricular septal defect (VSD) with Pp/Ps greater than 0.8 between 1 and 3 years of age under slightly hypothermic cardiopulmonary bypass, however, in the patients less than one year, profound hypothermia induced with surface cooling combined with cardiopulmonary bypass was used. The youngest patient of closure of VSD was 4 months of age and the oldest was 18 years.

Prior to 1970, 35 patients were subjected to primary closure of the defect, resulting in 11 operative deaths. Since 1971, however, the surgical results have been much improved and 29 such patients were operated on without death (Table I). Analysis of clinical data would clarify the reason of this improvement in our surgical results and possible difference in the severity of the patients between the two groups.

1) The ratio of pulmonary to systemic resistance (Rp/Rs).

Table I

<table>
<thead>
<tr>
<th></th>
<th>Rp/Rs &lt; 0.75</th>
<th>Rp/Rs &gt; 0.75</th>
</tr>
</thead>
<tbody>
<tr>
<td>1970</td>
<td>25 (6)</td>
<td>10 (5)</td>
</tr>
<tr>
<td>1971</td>
<td>26 (0)</td>
<td>3 (0)</td>
</tr>
</tbody>
</table>

Pp/Ps, the ratio of pulmonary to systemic artery pressure
Rp/Rs, the ratio of pulmonary to systemic resistance

Until 1970, 6 of 26 patients with Rp/Rs of less than 0.75 died postoperatively, as did 5 of 10 patients with Rp/Rs of 0.75 or more. Since 1971, however, there has been no death in both 26 patients with Rp/Rs of less than 0.75 and 3 patients with Rp/Rs of 0.75 or more (Table I).

2) Pure oxygen inhalation test.

Reduction in pulmonary arterial systolic pressure was detected after breathing pure oxygen for 5 minutes in the patients with Pp/Ps of 0.8 or more (Fig. 1).

Until 1970, there were 6 deaths in 10 patients whose decrease in pulmonary arterial systolic pressure remained less than 20%, while only 5 death occurred in the remaining 25 patients who demonstrated decrease of more than 20%. Since 1971, however, no death occurred even in the 16 patients whose decrease in pressure was less than 20%, including 6 patients who demonstrated no decrease at all.

3) Cardiac murmur.

Until 1970, 3 of 25 patients with apical dia-

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TABLE II SURGICAL RESULTS ACCORDING TO TYPE OF VENTRICULAR SEPTAL DEFECTS

<table>
<thead>
<tr>
<th></th>
<th>Type I</th>
<th>Type II</th>
<th>Type III</th>
</tr>
</thead>
<tbody>
<tr>
<td>~ 1970</td>
<td>9 (0)</td>
<td>19 (6)</td>
<td>7 (5)</td>
</tr>
<tr>
<td>1971 ~</td>
<td>3 (0)</td>
<td>18 (0)</td>
<td>8 (0)</td>
</tr>
</tbody>
</table>

(Type I, II and III are according to the classification by Kirklin et al.3)

TABLE III SURGICAL RESULTS OF PATENT DUCTUS ARTERIOSUS WITH Pp/Ps RATIO GREATER THAN 0.8

<table>
<thead>
<tr>
<th>Rp/Rs</th>
<th>Isolated PDA</th>
<th>PDA with VSD*</th>
</tr>
</thead>
<tbody>
<tr>
<td>&lt; 0.75</td>
<td>12 (0)</td>
<td>20 (3)</td>
</tr>
<tr>
<td>≥ 0.75</td>
<td>3 (0)</td>
<td>0</td>
</tr>
</tbody>
</table>

* Closure of PDA or closure of PDA with pulmonary artery banding

Toxic rumble died postoperatively, as did 8 of 10 patients without it. Therefore, decreasing tendency of diastolic rumble on periodical observation since infancy compelled us to perform an earlier operation. Since 1971, it was only two patients who were subjected to operation without diastolic rumble, but both have been doing well.

4) Chest x-ray findings.

Until 1970, 8 patients died out of 18 operative patients who had not distinct enlargement of the left ventricle on the chest x-ray films, while only 2 deaths occurred in the 17 patients with it. Therefore, an earlier operation has been performed in the patients with decreasing tendency of the cardiac shadow in the patients with VSD and severe pulmonary hypertension. Since 1971, it was only 4 patients who were subjected to operation without significant enlargement of the left ventricle on the chest x-ray. There has been no surgical mortality. Twenty five patients with the left ventricular enlargement also have been successfully operated on.

5) Location of the defect.

The location of defects were classified into three types according to Kirklin2 (Table II). Until 1970, there were no death in 9 patients of type I, 6 deaths in 12 of type II and 5 deaths in 7 of type III. Therefore, transatrial approach has been adopted for closure of defects of type II and type III in order to preserve postoperative right ventricular function. Since 1971, 3 patients of type I, 18 of type II and 8 of type III have been operated on successfully, including one patient who has an endocardial cushion defect type VSD associated with multiple VSDs and three patients with mitral insufficiency.

PATENT DUCTUS ARTERIOSUS

Surgical results of patent ductus arteriosus (PDA) with Pp/Ps of 0.8 or more are shown in Table III. No patients with isolated PDA died even in the cases of Rp/Rs greater than 0.75.

Three deaths occurred in 20 patients of PDA with VSD, in whom closure of PDA and pulmonary artery banding were performed.

**Atrial Septal Defect**

Cardiac catheterization data before and after closure of atrial septal defect (ASD) with pulmonary hypertension in aged three patients are shown in Table IV. Postoperative pulmonary artery pressure decreased and clinical symptoms improved when the ratio of pulmonary to systemic flow (Qp/Qs) was more than 1.5. In a patient, who was 42 years old with Qp/Qs of 1.2, pulmonary artery pressure and Rp/Rs had markedly increased postoperatively.

**Discussion**

Until 1968, operative mortality of VSD with Pp/Ps of 0.8 or more had been higher as compared with that of Pp/Ps less than 0.8. Hallman and Cooley reported 26% mortality, while Cartmill and Kirklin (Pp/Ps > 0.75) 17%. Recently, however, much improvement has been generally observed. Though our operative mortality in the patients with Pp/Ps of 0.8 or more had been 31% prior to 1970, there has been no death since 1971.

With increasing experiences, it became apparent to what symptoms particular attention should be paid on periodical observation of the patients with severe pulmonary hypertension in order to decide the optimal time for closure of VSD. Earlier operations were performed in the patients of VSDs with severe pulmonary hypertension, when the left ventricular enlargement was less remarkable than expected or decreasing tendency was considered to be present in left ventricular enlargement or apical diastolic rumble. This is considered to be one of the important factors which brought the improvement in our operative results since 1971.

**Cartmill and Kirklin** reported that the operative results were unsatisfactory in the patients with Rp/Rs of 0.75 or more. Prior to 1970, our operative results were also disappointing in the patients with Rp/Rs of 0.75 or more. The mortality rate was 50%. Since 1971, however, even if catheterization revealed Rp/Rs of 0.75 or more, closure of VSD would safely be performed especially in younger patients when the left to

![Diagram](attachment:image)

**Fig.2.** Distribution of patients with ventricular septal defect under the age of one year seen at Pediatric Clinic of Keio University Hospital. (1971–1973)
right shunt was considered to be dominant from auscultatory and x-ray findings.

Prior to 1970, per cent degree of decrease in pulmonary arterial systolic pressure after pure oxygen inhalation test was considered to be very useful for prediction of operative risk. At present, however, the role of this test seems questionable for us, because after 1971, no death occurred even in the patients who did not show any reduction in pulmonary arterial pressure at all.

During three years from 1971 to 1973, 247 patients with VSD less than one year of age visited the pediatric clinic of Keio University Hospital (Fig. 2). Forty-nine patients required medical treatment because of heart failure, 40 of whom were well controlled medically and are waiting for the optimal time of surgery. Only one death occurred during medical treatment under one year of age. In 8 patients, operation was considered to be mandatory under one year of age because of intractable heart failure. For these infants, our previous policy was to attempt pulmonary artery banding, however, 3 patients of 1 to 3 months of age died postoperatively. Recently, primary closure of the defects has become the procedure of choice with the introduction of profound hypothermia and limited cardiopulmonary bypass even under 6 months of age. The youngest patient of successful closure of VSD was 4 months of age.

We suppose that left ventriculogram and aortogram are mandatory to determine surgical indications for patients of PDA and VSD with severe pulmonary hypertension. Our current policy is to attempt primary repair of VSD and PDA even less than one year of age when the shunt through VSD seems to be large enough to cause pulmonary hypertension.

Summary

Prior to 1970, 35 patients of VSD with Pp/Ps of 0.8 or more were subjected to primary closure of the defect, resulting in 11 operative deaths. Since 1971, however, the surgical results have been much improved and 29 such patients were operated on without death.

It has been considered the optimal time of elective closure of VSD with severe pulmonary hypertension is at the age of 1 to 3 years, however, if decrease in apical diastolic rumble and heart size are found, earlier operation less than one year of age is to be scheduled.

Surgical indications and results of PDA, PDA with VSD, and ASD associated with severe pulmonary hypertension were also discussed.

REFERENCES