Results of Surgical Treatment of Ventricular Septal Defect Associated with Pulmonary Hypertension under the Age of Two

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SUMMARY

Various operative treatments for ventricular septal defect below the age of 2 years accompanied by pulmonary hypertension were compared with reference to the operative results, PA and Rp/Rs.

Although it is ideal to carry out a primary repair before pulmonary vascular changes advance, pulmonary artery banding should be performed as soon as possible if primary repair is impossible to prevent the progress of further pulmonary vascular changes.

In some cases with a PA of 70 mmHg and an Rp/Rs around 0.5, despite adequate primary repair, postoperative persistence of pulmonary vascular change was inferred.

A rise of PA and Rp/Rs appeared to be significant as the index of progressive pulmonary lesions.

Additional Indexing Words:
Pulmonary artery banding Primary repair Two stage operation

Since 1964, various surgical treatments were given in our Department for ventricular septal defect with marked pulmonary hypertension and repeated cardiac failure in children below the age of 2. In the first half of the 8 year period up to July 1972, pulmonary artery banding mainly was carried out; while a two stage operation and primary repair were conducted under extracorporeal circulation in the second half.

Results of surgery varied depending upon the method of operation and the degree of pulmonary vascular changes. As an index for pulmonary vascular changes, the pulmonary to systemic resistance ratio (Rp/Rs) has been used; and recently, mean pulmonary arterial pressure (PA) was found to reflect the changes better. Rp/Rs and PA were therefore used to study the relationship between various operative methods and results in an attempt to evaluate the advantage and disadvantage of these methods.

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MATERIALS AND METHODS

Pulmonary artery banding:
There were 27 male and 24 female patients, a total of 51 in the series. Ages ranged 3/12 to 28/12 (average of 9/12) years, and body weight ranged between 3.9 and 11.5 Kg (average of 6.2 Kg).

In the operation, a left thoracotomy or midsternal incision was followed by the gradual, careful banding of the main pulmonary artery up to approximately 1/2 of its diameter with a teflon tape of 0.8 cm in width, taking special note of the cardiac beat. In many cases, blood pressure rose and thrill increased in the pulmonary artery peripheral to the site of banding. Upon banding of the pulmonary artery, care was taken not to constrict it below the diameter of aorta. Death occurred in 15 cases, giving the operative death rate of 29%.

Two stage operation:
There were 11 male and 9 female patients in this series. Ages ranged from 3 to 6 years (average of 54/12 years). Radical operation was carried out 3 years 9 months later on the average after pulmonary artery banding. Body weight ranged from 10.7 to 18.5 Kg (average of 15.8 Kg). In many cases, improvements such as body weight increase and subsidence of cardiac failure were noted.

Operation consisted of a midsternal incision followed by abrasion of various degree of adhesions. Tape was applied to the ascending aorta and vena cava superior and inferior. Extracorporeal circulation was carried out under mild hypothermia of 30°C by esophageal temperature. Closure of the ventricular septal defect and debanding of the pulmonary artery was then carried out. In most cases, the tape was easily removed but the posterior portion of the pulmonary artery was permitted to remain in some cases. Dilatation of the pulmonary artery after debanding was relatively easy and enlargement by the pericardial patch for the pulmonary artery was necessary in only 1 case.

Operative death occurred in only 1 case, giving the mortality rate of 5%.

Primary repair:
There were 6 male and 11 female patients in this series. Age ranged between 7/12 to 27/12 years (average of 17/12 years). Body weight ranged between 5.4 and 10.5 Kg (average of 7.7 Kg).

As in older patients, the operation was carried out under hemodiluted extracorporeal circulation with fresh ACD blood and Ringer-Lactate solution using bubble oxygenator under mild hypothermia (esophageal temperature, 30°C). The average duration of perfusion was 60 min. Death occurred in 3 cases, giving the rate of operative death of 18%. In 12 cases or 70%, a teflon patch was necessary for closure of the ventricular septal defect.

In these groups of patients, PA and Rp/Rs were calculated from the results of preoperative cardiac catheterization. Rp/Rs was calculated as total pulmonary artery resistance/total systemic resistance.

RESULTS

Pulmonary artery banding:
In 37 cases with satisfactory description of PA and Rp/Rs, operative
death was found to increase as $\overline{PA}$ and $Rp/Rs$ rose. On the other hand, death due to respiratory failure and cardiac failure was experienced even in cases with these values low (Fig. 1). This is probably related to the conservative nature of this operation and the addition of systolic pressure loading the presence of left to right shunt. It might also be explained by the fact that this method was frequently applied in cases of younger age group and poorer development than the primary repair. This method consequently gave a rather poor result, death in 8 (44%) of 18 cases with $\overline{PA}$ above 60 mmHg and $Rp/Rs$ above 0.25. Death also occurred in 4 (21%) of 19 cases with values less than these. This might indicate a limitation of this method and the need for special care in the postoperative period.

*Two stage operation*:  
In this group, $\overline{PA}$ before reoperation was decreased in 17 of 20 cases (85%) (Fig. 2). In 3 other cases (15%) mild rise was noted; but pulmonary artery banding was found to be effective.  
In the group with a fall in $\overline{PA}$, shrinkage of the defect was noted in 3 cases with favorable decrease following prebanding $\overline{PA}$ of about 45 mmHg.  
In 16 cases with satisfactory description of the $Rp/Rs$, a mild rise was noted in 3 (19%). In 13 others (81%), fluctuations within the normal range were always seen, and a fall was noted in many (Fig. 3). In 3 of these cases, the
The size of the VSD became smaller. Even in cases with a rise of Rp/Rs, banding was effectively carried out.

Both $PA$ and Rp/Rs rose in 1 of the 3 cases with a rise of $PA$ and 3 cases with the rise of Rp/Rs. In this case, respiratory failure developed postoperatively, and long term use of respirator and tracheotomy were required. Despite adequate banding, the influence of changes of pulmonary blood vessels probably modified the postoperative course in this case. Rise of $PA$ and Rp/Rs were significant.

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**Fig. 2.** $PA$ before and after pulmonary artery banding.

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**Fig. 3.** Rp/Rs before and after pulmonary artery banding.
Relationship between results of operation and $\bar{PA}$ and $Rp/Rs$ before reoperation was studied in 19 cases (Fig. 4). In 15 cases (79%), $\bar{PA}$ was 50 mmHg and $Rp/Rs$ less than 0.25. Even in 4 cases (21%) with values above these, the postoperative course was stable, except for a case described above.

This is probably due to the arrest of the progression of pulmonary vascular changes by banding, along with augmentation of physical development and resistance to operation before radical operation.

In the single fatal case, reoperation was carried out 6 years later. Thickening of the right ventricular outflow tract and right to left shunt was found and the patient died of low cardiac output syndrome postoperatively.

The time for reoperation appears to be crucial, and the operation after 3 to 4 years appears to be optimal.

As was stated before, sufficient postoperative control was especially necessary in cases with the rise of $\bar{PA}$ and $Rp/Rs$ after banding.

**Primary repair:**

Among these cases, 1 patient with a $\bar{PA}$ of 82 mmHg and $Rp/Rs$ of 0.8 was lost due to respiratory failure probably as the result of pulmonary vascular changes. In subsequent cases, however, none died of serious respiratory failure. However, 2 cases were lost in acute cardiac failure due to postoperative complete atrioventricular block (Fig. 5).

Consequently, the indications for operation should be carefully decided.
when the $\overline{PA}$ is above 80 mmHg and $Rp/Rs$ above 0.8. We are therefore conducting $O_2$ inhalation tests in cases with low flow, high resistance in preoperative cardiac catheterization and selecting cases with a $\overline{PA}$ above 70 mmHg and $Rp/Rs$ above 0.5 may stand the operation but tend to present problems postoperatively. Namely, in 1 case with a $\overline{PA}$ 73 mmHg and $Rp/Rs$ 0.57, cardiac failure occurred after discharge. A second cardiac catheterization revealed no change in these values without leakage. Congenital pulmonary vascular changes were suspected in this case.

Recently, all cases with transient atrioventricular block during operation are treated with pacemaker wire attached to the anterior wall of the right ventricle, to be connected with a demand pacemaker.

**DISCUSSION**

The possibility of operation for a ventricular septal defect accompanied by marked pulmonary hypertension is influenced by the degree of pulmonary vascular change. As the index, various hemodynamic data such as $Rp/Rs$ are used and the relationships with the operative results have been demonstrated.$^{2,3}$ $Rp/Rs$ is calculated as PAR/TSR$^1$ or TPR/TSR.$^2$ These changes readily occur as the influence of hypoxia and disturbance of acid base balance leading to an erroneous diagnosis. However, in view of the decision on oper-
ative indications and prognosis, the degree of pulmonary vascular change is important; and it is frequently difficult to evaluate these in clinical practice.

Hallidie-Smith\(^7\) is of the opinion that an overall evaluation is important based upon hemodynamic state, X-ray findings, and auscultatory findings, since histological diagnosis is certain but not generally available. Wagner et al\(^8\) pointed out the frequent occurrence of death in operated cases of large VSD with a $\overline{PA}$ above 65 mmHg and this is more closely related with prognosis than preoperative $Rp/Rs$ and flow rate. The $\overline{PA}$ is controlled by the amount of blood in the blood vessel and the pulmonary vascular bed. It is readily measured accurately and reflects the pulmonary vascular changes accurately.

Matsumoto\(^9\) studied vascular changes of the lung of VSD patients. A thickening of the media in the muscular-type pulmonary artery was pronounced in cases with rising systolic pressure of the pulmonary artery and $Rp/Rs$ but mild in cases with rising $Qp/Qs$.

In view of these results, we used $\overline{PA}$ and $Rp/Rs$ to study the relationship with operative results. Viz. the results were definitely poorer as both of these values rose in pulmonary artery banding and primary repair. In cases with elevation of one of these, results were not always poor. Consequently, decision on indication and prognosis using both the $\overline{PA}$ and the $Rp/Rs$ appears to be reasonable.

The following conclusions were drawn from our operative experience.

In pulmonary artery banding, a $\overline{PA}$ above 60 mmHg and an $Rp/Rs$ above 0.25 would in our hands result in a high surgical mortality of 44%. The pulmonary vascular changes apparently influence the operative results and care should be taken in postoperative control.

In the two stage operation, banding probably inhibits the progression of pulmonary vascular changes. In cases with a $\overline{PA}$ less than 50 mmHg and an $Rp/Rs$ less than 0.25, stable results are obtained. The best time of reoperation presents some problem, and it is desirable to perform reoperation within 4 years after banding. In small defects, the possibility of spontaneous closure is also present. In cases with persistently high $\overline{PA}$ values of 70 mmHg and $Rp/Rs$ of 0.45, despite adequate banding, pronounced respiratory insufficiency occurred at the time of reoperation; and the influence of pulmonary vascular changes markedly disturbed the postoperative course.

In primary repair, adequate postoperative control ensures radical healing without serious respiratory insufficiency, except for cases with marked pulmonary vascular changes ($\overline{PA}$ above 80 mmHg, $Rp/Rs$ above 0.8). In cases with a $\overline{PA}$ above 70 mmHg and $Rp/Rs$ above 0.5, problems are apt to remain postoperatively although the patient can stand operation.
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