SURGICAL RESULTS AND FACTORS AFFECTING OPERATIVE MORTALITY IN TOTAL ANOMALOUS PULMONARY VENOUS DRAINAGE

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Factors affecting the surgical results of total anomalous pulmonary venous drainage were evaluated in 17 patients in an attempt to establish an appropriate plan of management. The mortality rate correlated with age, size of interatrial communication, arterial oxygen saturation and left heart volume but not with pulmonary to systemic systolic pressure ratio, vascular resistance ratio and right ventricular volume. In cases where the symptoms appeared early in life, left heart volume was small and clinical features were severe. Four children with a left ventricular end-diastolic volume of less than 65% of normal died of low cardiac output syndrome. In those with an unusually small left heart, enlargement of the left atrium and/or delayed ligation of the anomalous vertical vein were considered more favorable. Left atrial and ventricular volumes were restored to normal after the surgery.

IN 1956, Burroughs and Kirklin reported the first complete repair of total anomalous pulmonary venous drainage (TAPVD). Though, the operative mortality and morbidity have decreased during the last 25 years, problems still remain. The present study was undertaken to assess the factors affecting the operative mortality and to establish a more appropriate plan of management.

Key Words:
TAPVD
Surgical result
Cardiac volume
Small left heart
Delayed ligation

MATERIALS AND METHODS

From 1969 to 1979, 17 children ranging in age from 3 months to 12 years were treated at the Department of the Cardiovascular Surgery of Kinki University and at Heart Institute of Amagasaki Hospital for repair of TAPVD. Drainage was of the supracardiac type in 11, cardiac type in 4 and mixed type in 2. There were 10 patients with TAPVD alone, 4 patients associated with patent ductus arteriosus (PDA), 2 with ventricular septal defect (VSD) and one with VSD plus PDA (Table I).

Cardiac catheterization and angiography were performed on all patients preoperatively.
TABLE I SEVENTEEN PATIENTS WITH TOTAL ANOMALOUS PULMONARY VENOUS DRAINAGE

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Age (months)</th>
<th>Drainage site</th>
<th>ASD</th>
<th>Associated anomalies</th>
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<td>13</td>
<td>LIV</td>
<td>-</td>
<td>-</td>
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<td>-</td>
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<td>43</td>
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<td>-</td>
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<tr>
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<tr>
<td>8*</td>
<td>9</td>
<td>SVC</td>
<td>L</td>
<td>-</td>
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<tr>
<td>9*</td>
<td>5</td>
<td>LIV + CS + SVC</td>
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<tr>
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<td>L</td>
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<td>L</td>
<td>PDA</td>
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<tr>
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<td>58</td>
<td>LIV</td>
<td>L</td>
<td>VSD + PDA</td>
</tr>
</tbody>
</table>

Abbreviations: LIV = left innominate vein, CS = coronary sinus, SVC = superior vena cava, L = large, S = small, PDA = patent ductus arteriosus, VSD = ventricular septal defect, * = Deaths

TABLE II CALCULATION OF CARDIAC VOLUME

\[
RVEDV = 0.65V \quad \text{Graham TP, 1973}
\]
\[
LVEDV = \begin{cases} 0.93V - 3.8 & \text{Dodge HT, 1960} \\ 0.97V - 3.1 (V > 15 \text{ cm}^3) & \text{Graham TP, 1971} \\ 0.73V (V < 15 \text{ cm}^3) & \text{Graham TP, 1971} \end{cases}
\]
\[
LAm_{\text{max}} = 1.14V + 0.14 \quad \text{Arcilla RA, 1973}
\]

Abbreviations: RVEDV = right ventricular end-diastolic volume, LVEDV = left ventricular end-diastolic volume, LAm_{\text{max}} = maximum left atrial volume, V = measured volume on angiocardiogram

TABLE III NORMAL CARDIAC VOLUMES

\[
RVEDV = 68.6 (\text{BSA})^{1.32},
\]
\[
LVEDV = 70.3 (\text{BSA})^{1.35},
\]
\[
LAm_{\text{max}} = 40.7 (\text{BSA})^{1.19}.
\]

Abbreviations: RVEDV = right ventricular end-diastolic volume, LVEDV = left ventricular end-diastolic volume, LAm_{\text{max}} = maximum left atrial volume, BSA = body surface area

and on 10 of the 11 survivors one to 2 months and/or one to 2 years postoperatively. The hemodynamic studies were carried out according to the following equations: 1) Ratio of pulmonary to systemic pressure (Pp/Ps) = systolic pressure of pulmonary artery / systolic pressure of aorta. 2) Ratio of pulmonary to systemic blood flow (Qp/Qs) = [VO2 / (PVO2 - PAO2)] / [VO2 / (A02 - VO2)] = (A02 - VO2) / (PVO2 - PAO2). 3) Ratio of pulmonary to systemic vascular resistance (Rp/Rs) = [(PA - LA) / Qp] / [(A02 - RA) / Qs] = [(PA - LA) / (A02 - RA)] / [Qs/Qp], where VO2 is oxygen consumption, PVO2 is oxygen content in pulmonary venous blood, A02 is oxygen content in arterial blood, VO2 is oxygen content in mixed venous blood, PA is mean pressure of pulmonary artery, LA is mean pressure of left atrium, A0 is mean pressure of aorta and RA is mean pressure of right atrium.

The preoperative left heart volume could be measured in all patients but the right heart volume in only 11 of these children. The left heart volume was calculated according to the area-length method and Simpson’s rule was used to assess the right ventricular volume as shown in Table II. The appendages were excluded from the atrial images. All volume estimations were normalized to body surface area and the

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values were expressed as a percent of the normal (% of N). As the normal values, our own equations\(^6\) were used (Table III).

Statistical analysis was performed by Student's t-test, and p values less than 0.05 were considered significant.

**Technique of Repair**

Profound hypothermia with surface cooling and limited cardiopulmonary bypass (Kyoto technique)\(^7\) was used for 10 patients. The lowest rectal temperature was 16.7\(^\circ\)C on average, ranging from 11\(^\circ\)C to 20\(^\circ\)C. The average circulatory arrest time was 61, ranging from 48 to 72 min. Routine cardiopulmonary bypass was performed on 7 patients. Primary complete repair was carried out on all except one patient, as described later.

With regard to the approach for the supr cardiac type, the posterior approach of Cooley and Hallman\(^8\) was used in 4, the left lateral approach of Roe\(^9\) in one and right lateral approach by Gersony et al.\(^10\) in 6 patients. In 4 with the cardiac type and drainage into the coronary sinus, the atrial septum between the fossa ovalis and coronary sinus was excised and a new atrial septum was prepared, with a pericardial patch, so as to divert the pulmonary venous blood into the left atrium. In 2 with the mixed type, the procedures used for the two types of TAPVD above mentioned were combined. PDA was ligated during the partial bypass and VSD was transven tricularly closed with a Dacron patch.

**RESULTS**

**Factors Affecting the Operative Mortality**

Age: The mortality rates according to age were 75% in 4 patients under 6 months of age, 40% in 5 from 6 to 12 months and 12.5% in 8 over 12 months of age.

Type: The mortality rates were 50% in 4 with the cardiac type and in 2 with the mixed type. The rate was 27% in 11 with the supr cardiac type.

Size of ASD: In 12 patients with an atrial septal defect (ASD) over 10 mm in diameter, the mortality rate was 25%, while it was 60% in 5 patients with small ASD or patent foramen ovale (PFO). In the 5 patients with small ASD or PFO, arterial oxygen saturation was less than 80%.

Associated anomalies: Three of 10 patients with TAPVD alone were lost and in 4 patients with PDA, 3 succumbed. There was no incidence of death in 2 with VSD and one with VSD plus PDA. In these patients, the mean LVEDV was 99% of N, while it was 85% of N in all.

Hemodynamics: There was no correlation between the operative mortality and pulmonary to systemic pressure ratio, vascular resistance ratio and flow ratio (Fig. 1). However, a close correlation was noted between the mortality rate and arterial oxygen saturation. One of 10 patients died when the arterial oxygen saturation was over 80%. In contrast, 5 of 7 with a saturation of less than 80% died.

**Cardiac Volume and Ejection Fraction**

The Right Ventricular End-diastolic Volume and Ejection Fraction

The right ventricular end-diastolic volume (RVEDV) was obtained in 11 patients. The average value of all patients was 168% of N, indicating that generally, the right ventricular chamber is larger than normal in TAPVD. The
average value of 185% of N in the 7 survivors did not significantly differ from that of 136% of N in the 4 who succumbed. On the other hand, the right ventricular ejection fraction (RVEF) was 57.5% on an average for all patients, this value being less than normal. There was no significant difference between the average value of 56.7% of the survivors and that of 58.8% in those who died. Thus, the operative mortality did not correlate with the right ventricular volume and function.

The Maximal Left Atrial Volume and Body Surface Area
Figure 2 shows the relationship among maximal left atrial volume (LAmax), body surface area (BSA) and the operative mortality. Of 17 patients, the average value of LAmax was 60% of N, indicating that left atrial volume is, in general, extremely small in TAPVD. The average value of LAmax was 64.0% of N in 11 survivors and 52.0% of N in the 6 deceased. These two values were significantly different (p < 0.05). With regard to BSA, there was a statistically significant difference between the survivors and those who succumbed (p < 0.001). The 6 deceased patients all had a LAmax of less than 60% of N and a BSA of less than 0.4 m².

The Left Ventricular End-diastolic Volume and Ejection Fraction
The relationship between the survival results and the left ventricular end-diastolic volume (LVEDV) or ejection fraction (LVEF) is shown in Fig. 3. The average value of LVEDV in all patients was 85% of N, indicating that LVEDV is generally within normal limits in TAPVD. Assuming that the normal value of LVEDV is in the range of 120 to 80% of N, LVEDV was normal in 10 patients and less than normal in 7 of 17 patients. LVEF was slightly diminished and the average value was 0.59 in all patients. Pertaining to the operative mortality, the mean LVEDV was 69% of N in 6 deaths, while it was 95% of N in the 11 survivors (p < 0.05). LVEDVs were all less than 65% of N in 4 deaths due to low cardiac output. In 13 patients with a LVEDV greater than 65%.
of N, 9 who had an LVEF greater than 60% survived while death occurred in 2 of 4 patients with a LVEF of less than 53%. These two fatalities were not due to low cardiac output but rather to respiratory failure. Thus, the operative mortality is apparently influenced by LVEDV but not to any great extent by ejection fraction, even when the LVEF is less than normal.

The Size of the Interventricular Communication Size of the interatrial communication did directly correlate with the size of the left heart cavity. The average values of the LAmax and LVEDV were 62.4% of N and 95% of N, respectively, in 12 patients with large ASD, while in 5 patients with small ASD or PFO, the former was 52% of N and the latter 61% of N.

Pre-and Postoperative Alteration of the Left Heart Volume and Ejection Fraction

Pre- and Postoperative Alteration of LAmax and LVEDV

Figure 4 shows pre- and postoperative changes of LAmax and LVEDV. With regard to LAmax, the average value was 64% of N in 10 patients preoperatively, 77% of N in 7 one or 2 months postoperatively and 87% of N in 5 one or 2 years after surgery. The LAmax consistently increased in all. On the other hand, the average value of LVEDV was 95% of N in 11 survivors before

Fig.5. Pre- and postoperative alteration of left ventricular ejection fraction. Dotted area indicates normal range of left ventricular ejection fraction according to Graham et al. Open circles indicate preoperative values, black circles one or 2 month-postoperative values and double circles one or 2 year-postoperative values.
surgery. The LVEDV was already within the normal range in 8 of 10, preoperatively. After surgery the value was 96% of N in 7 one or 2 months and 104% of N in 5 one or 2 years after surgery. The LVEDV increased with time, in all but 2. In these 2 patients, the absolute values of LVEDV increased but % of N was diminutive because body growth was more extensive than the development of the left ventricle.

Pre- and Postoperative Change of LVEF
The pre- and postoperative change of LVEF is shown in Fig. 5. Preoperatively, LVEF was within normal in 7 and abnormal in 3 of 10 patients. Postoperatively, the ejection fraction was normal in all but 2. In these 2 patients with a postoperative LVEF of less than normal, the LVEDV was greater than 124% of N.

A Case without Primary Ligation of the Vertical Vein
Figure 6 shows the postoperative angiography of a 3 year-old girl with an unusually small left heart (No. 3 in Table I, * in Figs. 2–5). She had an isolated TAPVD with drainage into the left innominate vein. Left atrium and common pulmonary venous trunk were exposed through Roe’s left lateral approach; the ASD was directly closed from the left side and a sufficiently large anastomosis was made between the left atrium and common pulmonary venous trunk. Nevertheless, as soon as the anomalous vertical vein had been ligated, left atrial pressure as well as pulmonary venous pressure elevated over 20 mmHg and pulmonary edema ensued. The ligature was then immediately removed and another silk thread was wound around the vertical vein to make a stenosis. Pulmonary venous pressure could thus be kept under 18 mmHg. Both ends of the silk thread were fixed to the chest wall. This patient had preoperatively an extremely small left heart with a LAMax of 43% of N and a LVEDV of 66.1% of N. Two months after surgery, the LAMax was 60% of N and LVEDV was 71% of N. These values were 70% of N and 78% of N, respectively, 20 months after surgery. Left-to-right shunt was 30% 2 months postoperatively and 20 months after surgery, no shunt was detected on blood gas analysis, though the vertical and innominate veins were dimly opacified on the pulmonary arteriogram.

When both LAMax and LVEDV are extremely small as in this patient, primary ligation of the anomalous vertical vein should be avoided and delayed ligation is preferable. Our procedure, is considered useful because reoperation is unnecessary as the anomalous vertical vein is gradually but steadily tightened with physical development.
DISCUSSION

With regard to major factors affecting the pathophysiology and the surgical results in TAPVD, there are anatomical defects including pulmonary venous obstruction, anatomical type of anomalous connection, the size of interatrial communication and the associated anomalies, and the hemodynamic factors including pulmonary artery pressure, pulmonary vascular resistance, pulmonary blood flow, etc!11–14 Recently, heart volumes, particularly, left heart volumes have also been discussed15,16 In the present study, the mortality rate correlated with age, the size of ASD, arterial oxygen saturation and the left heart volumes but did not correlate with pulmonary to systemic pressure ratio, flow ratio and resistance ratio. The type of anomalous connection did not correlate with the mortality rate though no infracardiac type of TAPVD was included in this series.

In surgical repair, the most crucial problem is considered to be the size of the anastomosis between the left atrium and the common pulmonary venous trunk. Roe9 stated that the size should be greater than the area of mitral valve. We hold the same opinion. Pressure measurement revealed no pressure gradient across the anastomosed portion, in any patient in this series.

In patients whose symptoms appear at an early age, the size of interatrial communication was small and arterial oxygen saturation was low because mixing of blood was poor at the atrial level. In such patients, left atrial and ventricular volumes were small and the mortality rate was high. On the contrary, in patients with a large VSD, arterial oxygen saturation was relatively high and LVEDV was relatively large regardless of the LAMax. Despite the extensive surgical invasion required to close the VSD, all 3 patients with VSD survived. Therefore, an evaluation was done regarding the relationship between heart volume and the surgical results.

RVEDV is consistently greater than normal in this entity15–16 Mathew et al17 reported that the mean RVEDV was 152% of N in the patients with pulmonary venous obstruction and 215% of N in the patients without pulmonary hypertension. In our study, the average value of RVEDV was 168% of N and similar to that indicated by Mathew et al. But the RVEDV did not correlate either with the pulmonary arterial pressure or the operative mortality. As to RVEF, Mathew et al17 reported that 0.53 of RVEDV was slightly smaller than the normal value of 0.59. Nakazawa et al15 stated that the average value was significantly less than normal in infants but normal in older children. In the present study, the mean RVEF was 0.58 in 11 patients and 0.57 in survivors, while it was 0.59 in the deceased patients.

It is now generally accepted that the left atrium in TAPVD is smaller than normal15,16,18,19 Prior to repair, the left atrium plays the role of conduit from the right atrium to the left ventricle. By contrast, after repair, a reservoir function to the left atrium is required as the left ventricle fills only from the left atrium. Other workers stated that a small atrial cavity may impair cardiac output14,20 According to Trusler et al21 reduction of atrial volume in dogs by more than 50% reduced the cardiac output by about 40%. We have often seen marked phasic pressure fluctuations in the left atrial pressure tracings in postoperative patients with a small left atrium, and such indicates a poor reservoir function in the atrium.14 Thus, the risk involved in surgery increases in patients with a small left atrium. In our study, LAMax was closely correlated with the operative mortality. Katz et al22 stated that lack of left atrial reservoir function was not a major problem providing all other aspects were favorable. However, their patients who did not survive had a small left atrium and ventricle.

Pertaining to LVEDV in TAPVD, Mathew et al17 and Graham et al16 stated that the average value was generally within normal. Nakazawa et al15 mentioned the average value of 79% of N in patients with a peak right ventricular pressure equal to or higher than a peak left ventricular pressure, while it was 85% of N in patients with a peak right ventricular pressure less than the peak left ventricular pressure. In the present study, the average value of LVEDV for the entire group was normal and LVEDV did not correlate with pulmonary artery pressure, but did correlate with the size of interatrial communication. Graham et al16 stated that LVEDV was less than 67% of N in one third of their patients. In our series also, the LVEDV was less than 65% of N in 4 of 17 patients and these 4 all died with low cardiac output syndrome.

In TAPVD, the LVEF is generally reduced. According to Mathew et al17 LVEF in the patients with severe pulmonary hypertension and pulmonary venous obstruction was less than that
in the patients without these complications. In our series, also, the average value of LVEF in the deceased was significantly less than that in the survivors. Thus, left heart function including LAmax, LVEDV and LVEF is impaired in TAPVD.

It has long been suggested that enlargement of the small left atrium may be necessary but an effective enlargement is difficult. Katz et al. reported good surgical results with no significant difference in survival with or without left atrial enlargement. Goor et al. described an excellent procedure for enlargement of the left atrium but also stated that the small size of the left atrium does not always hinder good operative results. However, since a reduced atrial volume impairs cardiac output, left atrial enlargement does seem reasonable. On the other hand, when both the left atrium and ventricle are extremely small, as in our patient, left atrial enlargement only does not ensure success. In such patients, a delayed ligation of the vertical vein is preferable. Particularly, the procedure in which both ends of the thread are wound around the vertical vein to the chest wall should be given strong consideration. Re-operation is not necessary as the vertical vein gradually shrinks with body growth.

Cardiac chambers and function are, generally speaking, restored to normal after surgery but in some patients, LVEDV increases to greater than normal. In our series, postoperative LVEDV was greater than 120% of N in 3 patients. In 2 of these 3 patients, LVEF was reduced to less than normal despite a normal postoperative LVEF. Although, it is unclear why increase in LVEDV was over the normal, myocardial ischemia during surgery or postoperative rapid increase in the volume overload probably results in an overexpansion of the left ventricle.

As a result of this evaluation, the following management plan was formulated:
1) When TAPVD is suspected, echo cardiography, cardiac catheterization and angiocardiography should be done as soon as possible. 2) Emergency or semi-emergency surgery should be done when pulmonary venous obstruction is evident. 3) In the absence of pulmonary venous obstruction, balloon atrial septostomy should be done regardless of the presence or absence of an intratral pressure gradient. 4) When symptoms do not improve, surgical intervention is required regardless of age or severity of symptoms. 5) At the time of surgical repair, the size of the anastomosis between the left atrium and pulmonary vein should be sufficiently large, at least, larger than the mitral valve area. 6) When pulmonary venous pressure is extremely elevated and results in pulmonary congestion, primary ligation should be avoided and delayed ligation should be considered.

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