One-Stage Correction of Ventricular Septal Defect, Tetralogy of Fallot and Complete Transposition of the Great Arteries in Infancy

by

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Recent progress in diagnostic techniques and hemodynamic studies in congenital heart disease has revealed that some cardiac anomalies should be corrected even in infancy.

In Amagasaki Hospital during the past six years surgery was performed on 165 patients under the age of 12 months to correct various kinds of congenital heart disease (Table 1).

The technical difficulties in these small infants with complicated cardiac anomalies forced us to develop a technique of "profound hypothermia with surface cooling and limited cardiopulmonary bypass (Kyoto technique)"1-8. It will be outlined here only briefly (Table 2).

The infant is anesthetized with nitrous oxide, halothane and Dialferine and cooled by

Table 1. Open-heart surgery under the age of 12 months
(Sept., 1968 to July, 1974)

<table>
<thead>
<tr>
<th></th>
<th>Hypothermia</th>
<th>Bypass</th>
</tr>
</thead>
<tbody>
<tr>
<td>Cases</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Deaths</td>
<td></td>
<td></td>
</tr>
<tr>
<td>VSD</td>
<td>46</td>
<td>60</td>
</tr>
<tr>
<td>TF</td>
<td>9</td>
<td>9</td>
</tr>
<tr>
<td>TGA I</td>
<td>6</td>
<td>0</td>
</tr>
<tr>
<td>II</td>
<td>6</td>
<td>0</td>
</tr>
<tr>
<td>TAPVC</td>
<td>3</td>
<td>3</td>
</tr>
<tr>
<td>Miscellaneous</td>
<td>9</td>
<td>14</td>
</tr>
<tr>
<td><strong>Total</strong></td>
<td><strong>79</strong></td>
<td><strong>86</strong></td>
</tr>
</tbody>
</table>

Legend: VSD, Ventricular septal defect. TF, Tetralogy of Fallot. TGA, Complete transposition of the great arteries. TAPVC, Total anomalous pulmonary venous connection.

(Read at the Seminar of Japanese Medicine, San Jose, Costa Rica, August 26 to 31, 1974)

Key words: Ventricular Septal Defect, Tetralogy of Fallot, Complete Transposition of the Great Arteries, Deep Hypothermia, Corrective Surgery in Infancy, One-Stage Correction.

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Table 2. Hypothermic procedures (Kyoto technique)

<table>
<thead>
<tr>
<th>Procedure</th>
<th>Dosage</th>
<th>Mode</th>
</tr>
</thead>
<tbody>
<tr>
<td>1) Premedication</td>
<td>Atropine sulfate 0.02mg/Kg</td>
<td>i.m.</td>
</tr>
<tr>
<td></td>
<td>Pethilorfan 1~2mg/Kg</td>
<td></td>
</tr>
<tr>
<td>2) Anaesthesia</td>
<td>halothane</td>
<td>(0.5~1.0%)</td>
</tr>
<tr>
<td></td>
<td>carbon dioxide</td>
<td>(1~3%)</td>
</tr>
<tr>
<td></td>
<td>nitrous oxide</td>
<td>(0~50%)</td>
</tr>
<tr>
<td></td>
<td>oxygen</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Dialferine 0.5mg/Kg</td>
<td>i.v.</td>
</tr>
<tr>
<td>3) Chlorpromazine</td>
<td>0.5mg/Kg</td>
<td>i.v. drip</td>
</tr>
<tr>
<td>4) Surface cooling (blanket + ice bags)</td>
<td>22~17°C</td>
<td></td>
</tr>
<tr>
<td>5) Bypass cooling</td>
<td>down to 20~13.5°C</td>
<td></td>
</tr>
<tr>
<td></td>
<td>(average 17.7°C)</td>
<td></td>
</tr>
<tr>
<td>6) Circulatory arrest</td>
<td>54~95min.</td>
<td>(average 83min.)</td>
</tr>
<tr>
<td>7) Bypass rewarming</td>
<td>up to 34~35°C</td>
<td></td>
</tr>
<tr>
<td>8) surface warming</td>
<td>up to 36°C</td>
<td></td>
</tr>
</tbody>
</table>

a posterior water blanket and anterior ice bags. Carbon dioxide (1 to 3 per cent) is added to the respiratory gases during the cooling period. As no important arrhythmia has ever occurred during the cooling phase, surface cooling is usually continued until the rectal temperature reaches 17~22 °C.

The heart is exposed through a sternal splitting incision, and after heparinization the infant is cooled to 13.5~20 °C with partial cardiopulmonary bypass, depending on the anticipated duration of circulatory arrest. The intracardiac repair is carried out in a totally bloodless and completely relaxed heart. At the end of repair, rewarming is carried out to 36 °C with partial cardiopulmonary bypass and surface rewarming. When the duration of total circulatory arrest is anticipated to exceed the permissible duration for total correction of complicated cardiac anomalies, intracardiac procedures can be divided into two or three steps. In five infants with complex lesions, circulatory arrest has been interrupted by a second or third short period of cold perfusion to allow an extension of circulatory arrest beyond 90 minutes (Fig. 1).

In the early period, this hypothermia technique was employed for all infants under 10 kg of body weight who required open-heart surgery for the correction of congenital cardiac anomalies. Thanks to the progress of total cardiopulmonary bypass technique in infants, we started to employ the usual type of cardiopulmonary bypass in May 1969, for patients weighing more than 6 kg in the correction of ventricular septal defect (VSD) and tetralogy of Fallot (TF), which are relatively simple procedures. In patients with the same lesions weighing less than 6 kg, the hypothermia technique was employed. This hypothermia technique has been applied not only during the first year of life but also during the second year of life for the correction of complete transposition of the great arteries (TGA), total anomalous pulmonary venous connection (TAPVC) and other complicated cardiac anomalies.

Among the various congenital cardiac problems of infants, this report focuses on three typical anomalies: VSD with severe pulmonary hypertension, TF, and TGA. The operative results of one-stage correction during infancy are discussed and compared with those in age groups above one year.
1. Ventricular Septal Defect (VSD)

Patients were classified into four groups based on their preoperative hemodynamic status. The hospital mortality rate of each age group is shown in Table 3. None of the patients had any palliative surgery prior to closure of the VSD.

In 212 patients, when the pulmonary artery to systemic artery peak systolic pressure ratio (Pp/Ps) was less than 0.75, the mortality rate was 0.9%, and there was no significant difference among the age groups. When Pp/Ps was greater than 0.75, especially when the pulmonary to systemic resistance ratio (Rp/Rs) exceeded 0.5, the hospital mortality rate of infants was 19%. The high rate of mortality was due mostly to failure of the cardiopulmonary bypass or of postoperative respiratory care. Since Feb. 1971, however, with improved cardiopulmonary bypass and postoperative care techniques, 78 patients with severe pulmonary

Table 3. Operative results of primary closure of ventricular septal defect

<table>
<thead>
<tr>
<th></th>
<th>0~12 mo.</th>
<th>12~24 mo.</th>
<th>2~38 yr.</th>
<th>Total</th>
</tr>
</thead>
<tbody>
<tr>
<td>Pp/Ps≤0.75</td>
<td>27 (4%)</td>
<td>39 (0%)</td>
<td>146 (0.7)</td>
<td>212 (0.9)</td>
</tr>
<tr>
<td>Pp/Ps&gt;0.75</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Rp/Rs≤0.5</td>
<td>36 (11%)</td>
<td>22 (9%)</td>
<td>15 (7)</td>
<td>73 (10)</td>
</tr>
<tr>
<td>Rp/Rs 0.5~0.75</td>
<td>27 (19%)</td>
<td>10 (10%)</td>
<td>18 (6)</td>
<td>55 (13)</td>
</tr>
<tr>
<td>Rp/Rs≥0.75</td>
<td>16 (19%)</td>
<td>9 (11%)</td>
<td>9 (22)</td>
<td>34 (18)</td>
</tr>
<tr>
<td>Total</td>
<td>106 (12%)</td>
<td>80 (5%)</td>
<td>188 (3%)</td>
<td>374 (6%)</td>
</tr>
</tbody>
</table>

No.: Number of cases, M.R.: Mortality rate (per cent) (Sept., 1968 to July, 1974)
CORRECTION OF VSD, TF, AND TGA IN INFANCY

Table 4. Operative results of primary closure of ventricular septal defect in patients whose Pp/Ps was greater than 0.75 (Feb., 1971 to July, 1974)

<table>
<thead>
<tr>
<th>Age (Years)</th>
<th>Under 1</th>
<th>1</th>
<th>2 ~</th>
<th>Total</th>
</tr>
</thead>
<tbody>
<tr>
<td>Rp/Rs</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>0.5 ~ 0.75</td>
<td>7 (0)</td>
<td>6 (0)</td>
<td>5 (1)</td>
<td>18 (1)</td>
</tr>
<tr>
<td>≥ 0.75</td>
<td>5 (1)</td>
<td>6 (0)</td>
<td>5 (1)</td>
<td>16 (2)</td>
</tr>
</tbody>
</table>

( ): Number of hospital deaths

hypothesis with Pp/Ps greater than 0.75, have been operated upon with only two deaths (Table 4).

Thus, good operative results are achieved with VSD infants, even in the presence of severe pulmonary hypertension, intractable congestive heart failure, marked growth retardation and recurrent prolonged lower respiratory infections, and one-stage operation is the first choice.

2. Tetralogy of Fallot (TF)

Patients were classified into three groups according to the ratio of the diameter of the pulmonary annulus to that of the aorta (PA/AO), which is obtained from the lateral view of the preoperative angiocardiogram (Table 5). In group I, the ratio (PA/AO) was 0.5 or greater. In group II, it was between 0.3 and 0.5, and in group III, it was less than 0.3.

In group I (35 patients) there were no deaths. In group II, 8 out of 57 died, a mortality rate of 14%. In group III, we lost 10 out of 22, a mortality rate of 45%. The PA/AO ratio shows a high correlation with the mortality rate. Thus, the high mortality rate of 22% in the first year of life is explained by the fact that 6 out of the 18 patients were in group III, with severely hypoplastic pulmonary arteries less than one third the diameter of the aorta. There was no significant difference in mortality rate among the three age groups.

The operative results of total correction of TF depend more on the development of the pulmonary artery than on the age and weight of the patient.

The pulmonary valve area after operative enlargement was measured in 87 patients. Figure 2 shows the mortality rate of these 87 patients in relation to the degree of

Table 5. Hospital mortality rate for total correction of Tetralogy of Fallot (Sept., 1968 to July, 1974)

<table>
<thead>
<tr>
<th>PA/AO</th>
<th>0~12 mo.</th>
<th>12~24 mo.</th>
<th>2~23 yr.</th>
<th>Total</th>
</tr>
</thead>
<tbody>
<tr>
<td>Group I ≥ 0.5</td>
<td>4 (0)</td>
<td>6 (0)</td>
<td>25 (0)</td>
<td>35 (0)</td>
</tr>
<tr>
<td>Group II 0.3~0.5</td>
<td>8 (13)</td>
<td>12 (17)</td>
<td>37 (14)</td>
<td>57 (14)</td>
</tr>
<tr>
<td>Group III ≤ 0.3</td>
<td>6 (50)</td>
<td>2 (50)</td>
<td>14 (43)</td>
<td>22 (45)</td>
</tr>
<tr>
<td>Total</td>
<td>18 (22%)</td>
<td>20 (15%)</td>
<td>76 (14%)</td>
<td>114 (16%)</td>
</tr>
</tbody>
</table>

No.: Number of cases, M.R.: Mortality rate (%)

PA/AO = \frac{\text{diameter of pulmonary annulus}}{\text{diameter of aorta}}
enlargement of the pulmonary annulus. The postoperative size of the pulmonary annulus is expressed in terms of pulmonary valve area per unit body surface area (PVA/BSA). The horizontal axis shows the BSA and the vertical axis the diameter of the pulmonary annulus. The two lines indicated as 2.0 and 1.5 cm²/M² (pulmonary valve area per unit body surface area) divide the cases into three groups: group A with PVA/BSA greater than 2.0 cm²/M²; group B, PVA/BSA between 1.5 and 2.0 cm²/M²; and group C, PVA/BSA less than 1.5 cm²/M².

In group A, 46 out of 47 patients survived. In these patients, satisfactory hemodynamic results were obtained postoperatively without outflow tract prosthesis. In groups B and C, on the other hand, out of 13 patients with a BSA of more than 0.6 M² (over 4 or 5 years of age), 12 survived, while out of 26 patients with a BSA of less than 0.6 M² (under 3 or 4 years of age), we lost 46% of the cases.

This fact clearly demonstrates that the pulmonary valve area should be enlarged at least up to 2.0 cm²/M² in patients under 3 or 4 years of age. It may also be said that in the older age group enlargement of the outflow tract has little effect on the prognosis, and that in the younger age group more extensive correction of the stenosis is required and there is more need for outflow tract prosthesis.

3. Complete Transposition of the Great Arteries (TGA)

Complete transposition of the great arteries is one of the major causes of high morbidity during early infancy. As shown in Figure 3, in Japan the number of operative cases during the first year is still limited and the results are not satisfactory. Recent improvements of diagnostic technique are beginning to allow more cases of TGA to reach the hands of surgeons. Complete transposition of the great arteries is classified into four groups according to Mustard's classification. In group I, the ventricular septum is intact and slight left
VENTRICULAR OUTFLOW OBSTRUCTION OR A SMALL VSD MAY EXIST. IN GROUP II, THERE IS A LARGE VSD. IN GROUP III, VSD AND LEFT VENTRICULAR OUTFLOW OBSTRUCTION ARE PRESENT. IN GROUP IV, THE VENTRICULAR SEPTUM IS INTACT AND THERE IS LEFT VENTRICULAR OUTFLOW OBSTRUCTION.

IN ALL GROUPS BALLOON ATRIAL SEPTOSTOMY IS PERFORMED AS SOON AS THE DIAGNOSIS IS ESTABLISHED. IN GROUP I, WHEN THE ATRIAL SEPTOSTOMY IS NOT EFFECTIVE, ATRIAL BAFFLE REPAIR IS USUALLY PERFORMED EARLY. IN GROUP II, SINCE SEVERE PULMONARY VASCULAR OBSTRUCTIVE DISEASE OFTEN DEVELOPS EARLY, WE PERFORM CLOSURE OF THE VSD AND ATRIAL BAFFLE REPAIR AS SOON AS POSSIBLE IN ONE STAGE WITHOUT BANDING THE PULMONARY ARTERY. IN GROUP III AND IV, INFANTS ORDINARILY SURVIVE THE FIRST TWO YEARS OF LIFE, AND IF LEFT VENTRICULAR OUTFLOW OBSTRUCTION IS SEVERE, THE SYSTEMIC-PULMONARY ARTERIAL ANASTOMOSIS (I.E. WATERSTON OR BLALOCK-TAUSSEJ SHUNT) IS A VERY EFFECTIVE MEANS OF PALLIATION IN INFANCY.

TOTAL CORRECTION WAS PERFORMED IN A TOTAL OF 24 PATIENTS. WE LOST 7 OUT OF 12 PATIENTS UNDER THE AGE OF 12 MONTHS, AS SHOWN IN Figure 4. THESE DEATHS WERE STILL RELATED TO AVOIDABLE ERRORS IN MANAGEMENT: LATE DISCOVERY OF TAMPOONADE, HEMOLYSIS, DIGITALIS INTOXICATION AND SURGICAL ERRORS. SINCE 1972, HOWEVER, IMPROVED OPERATIVE TECHNIQUES, AND PRE- AND POSTOPERATIVE CARE HAVE REDUCED THE MORTALITY RATE TO 36% IN GROUP II, THAT IS FOUR DEATHS IN 11 PATIENTS (Table 6). THIS RESULT IS ALMOST THE SAME AS THAT OF CHAMPSAUR IN TORONTO7, DANIELSON IN ROCHESTER8, VENUGOPAL IN BIRMINGHAM9, AND BARRATT-BOYES IN AUCKLAND10. THEY REPORTED 45%, 55%, 30%, AND 56% MORTALITY RATES, RESPECTIVELY. THE CAUSES OF THESE FOUR DEATHS WERE ANALYSED, AND SEVERE PULMONARY OBSTRUCTIVE DISEASE WAS NOTED IN THREE CASES
Fig. 4 Mustard operation for complete transposition of the great arteries
(March, 1970 to July, 1974). Open circle=survival, Solid circle=death

Table 6. Mustard operation for complete transposition of the great arteries associated
with ventricular septal defect (March, 1970 to July, 1974)

<table>
<thead>
<tr>
<th>Age</th>
<th>March, 1970 ~ 1971</th>
<th>1972 ~</th>
<th>Total</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>cases</td>
<td>died</td>
<td>cases</td>
</tr>
<tr>
<td>&lt; 1 Year</td>
<td>4</td>
<td>3</td>
<td>2</td>
</tr>
<tr>
<td>≥ 1 Year</td>
<td>2</td>
<td>1</td>
<td>9</td>
</tr>
<tr>
<td>Total</td>
<td>6</td>
<td>4</td>
<td>11</td>
</tr>
</tbody>
</table>

in which pulmonary arteriolar resistance was over 2500 dynes sec cm⁻². In retrospect it is
considered that surgery was not indicated in these three cases. The fourth patient died of
digitalis intoxication in the early postoperative period, an avoidable error.

In group II of TGA, determination of the indications for total repair has been
considered to be difficult because of the complex nature of the intracardiac shunts and the
difficulty of evaluating the pulmonary circulation.

As the systemic and pulmonary circuits are parallel in complete transposition of the
great arteries, flow and resistance levels in the two circuits may vary independently (Fig. 5)¹¹.
Therefore, the flow and resistance ratios between the two circuits, Qp/Qs or Rp/Rs, are
often misleading in the evaluation of the pulmonary circulation in TGA. To assess accurately
the status of the pulmonary vascular bed, the pulmonary circuit must be considered alone.
Decreasing total pulmonary blood flow, secondary to increasing pulmonary vascular disease,
results in decreased intercirculatory mixing, and a decrease in systemic arterial oxygen
saturation. This decrease in saturation is partially compensated for by an increase in
hemoglobin concentration.

From this point of view in about 10 of our 17 patients in group II, pulmonary
arteriolar resistance was calculated and precise studies were done to correlate pulmonary
arteriolar resistance (PAR) with operative results, age at operation, effective pulmonary
flow index (QEI) and systemic arterial oxygen saturation index (SAI).
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Fig. 5 Schematic representation of circulation in complete transposition of the great arteries (From Mair et al.11)

Fig. 6 Relation of pulmonary arteriolar resistance to age at time of total repair and operative results in complete transposition of the great arteries associated with ventricular septal defect

Table 8. Formulas for the calculation of pulmonary flow, effective pulmonary flow and effective pulmonary flow index

Pulmonary flow = \( \frac{O_2 \text{ consumption}}{(\text{pulmonary venous } O_2 \text{ content} - \text{pulmonary artery } O_2 \text{ content})} \)

Effective pulmonary flow (QE) = \( O_2 \text{ consumption} \)

(pulmonary venous \( O_2 \) content — systemic mixed venous \( O_2 \) content)

Effective pulmonary flow index (QEI) = \( \frac{\text{effective pulmonary flow}}{\text{body surface area}} \)
Figure 6 shows the relation of PAR to age at operation and operative results. No patient with a PAR over 2500 dynes sec cm\(^{-5}\) survived the operation. At autopsy, all these patients had changes of grade 4 or more according to the Heath-Edwards classification\(^{12}\) in the pulmonary arterioles. It is quite interesting that there are babies who had a PAR over 4000 dynes sec cm\(^{-5}\) at the age of three months or younger, while there are lucky patients who had a PAR under 2500 dynes sec cm\(^{-5}\) even at the age of 7 or 8 years.

The term “effective pulmonary flow” means the flow necessary to take up metabolic oxygen requirements if systemic mixed venous blood alone were perfusing the lungs. In
the calculation of pulmonary flow, effective pulmonary flow, and effective pulmonary flow index, the formulas in Table 8 were used.

The relation of PAR to QEI is shown in Figure 7. Effective pulmonary flow index was significantly related to PAR except in one case which had banding of the pulmonary artery prior to total repair. All four patients who had QEI of 1.2 L/min/M² or greater were successfully operated upon except one patient who died of digitalis intoxication postoperatively.

As mentioned above, decrease in effective pulmonary flow (i.e. decrease in intercirculatory mixing), results in a decrease in systemic arterial oxygen saturation. Figure 8 shows that in our patients with TGA and large VSD, effective pulmonary flow index was also significantly related to systemic oxygen saturation. When QEI was greater than 1.2 L/min/M² or systemic arterial oxygen saturation was greater than 55%, all patients survived except for three who died of avoidable causes: hemorrhagic tendency, surgical error and digitalis intoxication.

The relationship between pulmonary arteriolar resistance (PAR) and systemic arterial saturation index (SAI) is shown in Figure 9. This systemic arterial saturation index was obtained by dividing the systemic arterial saturation by the hemoglobin concentration. The three patients whose SAI was less than 2.5 did not survive operation. On the other hand, we lost only two patients out of seven whose SAI was greater than 2.5. One patient had
anemia, which caused a falsely high systemic oxygen saturation index; the other died of digitalis intoxication. Therefore, we suppose that all patients with SAI of 2.5 or greater can survive total repair.

As already mentioned, pulmonary arteriolar resistance showed a high correlation with QEI and SAI. With these indexes, we can now follow the babies and determine the indications and optimum time for total repair of TGA with large VSD. That is, if the systemic arterial saturation index is stable or increases, a decrease in total pulmonary blood flow secondary to increasing pulmonary vascular disease or pulmonary stenosis is extremely unlikely and recatheterization can be avoided.

In summary, for TGA associated with large VSD and severe pulmonary hypertension, total repair should be performed when the QEI is greater than 1.2 L/min/m², or the SAI is greater than 2.5.

One-stage corrections of VSD, TF and TGA in infancy have been discussed. It is believed that the operative results of complicated cardiac anomalies by one-stage correction will be greatly improved with the application of profound hypothermia (Kyoto technique), and improved postoperative management.

Considering the natural course of the disease and also the results of two-stage operation, we should work for the further development of the one-stage operation in infancy for these anomalies.

References

和文抄録

心室中隔欠損症．Fallot 四頭症，完全
大血管転位症の乳児期期的根治手術

兵庫県立心胸外科
森 湧悦，安藤史隆，節家直己，山口勝雄
奥 秀遠，神崎義雄，河井 深，城谷 均

過去6年間において兵庫県立心胸科心胸外科にて
行なった生後12か月未満の乳児期心根治手術例は、
1974年7月現在165例である（表1）。
われわれは以前より乳児期における心根治手術に
は表面冷却と体外循環による急速加温法を併用した超
低体温法（京大方式）が有力であることを報告し、こ
れを使用している。最近使用している具体的なこの低
体温法の方法は表2に示したものをある。以前は体重
10kg以下の全ての低体温法を使用していたが1969
年5月より体重6kg以下のものに使用しその他は通
常の人工心肺による完全体外循環下にて手術を行なって
いる。しかし6kg以上でも完全大血管転位症（TGA）、
緑肺静脈逆流異常症（TAPVC）などのような心房内
で手術操作を必要とする複雑心房型に対しては前述の
超低体温法を使用している。本論文では乳児期期心
根治手術が必要となる乳児症例のうちVSD, TF, TGA の
3症例について、生後12か月未満の症例と生後1年以
上の症例とを対比してその乳児期期的根治手術の手
術成績、手術適応などについて報告する。

1. 心室中隔欠損症（VSD）

生後1年以内の症例は106例で死亡率12％である（表
3）。これ術前肺・体動脈収縮期圧比（Pp/Ps）、
および肺動脈圧抵抗比（Rp/Rs）にわけ、さらに年令
別に検討してみると、Pp/Psや Rp/Rs比より考えて
重症と思われる症例でも乳児期根治手術の成績が特
他の年令層に比し有意に劣っていなかったことが判明す
る。すなわち乳児期根治手術を必要とする症例は重
症例が多いことが死亡率を高めているといえる。しか
しながら1971年2月より人工心肺操作や術後呼吸管理
の改善により表4のごとくPp/Psが0.75以上の症例
120例うちR/Rs0.75以上の16例中2例を失ったの
みというきわめて良好な手術結果を得た。

以上より乳児期心根治手術におけるVSDに関し
はいかに重症であっても現在では積極的に一期的根
治手術を行う方針をとっている。

2. ファロー四頭症（TF）

乳児期における一期的根治手術例は18例で死亡率22
％であった。TFの重症度を肺動脈・大動脈直径比
（PA/AO比）で分類した場合年令間で手術
成績にまったく差をみるとめない（表5）。われわれの
乳児期死亡の高かった理由はPA/AO比が0.3以下の
症例が非常に多かったためである。このことは
TF根治手術成績が乳児の年令や体重にかかわる関
係なく、その肺動脈の発育程度にのみ左右されること
をしめしている。なおこの結果より以前は肺動脈幹ま
では外科的に修復可能であるという観点から無選択に
一期的根治手術を行なえてきたが、現在では術前PA/
AO比0.3以上のものは一期的根治手術の対象と
し、0.3以下のものは二期的術手の方針をとっ
ている。

さらに手術成績を術後の肺動脈弁締部の拡大度とそ
の心室の体表面積との相関であらわされたのが図2であ
る。この図より手術時に右室流出路断面積が体表面積
あたり2.0cm²前後にまで拡大されなければならない
ことをしめし、特に3なしに4才以下の年少例はどこ
が強く要求されることをしめしている。

3. 完全大血管転位症（TGA）

完全大血管転位症はMustardの分類により4群に
分岐されるが乳児期においてはいずれの群に属するものでも診断が確定すればだちにballoon atrial septostomy (BAS)を行なっている。IおよびII群ではBASの効果が不良であり、さらにII群では後述するような血行動態上の悪化（肺血管病変の進行）がみられればいずれの群でも姑息的手術を行うと乳児期でもI群ではMustard手術、II群でも一期的にMustard手術とVSD開鎖を同時に行っている。IIIおよびIV群では通常生後1ないし2年間は枝脈なく生存しうる症例が多いが左室流出路狭窄が高度のもののみ乳児期において肺・体循環短絡手術を行なっている。

1974年7月までに行なったTGA根治手術例は計24例でそのうち1例のみ生後8カ月で肺動脈枝縮術を受けていたが他の症例は全例まったく姑息的手術を受けていなかった。24例中乳児期一期的根治手術例は12例（うち死亡7例（図4））であり満足すべき結果を得ていない。しかしながら手術成績、術前術後管理の改善により、1才以上の症例をも含めた2群では1971年以前には6例中4例の死亡であったが1972年以後は11例中4例（死亡率36％）の死亡を見るにすぎなくなり著しい改善がみられる（表6）。これは世界中の各施設の報告と大体同じ成績（約40％）であるが、これらのが報告例は大部分姑息的手術を以前に行っているのでその通算死亡率を考えればわざわざの成績は非常に良好といえる。I群についてもわざわざの症例は大部分が初期の症例であり、最近ではStarkも乳児期早期の一期的根治手術死亡率は4％と報告しており、乳児期一期的根治手術が世界的に行われている。

II群の根治手術成績が世界的にみても不良であるがこの原因はII群においては乳児期早期にしかも高度の閉塞性肺血管病変が起こることによるとされる。われわれは計算したII群症例10例について各種の術前血行動態の指標と手術成績を比較したが肺小動脈抵抗2500dynes sec cm⁻²以上の症例はすべて死亡していることが判明した（図6）。さらに有効肺血管流量係数1.2L/min/M²以下のもの（図7）、動脈血酸素飽和度55％以下（図8）、動脈血酸素飽和度係数2.5以下（図9）のものはすべて死亡した。これらの死亡例は肺血管病変が高度と考えられ現状では根治手術の適応がなかったものと思われる。なお動脈血酸素飽和度係数は外来においても検査できるので、心電カテーテル検査後にこの係数が次第に低下を示す症例では手術の適応となり、手術時期決定の有力な指標とすることができる。

以上VSD、TF、TGAについて乳児期における一期的根治手術の手術適応、手術成績について報告した。