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

by

ATSUMI MORI, FUMITAKA ANDO, NAOKI SETSUIE, KATSUO YAMAGUCHI,
HIDETAKA OKU, YOSHIO KANZAKI, JUN KAWAI, and HITOSHI SHIROTANI

The Department of Cardiovascular Surgery, Hyogo Kenritsu
Amagasaki Hospital, 27-Kitajyonai, Amagasaki, Hyogo, Japan

Received for Publication, May 10, 1975

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)"¹⁻⁵⁾. It will be outlined here only briefly (Table 2).

The infant is anaesthetized 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)

	Hypothermia		Bypass	
	Cases	Deaths	Cases	Deaths
VSD	46	5	60	8
TF	9	1	9	3
TGA I	6	3	0	0
II	6	4	0	0
TAPVC	3	2	3	1
Miscellaneous	9	5	14	5
Total	79	20	86	17

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,

Present address: The Department of Cardiovascular Surgery, Hyogo Kenritsu Amagasaki Hospital, 27-Kitajyonai, Amagasaki, Hyogo, Japan. 〒660

Table 2. Hypothermic procedures (Kyoto technique)

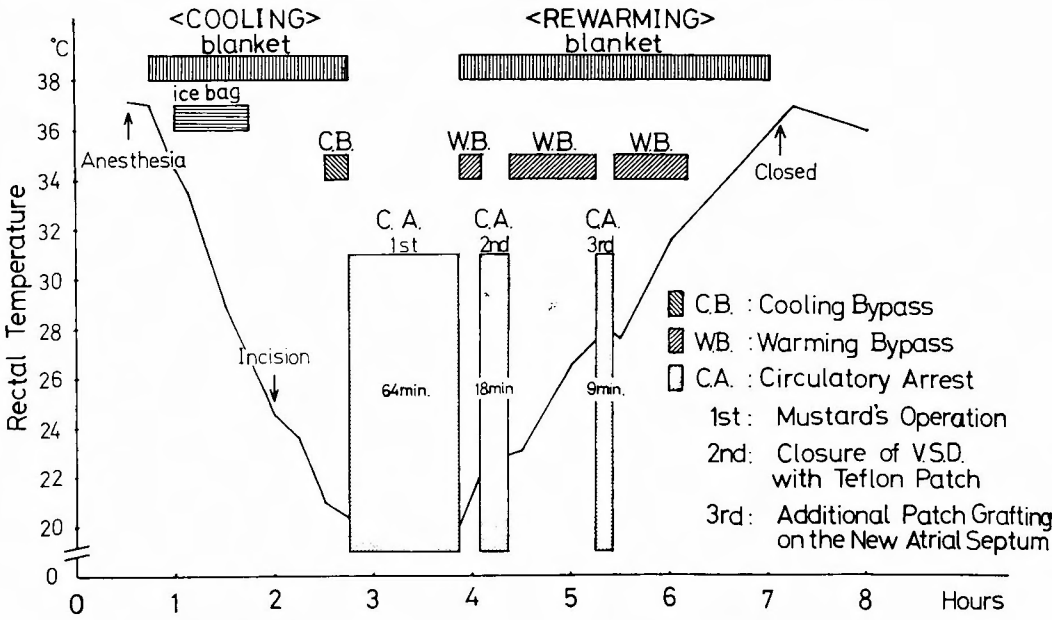
1) Premedication	Atropine sulfate	0.02mg/Kg	i. m.
	Pethilorfan	1~2mg/Kg	
2) Anaesthesia	halothane	(0.5~1.0%)	i. v.
	carbon dioxide	(1~3%)	
	nitrous oxide	(0~50%)	
	oxygen		
	Dialferine	0.5mg/Kg	
3) Chlorpromazine	0.5mg/Kg	i. v. drip	
4) Surface cooling (blanket + ice bags)	22~17°C (R. T.)		
5) Bypass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

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^{1,2)}. 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^{3,4)} 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 (P_p/P_s) was less than 0.75, the mortality rate was 0.9%, and there was no significant difference among the age groups. When P_p/P_s was greater than 0.75, especially when the pulmonary to systemic resistance ratio (R_p/R_s) 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

	0~12 mo.		12~24 mo.		2~38 yr.		Total	
	No.	M.R.	No.	M.R.	No.	M.R.	No.	M.R.
$P_p/P_s \leq 0.75$	27	(4)	39	(0)	146	(0.7)	212	(0.9)
$P_p/P_s > 0.75$								
$R_p/R_s \leq 0.5$	36	(11)	22	(9)	15	(7)	73	(10)
$R_p/R_s 0.5 \sim 0.75$	27	(19)	10	(10)	18	(6)	55	(13)
$R_p/R_s \geq 0.75$	16	(19)	9	(11)	9	(22)	34	(18)
Total	106	(12%)	80	(5%)	188	(3%)	374	(6%)

No. : Number of cases, M. R. : Mortality rate (per cent) (Sept., 1968 to July, 1974)

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)

Age (Years) Rp/Rs	Under 1	1	2 ~	Total
≤ 0.5	10 (0)	12 (0)	14 (0)	36 (0)
0.5 ~ 0.75	9 (0)	7 (0)	10 (0)	26 (0)
≥ 0.75	5 (1)	6 (0)	5 (1)	16 (2)
Total	24 (1)	25 (0)	29 (1)	78 (2)

() : Number of hospital deaths

hypertension 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 angiogram (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)

PA/AO	0~12 mo.		12~24 mo.		2~23 yr.		Total	
	No.	M.R.	No.	M.R.	No.	M.R.	No.	M.R.
Group I ≥ 0.5	4	(0)	6	(0)	25	(0)	35	(0)
Group II 0.3~0.5	8	(13)	12	(17)	37	(14)	57	(14)
Group III ≤ 0.3	6	(50)	2	(50)	14	(43)	22	(45)
Total	18	(22%)	20	(15%)	76	(14%)	114	(16%)

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

$$PA/AO = \frac{\text{diameter of pulmonary annulus}}{\text{diameter of aorta}}$$

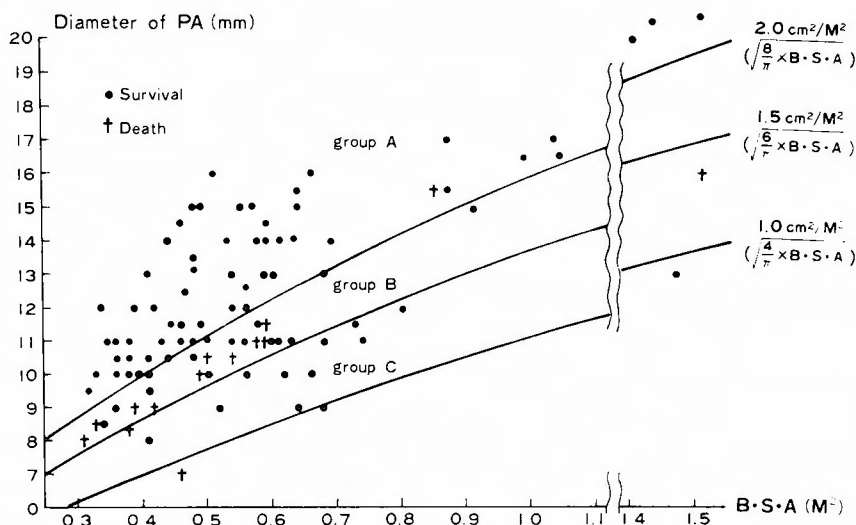


Fig. 2 Results of total correction of tetralogy of Fallot, in relation to degree of operative enlargement of pulmonary valve area.

Each curve, 2.0 or 1.5 cm^2/M^2 , shows pulmonary valve area per unit body surface area.

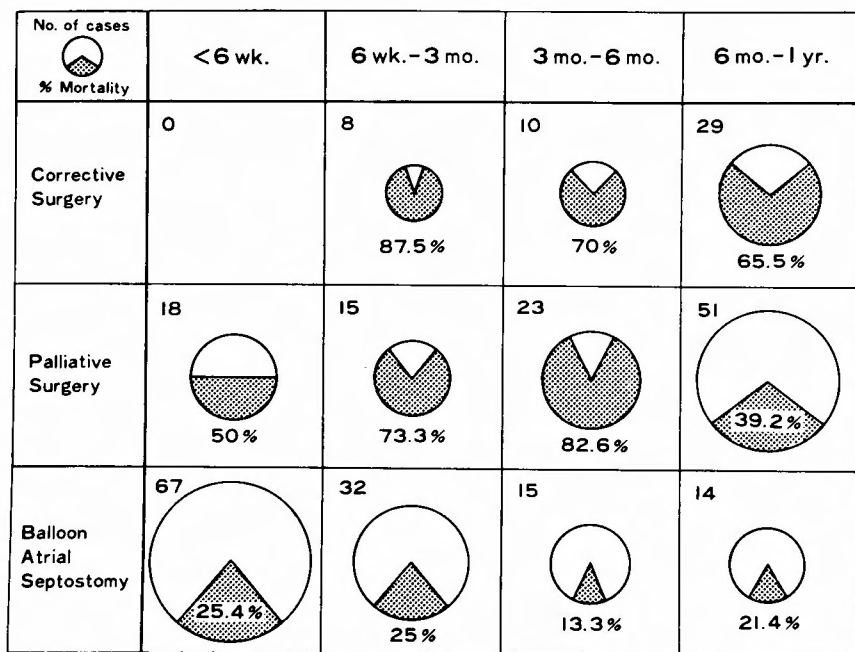
enlargement of the pulmonary annulus. The postoperative size of the pulmonary annulus is expressed in terms of pulmonary valve area per unit body surface area (PVA/BSA). The horizontal axis shows the BSA and the vertical axis the diameter of the pulmonary annulus. The two lines indicated as 2.0 and 1.5 cm^2/M^2 (pulmonary valve area per unit body surface area) divide the cases into three groups: group A with PVA/BSA greater than 2.0 cm^2/M^2 ; group B, PVA/BSA between 1.5 and 2.0 cm^2/M^2 ; and group C, PVA/BSA less than 1.5 cm^2/M^2 .

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 M^2 (over 4 or 5 years of age), 12 survived, while out of 26 patients with a BSA of less than 0.6 M^2 (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 cm^2/M^2 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 classification⁶⁾. In group I, the ventricular septum is intact and slight left



June, 1973.

Fig. 3 Surgery for complete transposition of the great arteries in the first year of life in Japan (from 32 main cardiac centers in Japan in June, 1973 studied by Dr. Atsushi Tanaka, Department of Pediatric Surgery, Juntendo University, School of Medicine)

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-Taussig 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 tamponade, 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

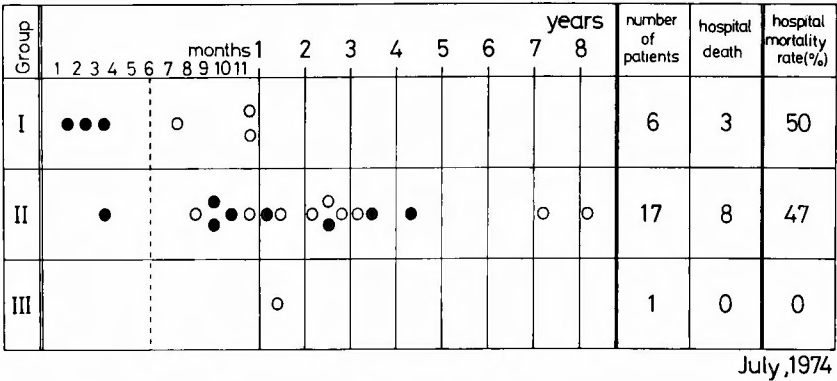


Fig. 4 Mustard operation for complete transposition of the great arteries (March, 1970 to July, 1974). Open circle=survival, Solid circle=death

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

Age	March, 1970 ~ 1971		1972 ~		Total	
	cases	died	cases	died	cases	died
< 1 Year	4	3	2	1	6	4
≥ 1 Year	2	1	9	3	11	4
Total	6	4 (67%)	11	4 (36%)	17	8 (47%)

in which pulmonary arteriolar resistance was over 2500 dynes sec cm⁻⁵. 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).

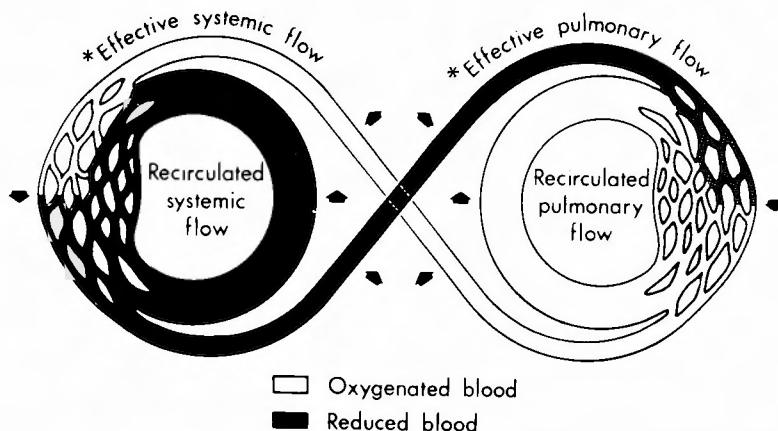


Fig. 5 Schematic representation of circulation in complete transposition of the great arteries (From Mair et al.¹¹)

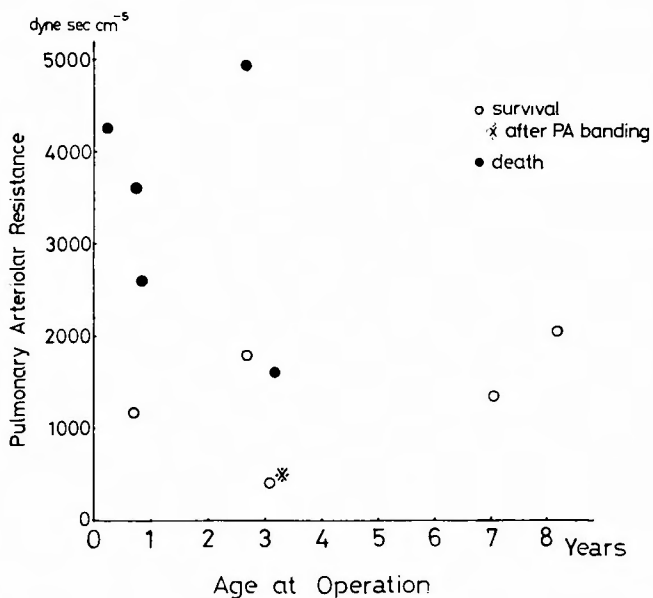


Fig. 6 Relation of pulmonary arteriolar resistance to age at time of total repair and operative results in complete transposition of the great arteries associated with ventricular septal defect

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

Pulmonary flow = O_2 consumption /
 (pulmonary venous O_2 content — pulmonary artery O_2 content)
 Effective pulmonary flow (QE) = O_2 consumption
 (pulmonary venous O_2 content — systemic mixed venous O_2 content)
 Effective pulmonary flow index (QEI) =
 effective pulmonary flow/body surface area

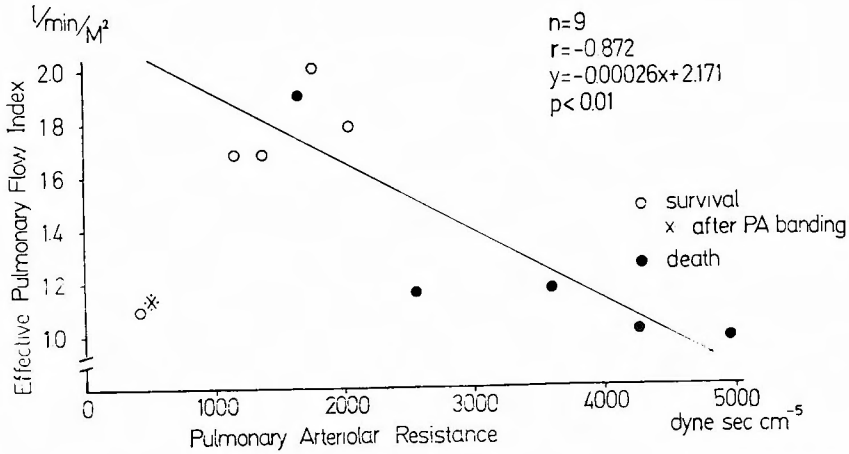


Fig. 7 Relation of pulmonary arteriolar resistance to effective pulmonary flow index and operative results in complete transposition of the great arteries associated with ventricular septal defect

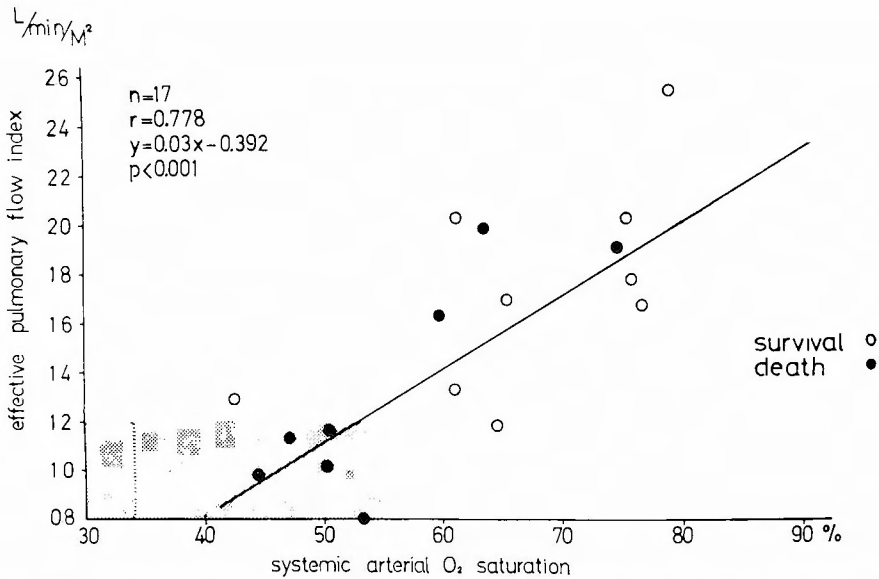


Fig. 8 Relation of systemic arterial oxygen saturation to effective pulmonary flow index and operative results in complete transposition of the great arteries associated with ventricular septal defect

Figure 6 shows the relation of PAR to age at operation and operative results. No patient with a PAR over 2500 $dyne\ sec\ cm^{-5}$ survived the operation. At autopsy, all these patients had changes of grade 4 or more according to the Heath-Edwards classification¹²⁾ in the pulmonary arterioles. It is quite interesting that there are babies who had a PAR over 4000 $dyne\ sec\ cm^{-5}$ at the age of three months or younger, while there are lucky patients who had a PAR under 2500 $dyne\ sec\ cm^{-5}$ 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

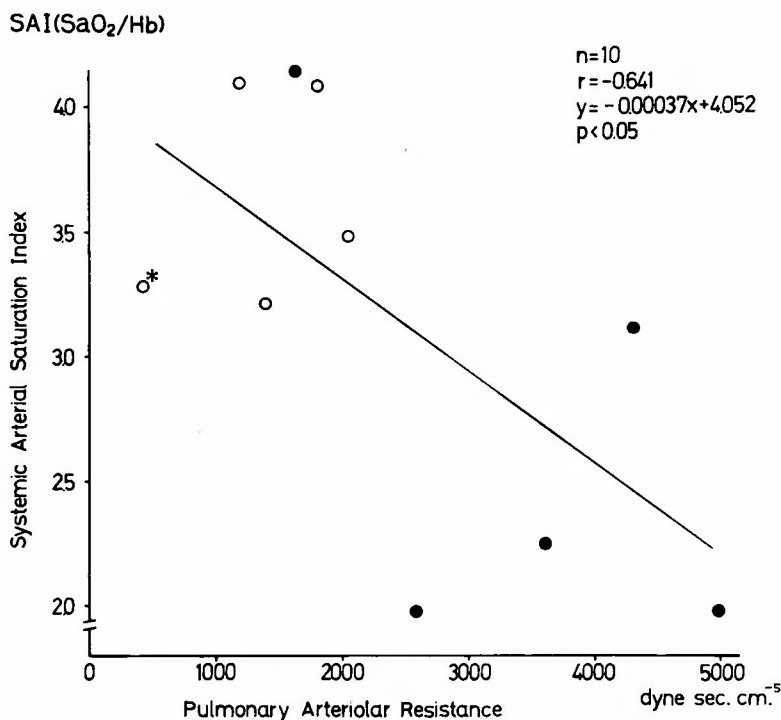


Fig. 9 Relation of systemic arterial oxygen saturation index to pulmonary arteriolar resistance and operative results in complete transposition of the great arteries associated with ventricular septal defect. *This patient had pulmonary artery banding prior to corrective surgery. SAI, systemic arterial oxygen saturation index. SaO₂, systemic arterial oxygen saturation (per cent). Hb, hemoglobin (gram per 100ml). Open circle=survival. Solid circle=death.

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/M^2 , 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.

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和文抄録

心室中隔欠損症・Fallot 四徴症、完全大血管転位症の乳児期一期的根治手術

兵庫県立尼崎病院 心臓外科

森 渥視, 安藤史隆, 節家直己, 山口勝雄
奥 秀喬, 神崎義雄, 河井 淳, 城谷 均

過去6年間に於いて兵庫県立尼崎病院心臓外科にて行なった生後12カ月未満の乳児期開心根治手術例は、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開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)が有利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に示したものである。以前は体重10kg以下の全例にこの低体温法を使用していたが1969年5月よりは体重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)、総肺静脈還流異常症(TAPVC)などのような心房内で手術操作を必要とする複雑心奇型に対しては前述の超低体温法を使用している。本論文では乳児期に開心根治手術が必要な諸疾患のうちVSD, TF, TGAの3疾患について、生後12カ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一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)。これを術前の肺・体動脈収縮期圧比(Pp/Ps)、および肺体血管抵抗比(Rp/Rs)にわけ、さらに年令別に考察してみると、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に他の年令層に比し有意に劣ってはいないことが判明する。すなわち乳児期に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78例のうちRp/Rs 0.75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。

以上より乳児期に内科的治療に抗するVSDに関して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なう方針をとっている。

2. ファロー四徴症(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)で分類した場合各年令間において手術成績にまったく差異をみとめない(表5)。われわれの乳児期死亡率の高かった理由はPA/AO比が0.3以下の重症例が非常に多かったためである。このことはTF根治手術成績が患児の年令や体重にはまったく関係なく、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ってきたが、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手術の方針をとっている。

さらに手術成績を術後の肺動脈弁輪部の拡大度とその患児の体表面積との相関であらわしたのが図2である。この図より手術時に右室流出路断面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ないし4才以下の年少例ほどこれ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

本論文の要旨は1974年8月26日より31日までコスタリカ共和国、サンホセ市において開催された日本医学セミナーにおいて発表した。

兵庫県尼崎市北城内27番地

分類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ただちに 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なわず乳児期でも I 群では Mustard 手術、II 群でも一期的に Mustard 手術と VSD 閉鎖を同時に行なっている。III および IV 群では通常生後 1 ないし 2 年間は支障なく生存す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カ月で肺動脈絞扼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。しかしながら手術手技、術前術後管理の改善により、1才以上の症例をも含めれば II 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、これらの報告例は大部分姑息的手術を以前に受けているので

その通算死亡率を考えればわれわれの成績は非常に良好といえる。I 群についてもわれわれの症例は大部分が初期の症例であり、最近では Stark も乳児期本症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なわれている。

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ることによるとされる。われわれは計算しえた 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 $2500 \text{ dynes sec cm}^{-5}$ 以上の症例はすべて死亡していることが判明した(図6)。さらに有効肺血流量係数 1.2 L/min/M^2 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、心臓カテーテル検査後この係数が次第に低下をしめ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。