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One Stage Corrective Surgery Early in Life for  
Tetralogy of Fallot, with Reference to  
Postoperative Cardiac Function

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Our indications for one stage total correction of tetralogy of Fallot and the results in 348 cases so treated at Kyoto University and Tenri Hospitals between 1964 and 1979 are reviewed here.

**Natural history and clinical classification**

Our early studies with serial angiocardiography have convinced us that tetralogy of Fallot is a progressively worsening disease. The ratio of pulmonary artery diameter to aorta diameter keeps decreasing (Fig. 1). Pulmonary peripheral vasculature also becomes more hypoplastic as the patient grows older. Therefore, the complete anatomical correction of tetralogy of Fallot becomes more difficult with age. Moreover, a gradual increase in the red cell count, hemoglobin value and right to left shunt were observed in serial examinations of the same patient (Fig. 2). Preoperative complications may be increased by such clinical disorders in older patients as hypoxic spells, thromboembolism, brain abscess, etc. Furthermore, much more serious postoperative complications, such as low cardiac output syndrome, pulmonary insufficiency, renal failure, etc., may occur in these

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Key words: Tetralogy of Fallot, One stage corrective surgery, Postoperative cardiac function, Kyoto technic, Pulmonary regurgitation.

索引語：ファロー四徴症，一期的根治手術，術後心機能，京都方式，肺動脈弁閉鎖不全。

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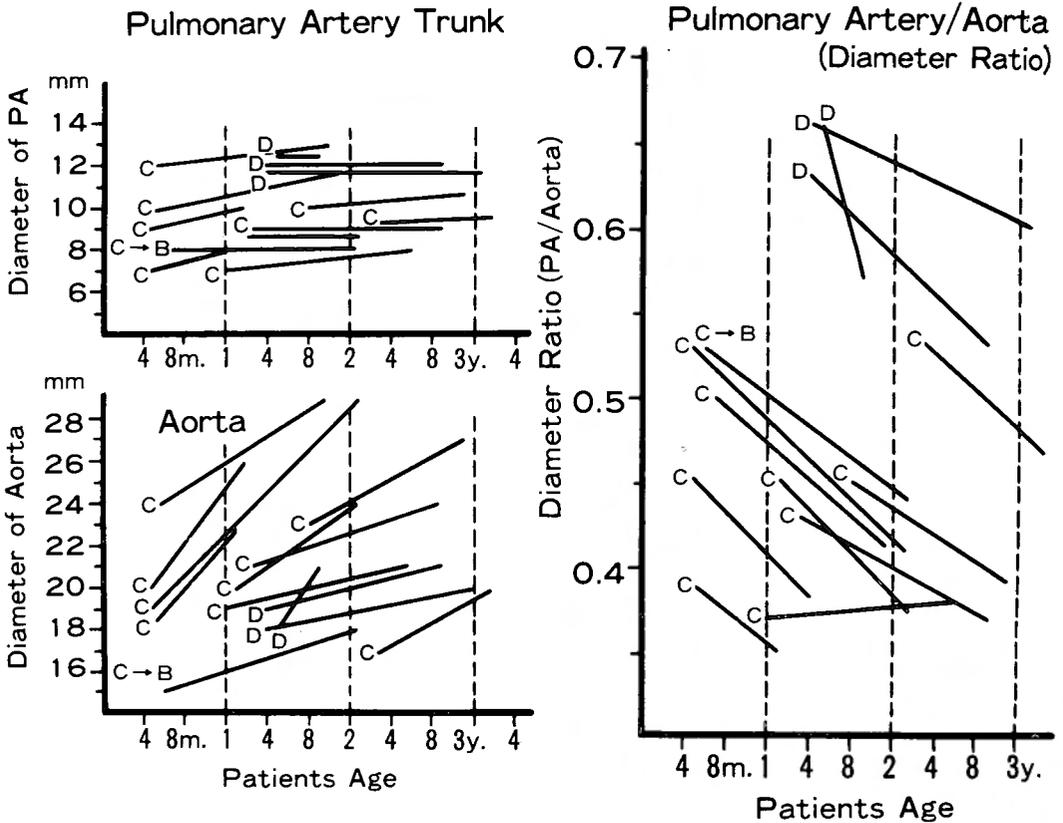


Fig. 1 Successive changes in diameters of main trunk of pulmonary artery and ascending aorta shown by serial angiocardiography in patients with tetralogy of Fallot.

The pulmonary artery does not grow because of the decreased blood flow, but the ascending aorta expands abnormally because of the right to left shunt.

Accordingly, the ratio of pulmonary artery diameter to aorta diameter keeps decreasing.

B : group B, C : group C, D : group D.

older patients. The severity of the disease, however, has a wide range from patients who die in early infancy to those who survive beyond adolescence.

We classified tetralogy of Fallot into four groups (A, B, C and D) according to the results of selective right ventriculography (Fig. 3). Group A patients had atresia of the pulmonary valve. Group B included patients with extremely severe hypoplasia of the pulmonary peripheral vasculature, pulmonary valve and outflow tract of the right ventricle (Fig. 3, 4). Group C comprised a wide variety of cases of tubular stenosis for various distances in the right ventricle, thickened hypoplastic pulmonary valve, a narrow pulmonary artery trunk, and sometimes branch stenosis. The pulmonary vasculature was scanty in group C. In group D, obvious localized muscular stenosis with a distinct third chamber was observed in the right ventricle.

Pulmonary valvular stenosis might or might not be present, but the pulmonary valve

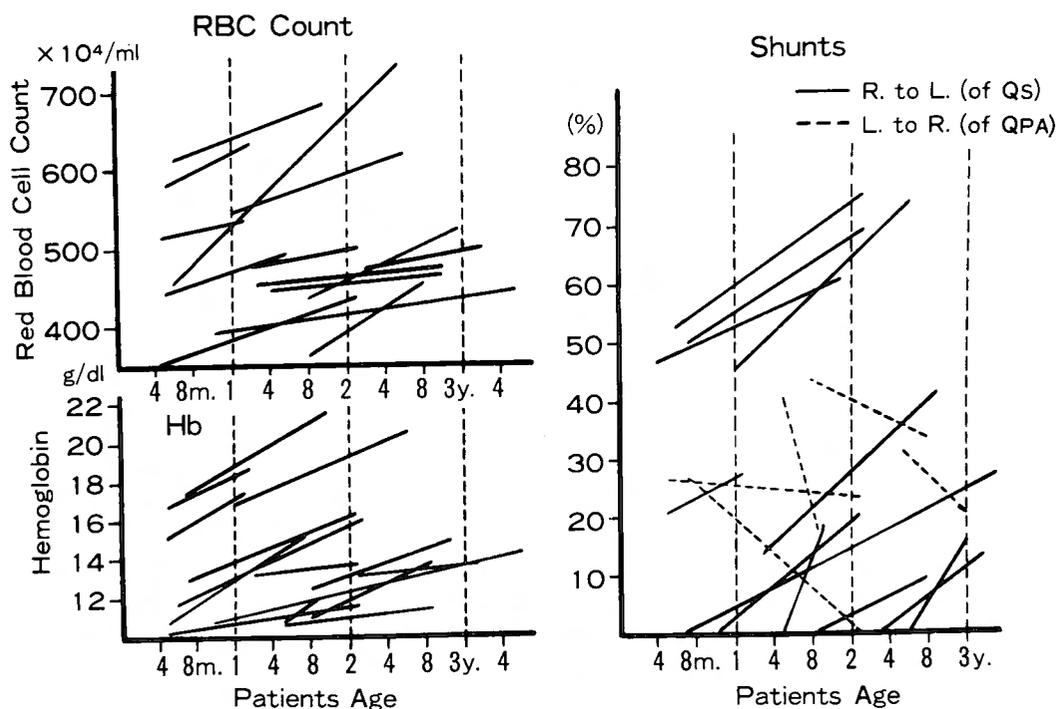


Fig. 2 Successive changes in red cell count in the same patients.

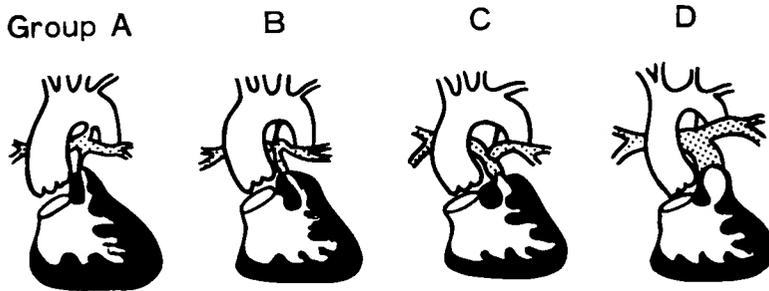
ring was usually almost normal in size, and the decrease in pulmonary vasculature was usually mild.

Fig. 3 shows the age distribution of the four groups in 1970. There are several characteristic findings. The majority of patients in group A are under four years of age, but several with sufficient collateral pulmonary blood flow survived until adolescence. There were no patients over five years of age in group B; three of the ten died during observation. In group C, the majority were under ten years of age, and one natural death occurred in a 15-year-old child. On the other hand, there were many adolescent patients in group D.

### Surgical policy and operative results

We based our surgical policy on the investigation mentioned above which indicated that corrective surgery should be done as early as possible in the treatment of tetralogy of Fallot. In group A, our choice of operation is an external conduit and a valve-bearing patch graft. However, a shunt operation is indicated in patients with severely hypoplastic pulmonary vasculature. Group B patients are the most difficult to treat surgically, since the mortality of corrective surgery is prohibitively high. A shunt operation seems to be the procedure of choice. However, the results of shunt operation are not completely satisfactory because of the poor run off in the peripheral pulmonary vascular bed. Corrective

## Age Distribution



Group	A	B	C	D	TOTAL
Age					
0	4 (1)	5 (2)	11	9	29
1, 2	3	2	16	7	28
3, 4	2	3 (1)	12	7	24
5, 6			6	4	10
7, 8	1		4	4	9
9, 10	1		5	4	10
11, 12				2	2
13, 14				1	1
15-20			2 (1)	2	4
21≤	1			5	6
TOTAL	12	10	56	45	123
Natural Death	(1)	(3)	(1)	(0)	(5)

Fig. 3 Our clinical classification of tetralogy of Fallot and age distribution.

surgery is usually recommended for groups C and D, and shunt operations are indicated for only a few patients in group C, who have extremely high hemoconcentration, such as a hematocrit above 70%.

We had already been doing a one stage corrective operation in infants with a large ventricular septal defect under deep hypothermia induced by surface cooling and core rewarming (Kyoto technic)<sup>6</sup>. We next used this technique in the corrective surgery of tetralogy of Fallot in infants<sup>7</sup>. This method was extremely useful in small babies weighing less than 10 kg, because the operative field appeared quite clear with complete elimination of bronchial blood return into the small heart during circulatory arrest.

Table 1 shows our surgical experience. Of the 348 patients treated with corrective surgery, 207 were under five years of age. The largest group of children under five years of age was in group C. In groups A and B, the patients were all under five years of age. The operative mortality in group B was prohibitively high (71%). The overall mortality

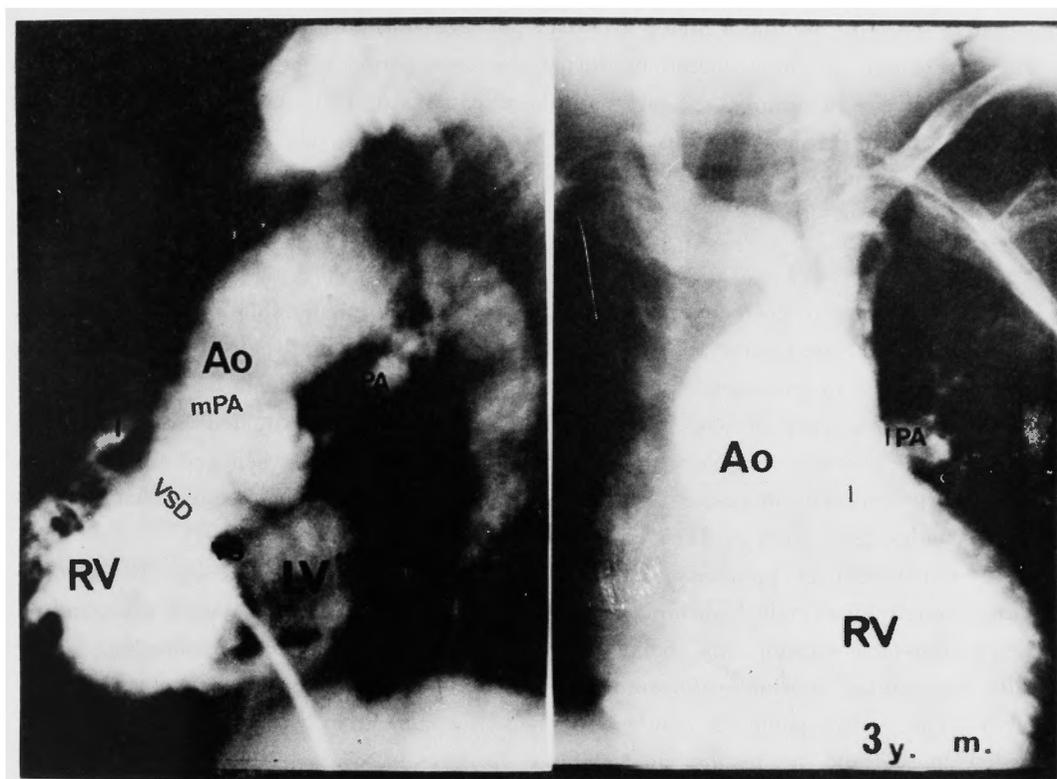


Fig. 4 Typical selective right ventriculography of a patient in group B.

PA : pulmonary artery    AO aorta,  
 RV : right ventricle,    LV : left ventricle,  
 I : infundibulum of right ventricle,    VSD : ventricular septal defect.

Table 1 Corrective surgery of tetralogy of Fallot (1964~1979)

	0 Y, 1-2Y, 3-4Y, 5-9Y, 10-14Y, 15Y,<						Total cases	Mortality	
								Over all (%)	Recent 5 years (%)
A Group		1(0)	1(0)				2(0)	0	0
B Group		2(2)	5(3)				7(5)	71	50
C Group	33(6)	41(8)	52(8)	45(9)	13(1)	2(1)	186(8)	18	9
D Group	17(3)	20(2)	35(3)	39(3)	20(0)	22(0)	153(11)	7	3
Total cases	50(9)	64(12)	93(14)	84(12)	33(1)	24(1)	348(49)	14	8
Mortality Over all (%)	18	19	15	14	3	4	14		
Recent 5 years (%)	15	9	9	7	4	0	8		

( ) : no. of deaths

in group C was 18% ; however, it was 9% during the last five years. Furthermore, the operative mortality for those under five years of age was 10% during the last five years. There were only 21 shunt operations during the same period : seven in group A, eight in group B and six in group C. Nearly all these patients (20/21) were under two years of age. The operative mortality was 14%, 50% and 0%, respectively in groups A, B and C; the overall mortality of shunt operations was 24%.

### Change in surgical treatment

The frequency of corrective surgery of tetralogy of Fallot in infants less than one year of age has decreased since 1970 because of improved conservative treatment to hypoxic spells especially in groups C and D with  $\beta$ -blocking drugs. Infants with severe polycythemia under one year of age (hematocrit over 70%) are mainly treated by shunt operations. Now corrective surgery with heart-lung bypass is done between one and four years of age in the majority of cases. Deep hypothermia is now used in the treatment of much more complex cases such as TGA, TAPVC etc.

The treatment of pulmonary stenosis has also been changing gradually. In the beginning, longitudinal right ventriculotomy without an outflow patch was used exclusively ; then a transverse incision was believed to preserve contractility of the outflow tract. After 1970, longitudinal incision reappeared in connection with the frequent use of the outflow patch. The outflow patch is now widely used in combination with minimal resection of cardiac muscle and has resulted in a definite decrease in surgical mortality and morbidity.

We are using a valve-bearing outflow patch in some cases to prevent postoperative pulmonary regurgitation. Pulmonary regurgitation caused by the use of the outflow patch may lead to deterioration of cardiac function in cases of severe hypoplastic pulmonary vasculature.

In these cases, pressure curves of the pulmonary artery and the right ventricle show similar patterns and can reach even higher levels than those of the systemic arteries because of severe pulmonary regurgitation and high resistance of the pulmonary vasculature after corrective surgery with the non-valved outflow patch. Weaning from the heart-lung bypass is impossible in this situation. We can reduce the deleterious effects of pulmonary regurgitation with the use of a monocusp-bearing outflow patch. The cusp is made of the patient's own pericardium to fit with native pulmonary cusps. This is effective, we believe, in the postoperative critical phase, although valve function had almost disappeared in many cases one month after operation. Complete prevention of pulmonary regurgitation is achieved with the use of a valve-bearing external conduit or prosthetic valve at the pulmonary valve ring.

We have been using a Björk-Shiley valve in the pulmonary position in corrective surgery for tetralogy of Fallot combined with complete absence of the pulmonary valve. The prosthetic valve is working well in the pulmonary position five years after operation and dilatation of the right ventricle has apparently been prevented. We prefer to use

mechanical valves rather than biological valves in children because of the superior durability of mechanical valves.

### Evaluation of postoperative cardiac function

In our clinical investigation of postoperative patients, right heart failure was observed most frequently in patients with residual severe pulmonary stenosis followed by those with residual ventricular septal defect and those with pulmonary regurgitation.

Fig. 5 shows the relationship between peak right ventricular systolic pressure and cardiac index at rest and during exercise. It is evident that residual severe pulmonary stenosis impairs cardiac performance; this tendency becomes more evident if tricuspid regurgitation is coexistent.

Our investigation of postoperative patients made it evident that left ventricular function is also impaired even many years after operation.

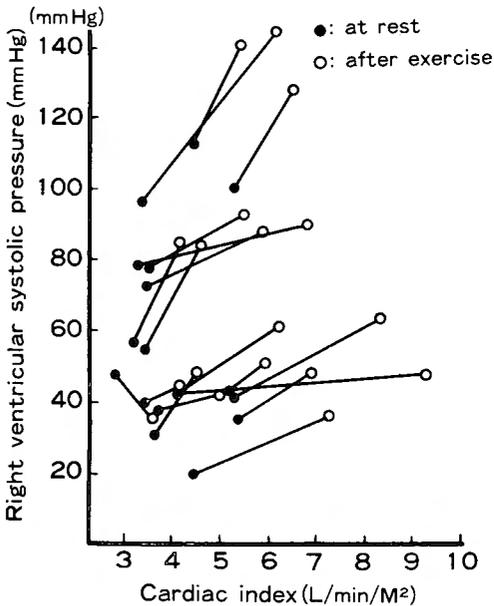


Fig. 5 Right ventricular performance after corrective surgery for tetralogy of Fallot

Only a slight increase of cardiac index is observed during the exercise test in spite of marked rise of right ventricular systolic pressure in patients with severe residual pulmonary stenosis who have high right ventricular systolic pressure at rest.

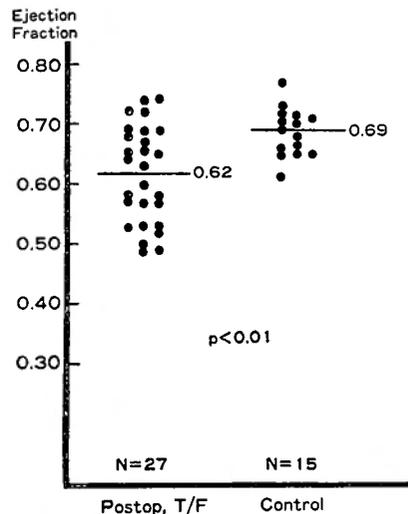
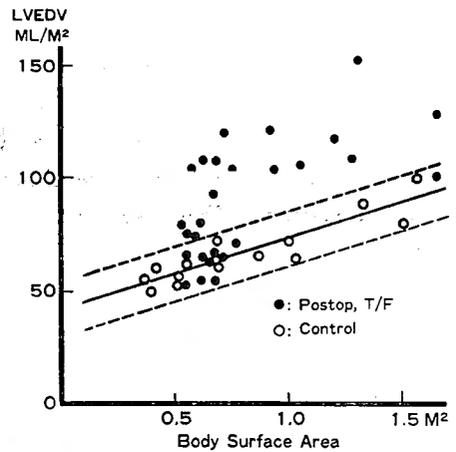


Fig. 6 LVEDV and LV ejection fraction in patients treated by corrective surgery for tetralogy of Fallot compared with those in controls without cardiac disease.

LVEDV: left ventricular end-diastolic volume.

Fig. 6 shows LVEDV and LV ejection fractions of postoperative patients in comparison with those of normal controls. The LVEDV increased significantly postoperatively, and the LV ejection fraction showed a significant decrease in patients with tetralogy of Fallot treated by corrective surgery.

Left ventricular contractility was also impaired significantly in our experience. All these tendencies to left ventricular functional impairment are evident in older children.

### Discussion

Recently, many authors have reported that the operative mortality of corrective surgery of tetralogy of Fallot is under 10%<sup>1)2)3)</sup>. However, there is a wide range in the severity of tetralogy of Fallot, from patients who will die in infancy unless a life-saving operation is performed to those who can survive until adolescence without surgery. Therefore, the operative results depend upon the clinical severity and age of the patients at the time of corrective surgery. They also depend upon the ratio of palliative operations to corrective surgery in infants.

Our investigations of the natural history of tetralogy of Fallot show that it is a progressively worsening disease (Fig. 1 and 2) and convince us of the necessity of early surgical treatment in selected cases (Fig. 3).

In considering which is better, one stage corrective surgery or a two stage operation, we find a curious similarity between tetralogy of Fallot and large ventricular septal defect with severe pulmonary hypertension, although, of course, there is a distinct difference between these two diseases, since ventricular septal defect has a high pulmonary blood flow and tetralogy of Fallot has a reduced pulmonary blood flow. However, pulmonary vascular changes progress to an irreversible point in both diseases without surgical treatment.

In the beginning, primary corrective surgery of large ventricular septal defect had a prohibitively high operative mortality. Then pulmonary banding in severely ill infants was proposed as a first stage operation to reduce pulmonary hypertension and postpone corrective surgery to a safer age<sup>10)</sup>. The operative mortality of corrective surgery reduced by this two-stage operation. Subsequently, improvements of medical therapy of infants with high pulmonary blood flow, of deep hypothermia technic.<sup>6)</sup>, and of heart-lung bypass made it possible to treat small babies with large ventricular septal defect by one stage corrective surgery is generally done under three years of age<sup>4)5)</sup>, and pulmonary banding is less commonly performed in cases of simple ventricular septal defect.

Our investigation of the natural history of tetralogy of Fallot showed a progressive polycythemia which increases blood viscosity, micro-thromboembolism in pulmonary arterioles and pulmonary vascular resistance. Shunt operations, which have a long history in the treatment of tetralogy of Fallot, can increase pulmonary blood flow and decrease vascular resistance with the development of pulmonary vasculature. However, there are many distinct differences between the results of successful corrective surgery and those of the shunt operation. When patients with similar pulmonary peripheral vasculature are opera-

ted on, some with corrective surgery and some with shunt operations, postoperative physical and social activities and physical development are all better in the former patients. Moreover, the latter must still face the risk of a second operation. If the operative risks of corrective surgery and a palliative procedure are not very different, it seems reasonable to choose corrective surgery.

Therefore, we have continued our efforts to reduce the risks of one-stage corrective surgery for tetralogy of Fallot with severe pulmonary vascular hypoplasia, and 348 corrective procedures were performed between 1964 and 1979 and only 21 shunt operations. The candidates for shunt operation were selected from patients under two years of age with severe hemoconcentration (hematocrit over 70%). The operative mortality of the shunt operation was very high (24%). The causes of death were postoperative pulmonary bleeding and obstruction of the anastomosis, probably because of hypoplastic pulmonary vasculature and poor run off of the peripheral pulmonary circulation. Furthermore, patients with severe hypoplastic pulmonary vasculature, as in our group B, were not helped much even by successful shunt anastomosis.

The operative mortality of corrective surgery for small infants with severe pulmonary vascular hypoplasia (our groups B and C) was also high at first, but it has been gradually decreasing with the changes in operative technique already mentioned and with improvement of heart-lung bypass. Deep hypothermia with surface cooling and core rewarming was extremely useful in providing a bloodless quiet operative field in small hearts, although heart-lung bypass is now used in almost all cases. During the last five years we have had reasonable results in our one-stage corrective surgery for children under five years of age in spite of our minimal use of shunt operations, although we have not yet solved the problem of how to treat patients with an extremely hypoplastic pulmonary peripheral vasculature (our group B). These cases might be candidates for heart-lung transplantation.

Although residual pulmonary stenosis is widely accepted as a limiting factor because of postoperative morbidity as well as operative mortality, there have been few reports on postoperative right heart function. Moller et al<sup>9)</sup> described exercise hemodynamics in severe cases of pulmonary valvular stenosis with fixed stroke index and elevated RVEDP. Our investigation of patients treated with corrective surgery for tetralogy of Fallot revealed that severe residual pulmonary stenosis impairs right ventricular performance especially during exercise (Fig. 5).

Pulmonary regurgitation causes decreased cardiac output in the immediate postoperative period in small babies with high pulmonary vascular resistance, especially in combination with tricuspid regurgitation. In this situation, a monocusp-bearing outflow patch, a valve-containing external conduit between RV and PA, or a prosthetic valve implantation in the pulmonary valve ring position prevents the deleterious effects of pulmonary regurgitation. A residual VSD and postoperative AV block are also factors in postsperative morbidity.

Several reports have described left ventricular function after corrective surgery of

tetralogy of Fallot. Jarmakani et al<sup>8)</sup> reported depressed LV function, although Sunderland et al<sup>11)</sup> found normal LV function. We noted an increase of the LVEDV and a decrease of the LV ejection fraction in postoperative angiocardiograms. The fact that these tendencies are more evident in older patients confirms the validity of early corrective surgery of tetralogy of Fallot.

### Summary

A total of 348 patients with tetralogy of Fallot treated by one stage total correction were reviewed and clinical classification, operative indications, operative techniques, including assist devices, and postoperative evaluation of cardiac function were described.

For the determination of surgical indications, we have classified tetralogy of Fallot into four groups (A, B, C and D) based on the findings of selective right ventriculography. While great difficulties remain in treating small babies with severely hypoplastic pulmonary vasculature (our group B) by total correction or shunt operation, the operative mortality of one stage correction in the other three groups (A, C and D) has recently become reasonably low (under 10%) in spite of a very small number of shunt procedures.

Primary total correction during deep hypothermia with surface cooling and core rearming (Kyoto technic) has long been the procedure of choice for small infants weighing less than 10 kg. Although the number of patients treated with the Kyoto technic is currently decreasing because of the progress in the medical treatment of small infants with anoxic spells, our policy for the management of tetralogy of Fallot is a one stage total correction in early life, as before.

Pulmonary regurgitation causes serious problems immediately after operation in cases of high pulmonary vascular resistance. In this situation, a monocusp-bearing outflow patch or prosthetic valve in the pulmonary valve position proved effective in tiding the patients over the critical postoperative phase.

A longterm evaluation of the survivors revealed that residual severe pulmonary stenosis presented serious problems especially for right ventricular function, and an increase in the left ventricular pump function after corrective surgery in older children.

In conclusion, our investigation of the natural history, operative results and postoperative cardiac function suggests that patients with tetralogy of Fallot should be treated by one stage corrective surgery between one and four years of age as far as possible.

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## 和文抄録

### ファロー四徴症に対する早期一期的根治手術， および術後心機能について

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ファロー四徴症は加齢と共に悪化する疾患であり早期外科治療を要する。1964年から1979年の間に根治外科手術の施行された348例を検討した結果を報告する。

超低体温麻酔完全循環遮断下の乳幼児開心術はファロー四徴症に対しても甚だ有効であるが、最近の保存的治療法の進歩に伴ない、1才以下で根治手術を必要とする症例は減少している。

しかし一方では肺動脈血管床の強い発育不全を伴な

う乳幼児症例は、その根治手術成績が不良であるばかりか、緩解手術成績も決して良好ではなく外科治療上大きい問題が尚残されている。

ファロー四徴症の自然歴、手術成績、術後心機能などの検討の結果、根治手術は1～4才の間に行われるべきであり、術後の強い肺動脈狭窄および閉鎖不全、心室中隔欠損遺残、心ブロックなどを残さぬ事が肝要であるとの結論を得た。