

Total Correction of Tetralogy of Fallot (under Extracorporeal Circulation)

by

YORINORI HIKASA, HITOSHI SHIROTANI, KUREO TSUSHIMI, HISAAKI KOIE,
JUN TAKEDA, NORIKAZU TATSUTA, JUN KAWAI and TOSHIHIKO BAN

From the 2nd Surgical Division, Kyoto University Medical School
(Director : Prof. Dr. CHUJI KIMURA)

MANNOSUKE MUNHEYUKI, YOSHIYUKI ASAWA, KIKUO NIŌ
and HITOSHI MITANI

From the Department of Anesthesiology, Kyoto University Medical School
(Director : Prof. Dr. AKIRA INAMOTO)

CHUZO MORI, TOKIO TAMURA and TETSURO KAMIYA

From the Department of Pediatrics, Kyoto University Medical School
(Director : Prof. Dr. HIDEO NAGAI)

Received for Publication Apr. 30, 1966

I. FOREWORD

Since safeness of selective heart cooling with coronary perfusion was assured experimentally by us one year ago, eight cases of tetralogy of FALLOT were operated radically in our clinic with selective cardiac arrest induced by the above method, under extracorporeal circulation. Another two cases, weighing below nine kilograms were repaired under deep hypothermia employing the surface cooling method (HIKASA-SHIROTANI's method). Altogether ten cases of tetralogy of FALLOT were operated radically and only one patient expired (Table 1).

Table 1. Result of Radical Operation of Tetralogy of FALLOT.

Case	Initial	Age (yr.)	Sex	Weight	Patch	Procedure employed	Result
1	F.A.	15	♂	48kg	teflon	extracorporeal circulation	alive
2	O.S.	12	♂	21kg	teflon	extracorporeal circulation	alive
3	I.Y.	14	♂	33kg	teflon	extracorporeal circulation	alive
4	M.M.	5	♂	19kg	teflon	extracorporeal circulation	dead
5	T.H.	7	♂	18kg	pericard	extracorporeal circulation	alive
6	Y.U.	6	♂	16kg	pericard	extracorporeal circulation	alive
7	K.M.	13	♂	39kg	pericard	extracorporeal circulation	alive
8	N.A.	11	♂	33kg	pericard	extracorporeal circulation	alive
9	S.M.	6/12	♂	6kg	pericard	deep hypothermia	alive
10	H.S.	15/12	♂	8.8kg	pericard	deep hypothermia	alive

II. CRITERIA OF DETERMINING SURGICAL INDICATION

Table 2. PA/AO Ratio of the Case with Tetralogy of Fallot on which Radical Surgery Was Performed.

PA/AO ratio	Number of Cases	
	extracorporeal circulation	deep hypothermia
0.30~0.39	0	1
0.40~0.49	0	0
0.50~0.59	3	0
0.60~0.69	2	0
0.70~0.79	1	1
0.80~0.89	2	0
total	8	2

The ratio of pulmonary arterial diameter to aortic diameter was measured by means of angiocardiogram. Those showing the ratio above 0.5 were roughly considered as indicating radical surgery. Being prepared for the surgery of radical repair, the patient is placed in FOWLER'S position and exploration of the heart is done. Indication of radical operation is finally confirmed by measuring directly the size of the each vessels. As shown by Table 2, this PA/AO ratio was above 0.5 in each case of older children who were treated under extracorporeal circulation with the heart lung

machine. One case of an infant who was operated under deep hypothermia revealed the PA/AO ratio of 0.33 (1/3), and was repaired successfully. The possible extension of this indication to elder children is to be considered seriously, and further study seems to be needed.

III. PROCEDURE OF RADICAL OPERATION

After the heart is exposed and is measured as described above, complete extracorporeal circulation is started. Decompression procedure of the left atrium is performed through interatrial septum, opening the right atrium first. Then, selective heart cooling

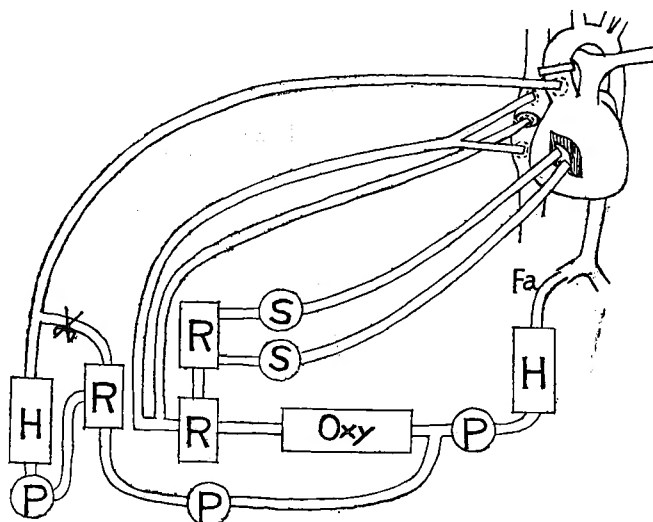


Fig. 1 Circuit of Selective Heart Cooling with Coronary Perfusion
 H : Heatexchanger, Oxy : Oxygenator, P : Pump,
 R : Reservoir, S : Suction pump, Fa : Femoral
 artery

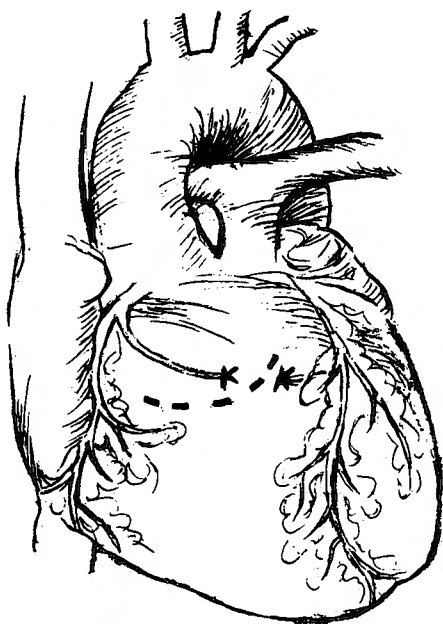


Fig. 2 Transverse Incision of Right Ventricle
(GERBODE)

with coronary perfusion is started by the circuit, as shown in Fig. 1. 5~10 cc/kg/min. of blood which is preparatorily cooled down to 4~5° centigrade is thus perfused under the pressure not exceeding 100 mmHg. When complete cardiac arrest is induced, a transverse incision as shown in Fig. 2 is placed to right ventricular wall. The purpose of the right atriotomy is to have a dry field in the right ventricle, by suctioning the return of coronary perfusate, besides making sure of the absence of ASD. Therefore, the right atrium is left open during the manipulation in the right ventricle. Selective coronary perfusion is continued intermittently in the duration of 15 to 20 seconds every ten minutes for the purpose of preventing the elevation of myocardial temperature, restart of heart beat and occurrence of metabolic derangement of the myocardium itself. The

right ventricle is incised transversely as described above for the purpose of giving least injury to the myocardium and of protecting from restenosis of the outflow tract when closing the ventriculotomy wound. In the first three cases, by resection of stenotic conus, outflow jungle was resected extensively, involving the right ventricular wall, in the extent from considerably central of the right ventricle up to the pulmonary valvular ring area. Extension of this resection was so wide that the total mass of resected muscles weighed averagely eight to ten grams and satisfactory dilatation of the outflow tract was thus attempted. Closure of the defect of ventricular septum was done with Teflon cloth which was sutured with about twenty individual stitches (Fig. 3). Postoperatively, in all three cases, blood pressures were not maintained at a level of 15 to 20 cmH₂O for venous pressure and 90 to 100 mmHg for arterial pressure, without the help of continuous intravenous dripping of Isuprel (Fig. 4). Even with the aid of Isuprel, maintenance of blood pressure to such levels was rather difficult. The amount of blood drained from the thoracotomy tubes totaled to 1,000 to 1,650 cc by the time of removal of the tube.

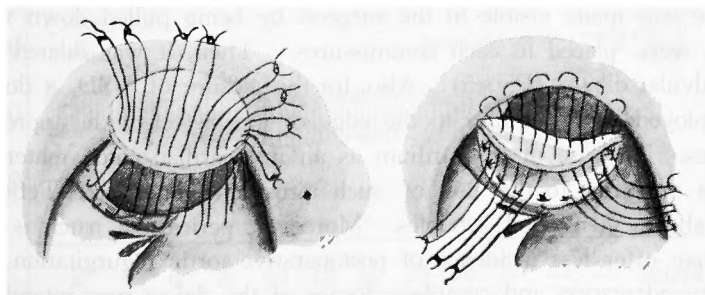


Fig. 3 Closure of Ventricular Septal Defect

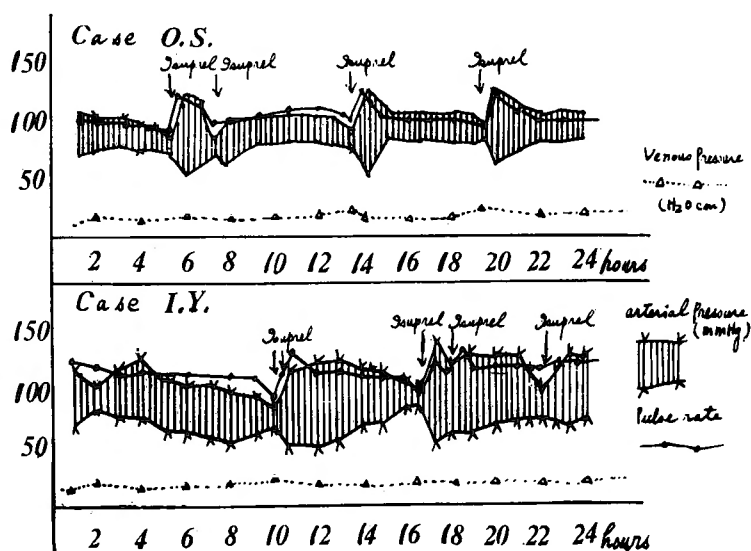


Fig. 4 Postoperative Course in Early Cases

The amount of fluid retention in the chest cavity thereafter, reached to 740 to 1,160 cc, thus necessitating puncture and drainage. Case 4 was unfortunately lost during the course of the same operative procedure as the above three cases, due to the injury of left branch of the coronary artery while repairing accidental injury of the pulmonary arterial wall. However, the experience of the above four cases gave us the following suggestions. 1) Attempt of thorough dilatation of the outflow tract tends to invite excessive resection of muscle pieces and to consequent right heart failure postoperatively. 2) Closure of VSD tends to be unsatisfactory with the use of Teflon cloth due to the possibility of a leakage of blood through its mesh for a time, after suturing. And there also is a tendency of incomplete adaptation of Teflon cloth to the edge of the defect. 3) With GERBODE's transverse incision, higher possibility of injury of the pulmonic ring area is suspected when manipulating around that area for dilatation, since operative field goes too deep. So, in all the cases after the fifth, including those cases repaired under deep hypothermia, removal of stenotic part around the conus, where stenosis is usually significant, were attempted as circumscribed as possible, when manipulating for the dilatation of the outflow tract. And removal of muscle was carefully performed not to go too far to the central part of the right ventricle. Altogether only about two to three grams of muscle pieces were resected. Pulmonic valve was made visible to the surgeon by being pulled down with a hook and small incisions were placed to each commissures. Then, it was dilated more by means of pulmonic valvular dilator (Fig. 5). Also, for the closure of VSD, a doubled pericardial patch was employed being sutured to the edge of the defect with approximately 20 individual stitches. The use of pericardium as an intracardiac patch material was preferred for the closure of VSD to the use of such prosthetic ones like Teflon cloth, which revealed the above mentioned troubles. Moreover, pericardial patch is considerably extensible and may offer less incidence of postoperative aortic regurgitation. By this technique, satisfactory adaptation and complete closure of the defect was intended. At the end



Fig. 5 Dilatation of Pulmonary Valvular Ring Area by Means of Pulmonic Valvular Dilator

of extracorporeal circulation, assist perfusion was performed for a time, roughly equivalent to the duration of previous heart arrest. With such devices, in all these cases the necessity of administering Isuprel and similar drugs postoperatively had disappeared. It became possible to maintain venous pressure below $15 \text{ cmH}_2\text{O}$ and arterial pressure around 120 mmHg (Fig. 6). In addition, the amount of blood drained postoperatively from the thoracotomy tubes came to remain below $1,000 \text{ cc}$. Moreover, the retention of pleural fluid ceased to occur at all. Furthermore, by such improvements of operative techniques, total by-pass time had come to shorten markedly (Fig. 7). Comparison of oxygen saturation of arterial blood before and after the repair is shown in Fig. 8. After performing dilatation of outflow tract in the above manner and while the pericardial patch is being sutured

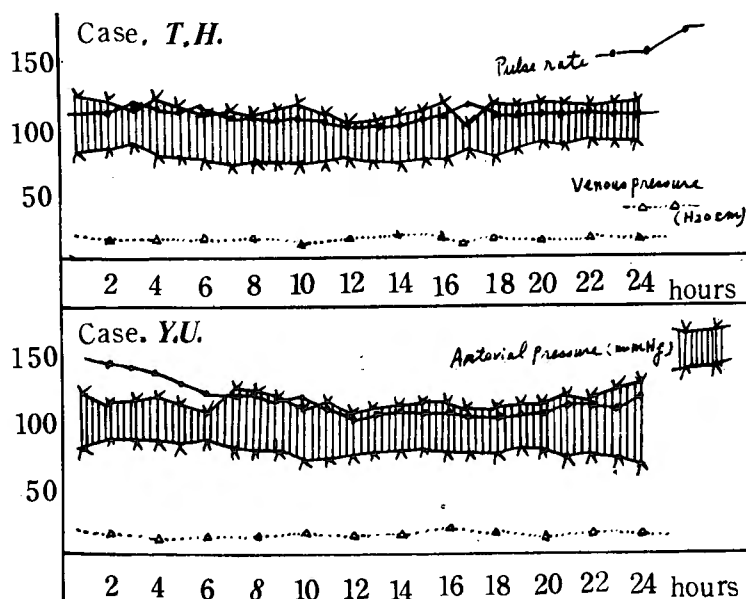


Fig. 6 Postoperative Course in Recent Cases

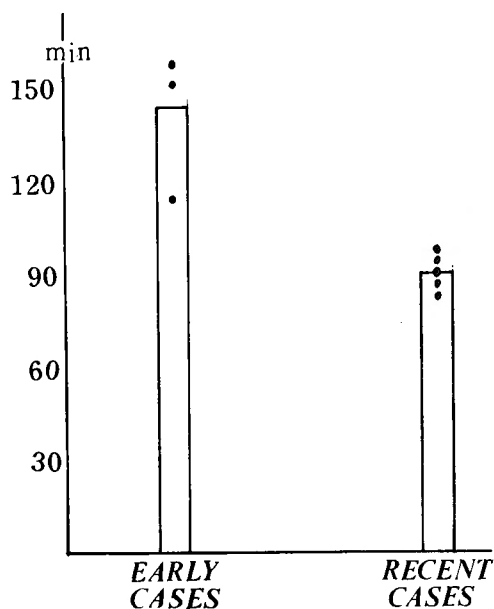


Fig. 7 Total-by-pass Time in Radical Surgery of Tetralogy of FALLOT

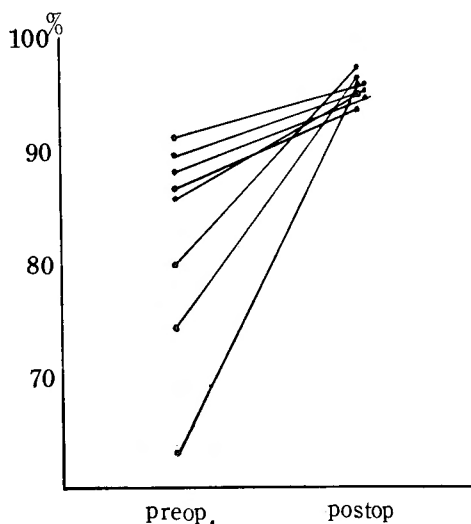


Fig. 8 Arterial Oxygen Saturation before and after Surgery

to VSD, the aortic clamp, which has been placed for selective coronary perfusion, is removed, and rewarming of the heart is attempted. By this procedure, the heart beat returned to normal quite smoothly in all cases without need of employing electric shock. The right ventriculotomy wound is closed by individual mattress, then over and over sutures. Extubation of the left auricular decompression cannula, closure of the interatrial hole through which this cannula has been inserted, then closure of right atriotomy wound are performed. And the radical operation is accomplished.

Due to the need of considerably long-term extracorporeal perfusion, enough attention is paid to the following points. 1) The patient is placed on FOWLER'S position and attention is paid to SVC pressure, not to exceed 30 cmH₂O at most, during the entire course of perfusion. 2) The suction system contributes to the cause of marked destruction of the blood. So, the decompression system of the left atrium is used as much as possible, instead. 3) Before and after extracorporeal circulation, maintenance of a normal acid base balance is to be attempted by positive and negative pressure breathing by respirator. In addition, during extracorporeal circulation, 4% sodium bicarbonate solution was added in an adequate amount and it was deliberately attempted to maintain a normal acid base balance. This solution was administered as a rule, by the amount of 1.0 cc/kg for the depression of 0.05 of blood pH. 4) Moreover, for the purpose of preventing the occurrence of renal shut down, mannitol was added preliminary into the priming blood at the amount of two grams per one kilogram of the patient's body weight.

In addition, for the radical operation of tetralogy of FALLOT, the following points are deliberately considered. 1) In this malformation, collateral circulation from bronchial artery system is usually developed. Therefore, it is necessary to notice well that a large amount

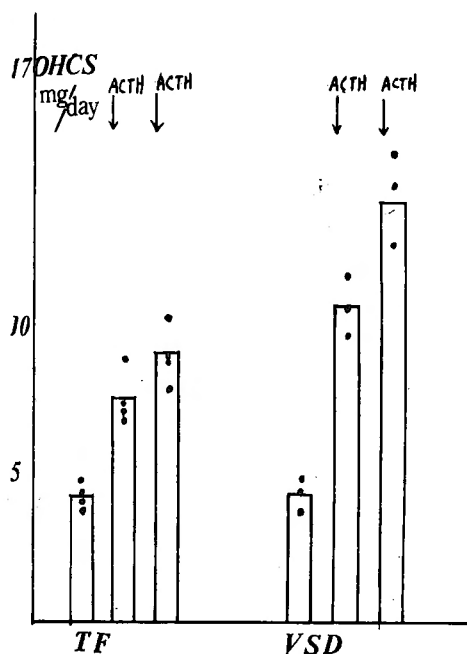


Fig. 9 Result of ACTH-Test, Performed before Surgery. (Comparison between TF Group and VSD Group)

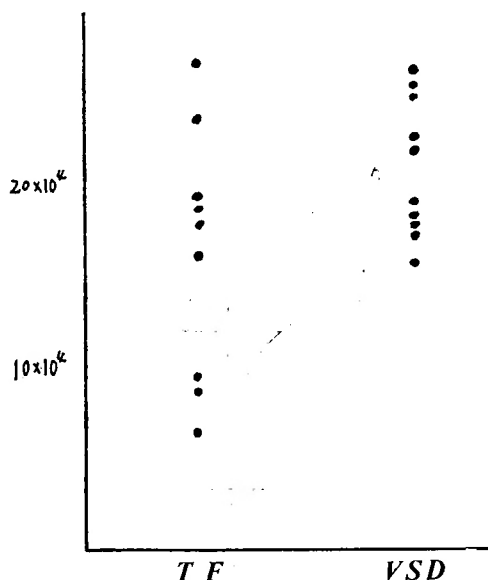


Fig. 10 Thrombocyte Count in the Blood (Comparison between TF Group and VSD Group)

of blood escapes through this circulation system when performing extracorporeal perfusion. From this standpoint, heart lung by-pass currently has been done with such high flow volume as 120 cc/kg/min.. And it was attempted to maintain the mean arterial pressure above 60 mmHg and venous pressure preferably below 20 cmH₂O. 2) In this malformation, the cases with hypoadrenocortical function are occasionally seen (Fig. 9). So, if needed, the administration of hydrocortison for two or three postoperative days is to be remembered. 3) Also, as a characteristics of tetralogy of FALLOT, a decrease of thrombocyte count in the blood below 100,000, is occasionally seen (Fig. 10). So, a preliminary administration of Vitamin K₁ preparation before the operation is to be done sufficiently. Moreover, Ipsilon (ϵ -Aminocaproic acid) and PVP are always added in priming blood at the amount of one volume for ten volumes of fresh blood and extracorporeal circulation is performed with it. Postoperatively, if needed, such hemostatic drugs like Vitamin K₁ preparation, Ipsilon, Thromboplastin preparation, Fibrinogen, and so on are used. 4) It is assumed by us, that dilution of priming blood with low molecular dextran or amino acid solution is to be avoided. Because, in tetralogy of FALLOT, even though there exists a concentration of blood, such changes like decrease of thrombocyte count is already seen preoperatively. Consequently bleeding tendency is rather manifest compared with the other congenital heart diseases. 5) As to supplying fluid, 500 cc/m² of fluid was given postoperatively in 24 hrs. following the way which STURTZ and others recommended. In the next and following 24 hours periods, 750 cc/m² of fluid was given in each period. For this purpose, 10% D/W was used. 6) In every case, pericardial space was opened enough

to right chest cavity by dividing both right pericardium and mediastinal pleura in sufficient length. Thoracotomy tubes were inserted not only to the anterior mediastinum but also to the right chest cavity and continuous postoperative drainage was attempted.

IV. SUMMARY

As was described above, since the indication for radical operation of tetralogy of FALLOT and the technique of selective heart cooling by coronary perfusion were established experimentally in our department one year ago, ten cases of tetralogy of FALLOT were operated radically. Among them only one case was lost, and this result seems roughly satisfactory. So, the course of improvement and progress which enabled this result, was discussed mainly in relation with operative procedures.

Table 3. Findings before and after Surgery.

		Befroe op.	After op.
Blood Cell Count	Erythrocyte	648×10^4	459×10^4
	Ht	50.5%	44%
Arterial Oxygen Saturation (F.A.)		75.2%	94.6%
Circulating Blood volume		2.25 l	3.62 l

(Age : 19 yr., Sex : ♀, Weight : 50kg)

course (Table 3). We are now coming to have an impression that our present criteria for surgical indication as mentioned early in this report, may be revised.

REFERENCES

- 1) Abe, K. : Experimental studies on bleeding diathesis not uncommonly accompanied with extracorporeal circulation. Arch. Jap. Chirurgie, **31** : 431, 1962.
- 2) Ban, T. : Über Blutgerinnungsstörungen beim extrakorporalen Kreislauf, mit besonderer Berücksichtigung der Frage des Perfusionsblutes und des gefahrlosen Durchführens der längeren Perfusion. Arch. Jap. Chirurgie, **35** : 1966 (to be published).
- 3) Bahnson, H. T. et al. : Surgical treatment and follow-up of 147 cases of tetralogy of Fallot treated by correction. J. Thorac. Cardiovasc. Surg., **44** : 419, 1962.
- 4) Ehrenhaft, J. L. et al. : Evaluation of results after correction of tetralogy of Fallot. J. Thorac. Cardiovasc. Surg., **45** : 224, 1963.
- 5) Kirklin, J. W. et al. : Factors affecting survival after open operation for tetralogy of Fallot. Ann. Surg., **152** : 485, 1960.
- 6) Klinner, W. : Operativ-technische Probleme der Radikaloperation bei Fallotscher Tetralogie. Thoraxchirurgie, **9** : 274, 1961.
- 7) Klinner, W. : Klinische und experimentelle Untersuchungen zur operativen Korrektur der Fallotschen Tetralogie. Ergebnisse der Chirurgie und Orthopädie, **44** : 58, 1962.
- 8) Lillehei, C. W. et al. : Complete anatomic correction of the tetralogy of Fallot defects. Arch. Surg., **73** : 526, 1956.
- 9) Kirklin, J. W. et al. : Surgical treatment for the tetralogy of Fallot by open intracardiac repair. J. Thorac. Surg., **37** : 22, 1959.
- 10) Malm, J. R. et al. : An evaluation of total correction of tetralogy of Fallot. Circulation, **27** : 805, 1963.
- 11) Ross, D. N. : The surgical treatment of Fallot's tetralogy. Thoraxchirurgie, **9** : 267, 1961.
- 12) Tsushimi, K. : Experimental and clinical studies on open heart surgery using blood stream cooling. Arch. Jap. Chirurgie, **34** : 704, 1965.
- 13) Zerbini, E. J. et al. : Total correction of complex of Fallot under extracorporeal circulation : Immediate results in a group of 221 patients. J. Thorac. Cardiovasc. Surg., **49** : 430, 1965.