

Long-Term Hemodynamic Evaluation of Primary Total Correction of Tetralogy of Fallot During The First Two Years of Life

by

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Received for Publication, Aug. 31, 1973

Introduction

Primary total correction of symptomatic tetralogy of Fallot in children less than two years of age has been performed since 1964 in Kyoto University Hospital, using our technique of deep hypothermia with surface cooling and partial cardiopulmonary bypass¹⁾⁻⁶⁾. There has been only one operative death in 26 procedures during this time. Operative mortality is no longer the limiting factor for performing total correction of this lesion in this age group. The long-term hemodynamic and anatomical results are now the most important factors which determine whether the early primary total correction is more advantageous than the conventional two stage operation.

Late hemodynamic results of tetralogy of Fallot under 2 years of age have been reported only by Starr and his associates⁷⁾⁸⁾. They often used an outflow patch to relieve the pulmonic stenosis. We did not use an outflow patch in our series of 26 patients even in cases of markedly hypoplastic outflow tract of the right ventricle and the pulmonary artery.

The purpose of this paper is to describe the late postoperative hemodynamic and angiocardigraphic findings in 15 children who underwent total correction in their first two years of life.

Material and Method

From December 1964 through December 1972, twenty-six patients younger than two years of age underwent corrective operation for tetralogy of Fallot as the primary procedure in Kyoto University Hospital. The patients ranged in age from 3 to 23

months with an average of 16.3 months, and their weight at the time of surgery ranged from 4.9 to 11.8 kg with an average of 8.9 kg.

Preoperative right heart catheterization with or without combined left heart catheterization was performed in all 26 patients. Except for the first 5 patients selective biplane angiocardiology and cine-angiocardiology were also performed. The diagnosis of tetralogy of Fallot was established preoperatively by these examinations.

All 26 patients exhibited cyanosis at rest, and 19 out of 26 patients experienced anoxic spells. Medical treatment including Propranolol was usually effective to reduce the frequency and severity of these spells, however, 4 of 19 patients did not respond to any medical measures including administration of Propranolol, sedatives, narcotics, oxygen and fluid therapy. Primary total correction was performed as an emergency procedure to these 4 patients.

In all but the first patient the technique of deep hypothermia, which has been developed by our group, was used. This procedure consists of surface cooling, total circulatory arrest, and use of partial cardiopulmonary bypass. The partial bypass was used at the terminal stage of the cooling process, for cardiac resuscitation, and central rewarming. The first patient was operated upon using simple deep hypothermia, that is, surface cooling and surface rewarming.

There was only one hospital death out of 26 patients. One late death occurred 2 years and 2 months after surgery, unrelated to the operation.

All patients had similar operations. A transverse right ventriculotomy was used. Infundibular stenosis was relieved mainly by wedge resection of the medial muscle of the infundibulum, more specifically the parietal band and the right portion of the crista supraventricularis. The anterior wall of the right ventricular outflow tract was thinned by excising the deeper layer of the myocardium as necessary. However, the septal band was left intact in most cases to maintain the proper postoperative right ventricular contraction. The valvular stenosis, if any, was relieved by incising the commissures at 2 or 3 points. The small annulus of the pulmonary valve, which was the most formidable type of obstruction, was incised medially and laterally and was dilated as much as possible using a miniature type of Gerbode's tranventricular mitral valve dilator and/or Hegar dilators. No outflow patches were inserted even in the cases of severely hypoplastic pulmonary valve annuli, as small as 5 mm in inner diameter or one-third of the diameter of the aortic root. The ventricular septal defect was closed in all cases with duplicated autologous pericardial patch. Intracardiac pressures were measured with a strain-gauge manometer after the repair before the chest was closed.

Result

Postoperative cardiac catheterization and selective biplane angiocardiology have been performed in 15 unselected patients from 5 months to 6 years and 3 months

(average 2 years and 3 months) after surgery to obtain an objective analysis of the early operation.

The systolic pressure of the right ventricle in 15 patients in the late postoperative period ranged from 24 to 71 mmHg (average 45 mm Hg). The systolic pressure of the main pulmonary artery was in the range of 20 to 31 mm Hg (average 25 mm Hg). The systolic pressure gradient between the right ventricular inflow and the pulmonary artery ranged from 0 to 55 mm Hg (average 21 mm Hg).

Five of 15 cases had mild pulmonary regurgitation, which was detected by angiocardiography and or auscultation and phonocardiography. All 5 cases, had a diastolic pressure gradient across the pulmonary valve from 5 to 13 mm Hg (average 9.6 mm Hg). In these cases the right ventricular end-diastolic pressure was less than 8 mm Hg, ranging from 0 to 8 mm Hg (average 5.2 mm Hg).

No residual left to right or right to left shunt was found by cardiac catheterization and/or radiocardiography in any of these cases. Neither complete heart block nor pulmonary hypertension was noted. Arterial oxygen saturation was normal in all but 2 patients who were crying during the study (Table 1).

Table 1. Results of postoperative catheterization in 15 patients.

Patient	Age at operation (yr.)	Time to postop. study (yr.)	Pressures (mmHg)					Max. dp/dt of RV (mmHg/sec.)	Pulm. insuff.	Resid. shunt	Art. O ₂ saturation (%)
			RA mean	RV syst./diast.	end-diast.	PA syst./diast.	Systemic artery				
S.N.	6/12	6 3/12	3	30/-10	0	30/ 2	98/60	300	Mild	None	95.8
H.S.	1 3/12	1 6/12	4	63/-1	6	27/10	125/72	—	None	None	96.2
T.O.	10/12	5 0/12	2	36/ 0	5	21/ 5	122/51	310	Mild	None	95.1
H.Y.	1 7/12	5/12	2	38/ 1	3	—	105/68	300	None	None	93.4
T.I.	1 9/12	3 2/12	5	45/ 1	7	31/11	100/76	570	None	None	95.2
Y.I.	1 10/12	3 9/12	4	33/-7	4	25/ 4	110/72	380	Mild	None	97.7
M.K.	1 6/12	3 4/12	6	64/ 0	6	28/ ?	93/53	380	None	None	97.3
K.S.	1 11/12	3 1/12	5	24/ 0	3	21/ 8	112/62	310	None	None	95.7
N.S.	1 9/12	3 3/12	3	33/-9	4	22/ 4	113/70	400	Mild	None	94.9
K.O.	1 7/12	7/12	3	68/-2	7	26/ 7	115/70	610	None	None	95.5
T.K.	1 10/12	11/12	4	56/-8	8	—	112/60	400	None	None	92.9*
S.N.	1 6/12	6/12	3	51/ 0	4	20/ 7	115/67	410	Mild	None	89.8*
H.S.	9/12	9/12	6	43/ 0	7	22/ 5	100/47	430	None	None	96.2
M.O.	1 11/12	1 0/12	8	71/-3	8	25/11	126/80	570	None	None	93.9
S.T.	1 3/12	8/12	4	29/-3	7	29/ 7	81/42	270	None	None	94.5

* At crying

The right ventricular systolic pressure and the systolic pressure gradient between the right ventricular inflow and the pulmonary artery immediately after the repair were remarkably high in some cases, probably related to our technique of relieving the obstruction yet not inserting the outflow patch to relieve the annular stenosis of the pulmonary valve as described above. As the postoperative period lengthened, the

right ventricular systolic pressure and the systolic pressure gradient between the right ventricular inflow and the pulmonary artery lessened (Fig. 1, 2). This may be due to the natural corrective growth of the right ventricular outflow tract and the annulus

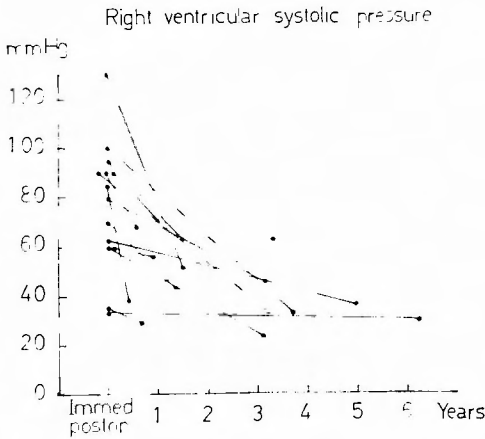


Fig. 1. Comparison of right ventricular systolic pressures obtained in the operating room after correction with those found later at catheterization.

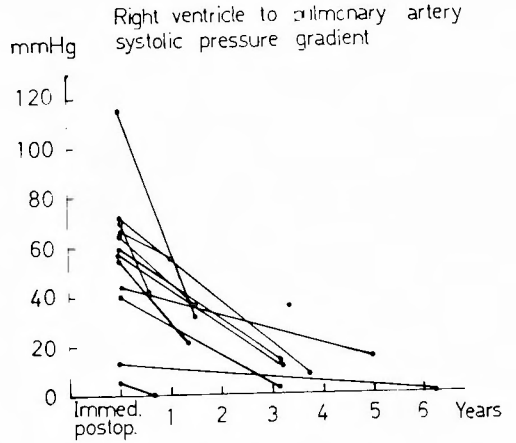


Fig. 2. Comparison of systolic pressure gradients between the right ventricular inflow and the pulmonary artery obtained in the operating room after correction with those found later at catheterization.

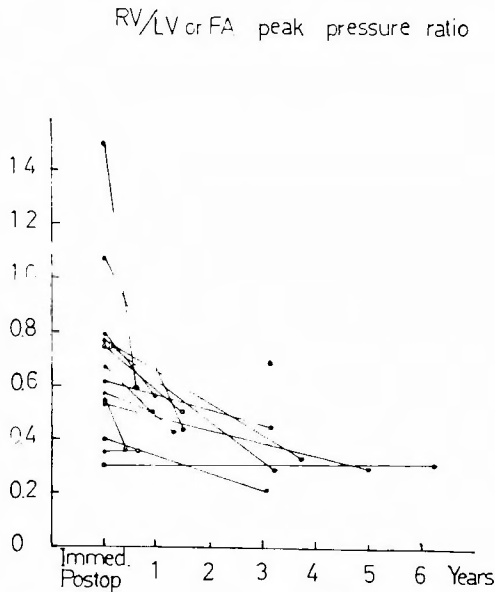


Fig. 3. Comparison of right ventricular/left ventricular or femoral arterial peak pressure ratios obtained in the operating room after correction with those found later at catheterization.

of the pulmonary valve which would occur postoperatively in such young children.

The right ventricular/left ventricular or systemic arterial peak pressure ratio seemed to be one of the most convenient indicators to evaluate the success of the relief of the pulmonary stenosis. In the late postoperative period it was lower than 0.7 in all patients, ranging from 0.21 to 0.69 (average 0.42), whereas the values immediately after the repair in these cases were from 0.31 to 1.50 (average 0.69) (Fig. 3).

The measurement of the diameters of the pulmonary valve annulus and the aortic root was made from the angiocardiology in 14 out of 15 patients in whom the angiocardiology was performed preoperatively and/or postoperatively. Twelve of 14 patients were studied both pre- and postoperatively, 2 only postoperatively. The diameters of the pulmonary valve annulus at the base of the cusps and the ascending aorta at the midpoint between the aortic arch and the aortic valve were measured on the lateral view of serial angiocardigrams in their late systolic periods.

Comparison of the diameters of the pulmonary valve annulus pre- and postoperatively revealed that the preoperative small annulus (range: 9-18 mm, average 11.8 mm on angiocardigrams) had become larger in the course of the postoperative period (range: 15-23 mm, average 18.7 mm). On the other hand, the diameter of the ascending aorta,

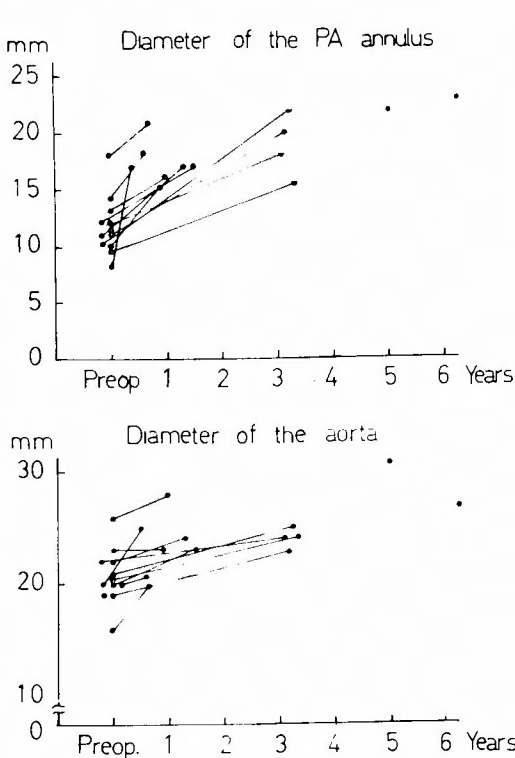


Fig. 4. Comparison of diameters of the pulmonary valve annulus and the ascending aorta measured on pre- and postoperative angiocardigrams.

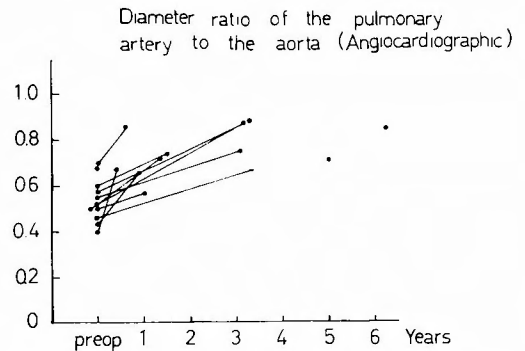


Fig. 5. Diameter ratio of the pulmonary valve annulus to the ascending aorta by pre- and postoperative angiocardiology.

which was uniformly large preoperatively (range: 16-26 mm, average 20.6 mm), had not enlarged as significantly as that of the pulmonary valve annulus in the late postoperative period (range: 21-31 mm, average 24.8 mm) (Fig. 4). The ratio of the diameters of the pulmonary valve annulus to the ascending aorta increased from the preoperative average value of 0.58 to a postoperative one of 0.75 (Fig. 5). This suggests that the abnormally small pulmonary valve annulus and abnormally large aorta

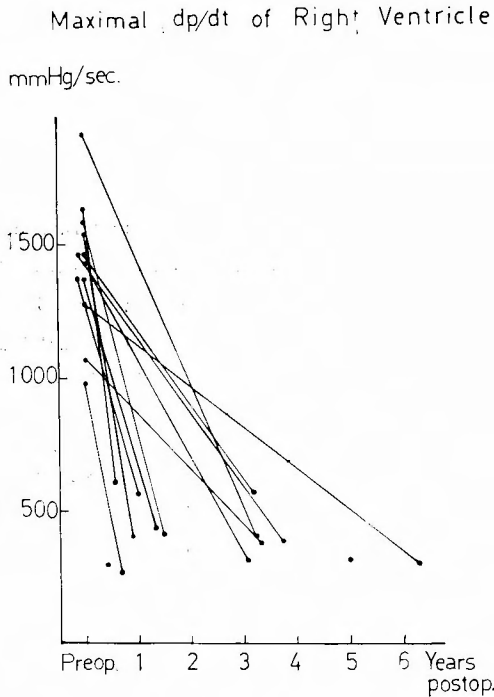


Fig. 6. Comparison of pre-and postoperative maximal dp/dt of the right ventricle.

of the tetralogy may be corrected in the course of the postoperative period in this group.

Maximal dp/dt of the right ventricle markedly decreased postoperatively, concomitant with the decrease of the right ventricular systolic pressure (average preoperative value of 1425 mm Hg to average postoperative 403 mmHg) (Fig. 6).

Clinically, all surviving patients are now asymptomatic and their growth patterns are normal.

Discussion

The management of severe, symptomatic infants and children with tetralogy of Fallot remains a matter of debate. A palliative procedure followed later by a secondary correction has been recommended in most centers except for a few where a primary total correction is performed in selected cases⁹⁾¹⁰⁾¹¹⁾.

We have performed primary total correction of symptomatic tetralogy of Fallot even in patients younger than two years of age, using the deep hypothermia technique described previously¹⁾⁻⁶⁾.

In our experience, the total correction can be well tolerated even at this early age, and the postoperative clinical improvement is dramatic. However, the long-term results of this procedure must await the test of time. Taussig¹²⁾ stated at the 8th International Congress of Cardiology that "the great unanswered question is the long-term results, especially when the operation is performed on a child". Malm also stated "it is important that not only the mortality rate influence our decision, but the late hemodynamic results as well" in the discussion following Dobell's presentation¹⁰⁾.

The late results described here strongly suggest that early primary correction of the tetralogy of Fallot may be a reasonable procedure and may have some hemodynamic and anatomic advantages over total correction on older children. The right ventricular systolic pressure, the systolic pressure gradient between the right ventricular inflow and the pulmonary artery, and the right ventricular/left ventricular or systemic arterial peak pressure ratio fell markedly during the postoperative period, and there was a tendency for further decrease as the postoperative period lengthened.

Anatomically, in accordance with these pressure measurements, it was demonstrated that the pulmonary valve annuli and the pulmonary arteries increased in size but the aorta did not show a comparative increase in the late postoperative period. This may be explained by the postoperative development of the right ventricular outflow tract and the pulmonary arteries owing to the increased blood flow in the pulmonary circulation.

Even if the right ventricular pressure, the pressure gradient between the right ventricle and the pulmonary artery, and the right ventricular/left ventricular or systemic arterial peak pressure ratio remain high immediately after primary total correction in young children, they will improve markedly in the late postoperative period. This was true even when the immediate postoperative pressure in the right ventricle exceeded left ventricular pressure. These results differ from other reports¹³⁾¹⁴⁾¹⁵⁾¹⁶⁾¹⁷⁾ on older children, in which the late postoperative pressures and the pressure ratio were similar or slightly less than immediate postoperative period.

Considering the natural course of tetralogy of Fallot without surgery that the severity of obstruction of the right ventricular outflow tract, the pulmonary valve annulus, and the pulmonary artery progress, it would seem that the patient who can not tolerate correction without outflow tract patch in older age may tolerate in early age without a patch.

Incidence and severity of postoperative pulmonary regurgitation were less than that reported by others⁷⁾¹³⁾¹⁵⁾¹⁶⁾¹⁷⁾¹⁸⁾, perhaps because we do not insert the outflow tract patch across the pulmonary valve annulus even in cases of severe stenosis, if the inner diameter of the pulmonary valve annulus is more than 5 mm or one-third of the diameter of the aortic root. The long-term results of the postoperative pulmo-

nary regurgitation and also those of the outflow patch including that of an aortic or pulmonary homograft prostheses have been under debate. It is apparent, however, that the insertion of the outflow patch must be avoided if possible.

Residual shunt is one of the most important factors that influence the long-term results of the correction of tetralogy of Fallot. No residual shunt was found in this series by cardiac catheterization and/or radiocardiography. Even a ventricular septal defect in the small heart of an infant is easily closed using our deep hypothermia technique which offers a completely bloodless and motionless operative field with a flaccid myocardium.

Summary

Twenty-six patients younger than 2 years of age underwent corrective surgery for tetralogy of Fallot as the primary procedure from December 1964 to December 1972 in Kyoto University Hospital. There were one hospital death and one late death.

Fifteen unselected patients out of 24 survivors were evaluated by cardiac catheterization and angiocardiography from 5 months to 6 years and 3 months (average 2 years and 3 months) after surgery.

Total correction was performed using our unique technique of deep hypothermia with surface cooling and combined use of the partial cardiopulmonary bypass in all except for the first case.

Relief of the outflow tract obstruction was obtained mainly by wedge resection of the hypertrophied medial muscle and incision and mechanical dilatation of the pulmonary valve and the pulmonary valve annulus as necessary without insertion of an outflow patch.

Remarkably high right ventricular systolic pressure (average 78 mmHg) and the pressure gradient between the right ventricular inflow and the pulmonary artery (average 55 mmHg) immediately after the repair became lower in the late postoperative period (average 45 mmHg and 21 mmHg respectively). Average right ventricular/left ventricular or systemic arterial peak pressure ratio also decreased from 0.69 to 0.42. Pulmonary regurgitation was observed in 5 out of 15 patients but, when present, it was mild.

Preoperative small annuli of the pulmonary valve (average 11.8 mm on angiocardiograms) became relatively larger (average 18.7 mm) and preoperative large aorta (average 20.6 mm) became relatively smaller (average 24.8 mm) in the late postoperative period when evaluated by angiocardiography. The diameter ratio of the pulmonary valve annulus to the ascending aorta increased from 0.58 to 0.75.

These results indicate that natural corrective growth of the heart may occur postoperatively in the young children, and excellent late hemodynamic and anatomic results can be expected by the early primary total correction of tetralogy of Fallot without excess outflow tract reconstruction.

References

1. Hikasa, Y., Shirotani, H., Satomura, K., Muraoka, R., Abe, K., Tsushimi K., Yokota, Y., Miki, S., Kawai, J., Mori, A., Okamoto, Y., Koie, H., Ban, T., Kanzaki, Y., Yokota, M., Mori, C., Kamiya, T., Tamura, T., Nishii, A., and Asawa, Y.: Open Heart Surgery in Infants with an Aid of Hypothermic Anesthesia, *Arch. Jap. Chir.*, **36**: 495, 1967.
2. Hikasa, Y., Shirotani, H., Satomura, K., Koie, H., Tsushimi, K., Muraoka, R., Okamoto, Y., Kawai, J., Yokota, M., Yokota, Y., Mori, C., Kamiya, T., Tamura, T., Nishii, A., Muneyuki, M., Asawa, Y.: Open Heart Surgery in Infants with an Aid of Hypothermic Anesthesia (II), *Arch. Jap. Chir.*, **37** : 399, 1968.
3. Hikasa, Y., Shirotani, H., Muraoka, R., Sugitani, A., Okamoto, Y., Miki, S., Tsushimi, K., Abe, K., Koie, H., Yokota, M., Mori, C., Kamiya, T., Nishii, A., Tamura, T., Tatsuta, N., Ban, T., Muneyuki, M., Hazama, H., and Asawa, Y.: Total Correction of Tetralogy of Fallot (III) Experience with Nine Infants under the Age of Two Years, *Arch. Jap. Chir.*, **37**: 867, 1968.
4. Muraoka, R., Yokota, M., Shirotani, H., and Hikasa, Y.: Open Heart Surgery under the Age of Two Years, 10th International Congress of the International Cardiovascular Society, August, 26-28, 1971, Moscow, U.S.S.R.
5. Mori, A., Muraoka, R., Yokota, Y., Okamoto, Y., Ando, F., Fukumasu, H., Oku, H., Ikeda, M., Shirotani, H., Hikasa, Y.: Deep Hypothermia Combined with Cardiopulmonary Bypass for Cardiac Surgery in Neonates and Infants, *J. Thorac. Cardiovasc. Surg.*, **64**: 422, 1972.
6. Muraoka, R., Hikasa, Y., Shirotani, H., Mori, A., Okamoto, Y., Koie, H., Abe, K., Yokota, M., Shirai, O., and Konishi, Y.: Open-heart Surgery under Two Years of Age Using Deep Hypothermia with Surface Cooling and Partial Cardiopulmonary Bypass, *J. Cardiovasc. Surg.*, In Press.
7. Burnell, R.H., Woodson, R.D., Lees, M.H., Bristow, J.D., and Starr, A.: Results of Correction of Tetralogy of Fallot in Children under Four Years of Age, *J. Thorac. Cardiovasc. Surg.*, **57**: 153, 1969.
8. Starr, A., Bonchek, L. I., Sunderland, C.O.: Total Correction of Tetralogy of Fallot in Infancy, *J. Thorac. Cardiovasc. Surg.*, **65**: 45, 1973.
9. McMillan, I. K. R., Johnson, A. M., and Machell, E. S.: Total Correction of Tetralogy of Fallot in Young Children, *Brit. Med. J.*, **1**: 348, 1965.
10. Dobell, A.R.C., Charrette, E. P., and Chughtai, M. S.: Correction of Tetralogy of Fallot in the Young Child, *J. Thorac. Cardiovasc. Surg.*, **55** : 70, 1968.
11. Barratt-Boyes, B. G., Simpson, M., and Neutze, J.M.: Intracardiac Surgery in Neonates and Infants Using Deep Hypothermia with Surface Cooling and Limited Cardiopulmonary Bypass, *Suppl. I to Circulation* **43** and **44**, 1-25, 1971.
12. Taussig, H. B.: Indications and Contraindications for Surgery in the Common Malformations of the Heart, 8th Interamerican Congress of Cardiology, April, 21-27, 1968, Lima, Peru.
13. Bristow, J.D., Adrouny Z.A., Pooter, G.A., Menashe, V.D., Starr, A., and Griswold, H. E. : Hemodynamic Studies After Total Correction of Tetralogy of Fallot, *Amer. J. Cardiol.*, **9**: 924, 1962.
14. Bircks, W., Loogen, F., Wilke, K. H.: Hemodynamic Findings After Surgical Correction of Tetralogy of Fallot, *J. Cardiovasc. Surg.*, **11** : 15, 1970.
15. Hawe, A., Rastelli, G. C., Ritter, D. G., DuShane, J. W., and McGoon, D.C. : Management of Right Ventricular Outflow Tract in Severe Tetralogy of Fallot, *J. Thorac. Cardiovasc. Surg.*, **60** : 131, 1970.
16. Hawe, A., McGoon, D. C., Kincaid, O. W., and Ritter, D. G. : Fate of Outflow Tract in Tetralogy of Fallot, *Ann. Thorac. Surg.*, **13**: 137, 1972.
17. Kirklin, J. W., and Karp, R.B.: The Tetralogy of Fallot from a Surgical Viewpoint, W. B. Saunders Co., 1970.
18. Hallidie-Smith, K.A., Dulake, M., Wong, M., Oakley, C. M., and Goodwin, J.F.: Ventricular Structure and Function After Radical Correction of the Tetralogy of Fallot, *Brit. Heart J.*, **29**: 533, 1967.

和文抄録

2才未満で1期的根治手術を行なったフアロー
四徴症の血行動態的遠隔成績

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昭和39年12月から47年12月までの約8年間に京都大学第2外科に於て2才未満のフアロー四徴症26例に対して1期的根治手術を行ない、手術死1例、手術と関係のない遠隔死1例の成績を得た。

24例の生存例は全て健康な日常生活を営んでいるが、そのうちの15例に対して、術後5カ月から6年3カ月 (平均術後2年3カ月) に亘って心臓カテーテル検査および心血管造影を行なって血行動態的遠隔成績を検討した。

尚、開心術補助手段としては第1例を除いては全例教室で開発発展せしめた表面冷却による超低体温法に部分体外循環を併用する方法を用い、右室流出路狭窄の解除には主として Parietal band およびこれと肺動脈弁輪までの間の右室流出路右側壁筋肉の楔状切除により対処し、肺動脈弁狭窄および弁輪狭窄には弁および弁輪部の切開と拡張器による可及的拡大にとどめ、たとえ重症例といえども流出路パッチは使用しない方針をとった。

その結果は、術直後の比較的高い右室収縮期圧 (平

均 78mmHg)、右室・肺動脈収縮期圧較差 (平均 55 mmHg) および右室/左室・収縮期圧比 (平均 0.69) は遠隔時には著明に低下し、夫々平均 45mmHg, 21 mmHg, および 0.42 の値を示した。特に興味深いのは、術後の経過年月の長い症例程これらの諸値が正常化する傾向を示したことである。

また心血管造影像から測定した肺動脈弁輪部と上行大動脈との直径比は術後遠隔時には術前の平均 0.58 から 0.75 へと増大し、これは肺動脈が術後大動脈に比して著るしく発育が促進され正常化する傾向を有することを示すものである。尚、術後遺残短絡例はなく、肺動脈弁閉鎖不全は軽度のものが5例にみられたに過ぎない。

以上の成績は、本症に対する早期根治手術では血行動態的にも解剖学的にも術後年月の経過と共に自然に矯正されて正常化する傾向が強く、乳児期ないしは2才未満に於ける根治手術の妥当性を示すものと考えらる。