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Total Correction of Tetralogy of Fallot (III) Experience with Nine Infants under the Age of Two Years

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INTRODUCTION

Since the first successful intracardiac correction of the tetralogy of Fallot by Lillehei and his associates in 1954, total correction has become the method of choice for treatment of the tetralogy of Fallot.

Neverthless, despite great advance in surgical technique and in knowledge of the pathophysiology of the tetralogy, most centers have avoided performing total correction in the group under 4～5 years of age owing to the high operative mortality.

The maximum mortality in tetralogy of Fallot occurs during the first or the second year of life. The appearance of life-endangering anoxemic spells usually dictates surgical intervention. For these patients, a systemic-pulmonary shunt has generally been performed for the reason of the poor tolerance of small, hypoxic infants to open heart surgery.

With the combined use of deep hypothermia by surface cooling and partial extracorporeal perfusion for resuscitation and rewarming, we have performed total correction on 9 cases of tetralogy of Fallot weighing between 4. 9 and 9. 8 kg. and aging 3 to 21 months.

MATERIAL AND METHODS

Nine cyanotic patients underwent total correction of tetralogy of Fallot between December, 1964, and May, 1968. All of them were under the age of 2 years and weighed less than 10 kg.. The youngest infant was 3 months old weighed 4. 9 kg..

All patients were investigated in the Department of Pediatrics of Kyoto University

Hospital with cardiac catheterization which documented the presence of arterial desaturation ranging from 60.4 to 88.0 per cent and right ventricular hypertension at systemic levels. Angiocardiogram demonstrated the site and severity of the outflow tract obstruction in each case.

Most of these infants had frequent cyanotic spells, and necessitated surgical intervention as a life-saving procedure (Table 1).

Table 1 Preoprative findings in 9 infants undergoing total correction of tetralogy of Fallot

Patient No.	Age (mo.)	Weighlt (kg)	Indications for Surgery	Red Blood Cells ($\times 10^4$)	Hemoglobin* (Gm.)	Hematocrit (%)	O_2-saturation (%)
1	6	6.0	Dyspnea, Incapacity	574	13.5	49	85
2	15	8.8	Dyspnea, Incapacity	538	14.3	48	87
3	10	7.0	Dyspnea, Incapacity	564	15.2	52	79
4	12	9.0	Syncope, Dyspnea	543	14.4	54	88
5	20	9.5	Syncope, Convulsion, Dyspnea	558	15.1	50	69
6	19	9.0	Syncope, Convulsion, Dyspnea	727	20.5	58	66
7	14	7.9	Dyspnea, Incapacity	629	19.8	58	62
8	21	9.8	Syncope, Dyspnea	552	13.8	48	60
9	3	4.9	Syncope, Dyspnea	418	13.2	38	67

* Normal value at 1 year is 11 Gm./100ml.

The patients with cyanotic spells have a size of pulmonary arterial trunk and its branches large enough to endure the total correction, and the cause of cyanotic spell is due to the spasm of hypertrophied infundibular muscle as will be described later. Accordingly, the infants with frequent anoxic attacks can undergo total correction instead of palliative shunt operation which is now widely accepted as a surgical intervention for tetralogy of Fallot in infancy.

We have applied a deep hypothermia using surface cooling combined with partial extracorporeal perfusion (Hikasa-Shirotani method), the latter was used for resuscitation and rewarming, to total correction of tetralogy of Fallot as well as open heart surgery of ventricular septal defect in infancy as described previously. With this method, one can obtain flaccid, motionless and dry operative field to facilitate the intracardiac manipulation of the small heart of infants, and settle the problems concerning heart massage such as retarded resuscitation, myocardial damage and breakage of repaired defect. Moreover, it is also

useful as assist-perfusion in cases of a surgical block, a temporary disturbance of conducting system and after the total correction of tetralogy of Fallot if necessary.

Our operative procedure for total correction of tetralogy of Fallot was the same as previously described. The outline are as follows. 1) Transverse right ventriculotomy was adopted to avoid the restenosis of the right ventricular outflow tract and postoperative hypofunction of the right ventricle. 2) Incision and resection of the right wing (parietal band) connected with the anterior wall of the right ventricle, which was a extension of the hypertrophic supraventricular ridge were performed. Thus the reconstruction of the right ventricular outflow tract was attempted at the site of right wing. 3) In the case of definite obstructing element to the blood flow noticed at the site of left wing (septal band) by preoperative angiocardiogram, muscles of this part were exised as minimally as possible. Thus the myocardium of the left wing was preserved as much as possible avoiding the injury. 4) At the anterior and septal wall of the right ventricle, no surgical intervention upon the myocardium was intended, but the myocardium of the upper and lower margin of the transverse right ventriculotomy wound was sliced off, so that the closure of this wound may not cause the restenosis of the outflow tract. 5) Then, the fibrously thickened subvalvular endocardium was excised in wedge-shape usually only at the right side. 6) If the valvular pulmonic stenosis was present, using the nerve hook, pulmonar valve was turned upside down and was pulled down to the operative field, then the each commissure was divided with knife, and pulmonary valve dilator which was a miniature form of Gerbode's transventricular mitral valve dilator, was inserted to the pulmonary valve orifice, and the satisfactory widening of the valvular annulus as well as the correction of the valvular stenosis was attempted. 7) Ventricular septal defect was completely closed with the doubled autogenous pericardial patch. 8) Lastly, without using patchgraft for the widening of the outflow tract, transverse right ventriculotomy was sutured and closed.

RESULTS

The operative findings and procedures were listed in Table 2. Average minimum rectal temperature was 20°C (17.2-22.1°C), and average duration of circulatory arrest

Table 2 Operative findings, proceduresand results

Patieat No.	Minimum Rectal Temp. (°C)	Duration of Circulatory Arrest (min.)	PA/Ao Diametric Ratio	Type of Pulmonic Stenosis	VSD Patch	Outflow Patch	Results
1	20.2	54	0.50	Valvular	+	−	Excellent
2	19.4	53	0.33	Infundibular	+	−	Excellent
3	20.0	51	0.47	Infundibular	+	−	Excellent
4	20.0	57	0.44	Infundibular	+	−	Excellent
5	20.5	57	0.34	Infundivulovalvular	+	−	Excellent
6	20.2	59	0.31	Infundibular	+	−	Excellent
7	20.9	58	0.25	Infundibulovalvular	+	−	Died, 20 hours
8	17.2	65	0.38	Infundibulovalvular	+	−	Excellent
9	22.1	55	0.46	Infundibular	+	−	Excellent

was 57 minutes (51–65° minutes). Ventricular septal defect was closed with doubled pericardial patch in all cases, but the outflow patch was never used.

Eight of 9 patients totally corrected survived and their convalescence was uncomplicated, and their improvement was dramatic, immediate and has persisted. All 8 infants showed no cyanosis and their physical and mental development became normal.

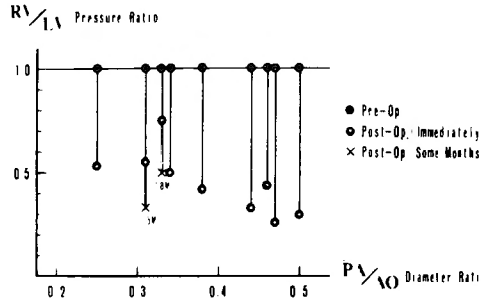

Fig. 1 Relation between pressure ratio of right ventricle to left ventricle and diameter ratio of pulmonary artery to aorta after total correction of tetralogy of Fallot in infants.

Right ventricular pressure in small children seemed to fall more rapidly and readily than in the adults even in the cases which have the pulmonary arterial to aortic diametric ratio less than 50 per cent (Fig. 1).

There was one operative death caused by acute heart failure.

Case 9 might be the youngest not only in our series but also in the world who survived the total correction of tetralogy of Fallot. The case history follows.

Case 9 H. A., a male infant, born by Caesarian operation in full-time weighed 2900 g.. He had been cyanotic from birth, with progressively severe manifestations of dyspnea, cyanosis, and failure to develope, and experienced episodes of anoxic spells associated with extreme cyanosis and dyspnea occuring several times each day since 1.5 months of age.

He was admitted to the Pediatric Department of Kyoto University Hospital and examined by pediatric cardiologists with cardiac catheterization and angiocardiography, and was diagnosed as having tetralogy of Fallot with severe infundibular obstruction. He was transferred to our Surgical Department for emergent surgical intervention at the end of 3 months of age.

Corrective surgery was undertaken on May 23, 1968, using the method above described. Upon entering the chest the diameter of aorta was 13 mm. and of the pulmonary artery 6 mm.; the tension of the latter was low. The right ventricular cardiotomy was extended through a very tight infundibular stenosis of 2 mm. in diameter. The aorta dextoposed extremely and seemed as if the double-outlet right ventricle. The ventricular septal defect was 18 mm. in diameter. One bite of infundibular muscle of the right wing was resected and appeared to relieve the infundibular stenosis quite adequately. The pulmonary valve inverted with a small retractor was normal as well as pulmonary valve annulus. The ventricular septal defect was closed with a double layer of pericardium. In this case, the minimum rectal temperature was 22.1°C, duration of circulatory arrest was 55 minutes, and partial extracorporeal circulation with heart-lung machine in rewarming stage was 10 minutes which raised the rectal temperature from 22.4 to 31.4°C. Immediately after the surgery, the pressure of right ventricle and pulmonary artery revealed 30/0, and 20/7 mmHg. respectively, while left ventricular pressure was 70/0 mmHg..

Postoperatively, recovery was uneventful and symptoms disappeared immediately. He was discharged 1 month after the operation.

DISCUSSION

The significant advances made in the treatment of congenital heart diseases in adults and children have served to focus attention upon the many infants now succumbing to these same lesions during their first two years of life. However, as to the surgical treatment of teralogy of Fallot, patients less than 4-5 years of age with severe hypoxemia due to tetralogy of Fallot are subjected to a shunt procedure because of inordinately high mortality of open heart surgery in small infants.

We have made efforts to perform open operation of infants with congenital heart disease incompatible with life, using deep hypothermia by surface cooling combined with partial extracorporeal perfusion originated by our department. As previously reported, 62 cases of severely ill infants with ventricular septal defect under 2 years of age were treated by open heart surgery with the mortality of 8 per cent. In the same way, 9 infants with tetralogy of Fallot were totally corrected, and 8 of 9 survived.

The cyanotic attacks which endangered life in tetralogy of Fallot were apparently caused by spasm of infundibular muscle in our experience, and in all of these cases the size of the pulmonary arteries was sufficient to tolerate to radical repair of the defects (Fig. 2, 3). Contrally, the infants who have extremely hypoplastic pulmonary arteries

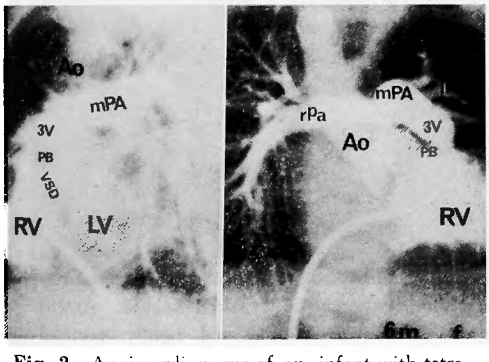

Fig. 2 Angiocardiograms of an infant with tetralogy of Fallot with frequent daily cyanotic spells.

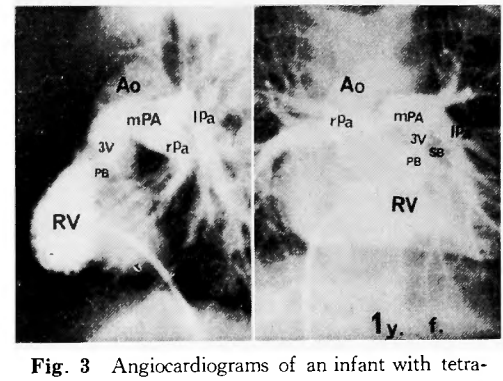

Fig. 3 Angiocardiograms of an infant with tetralogy of Fallot with cyanotic spells which occured every two or three days.

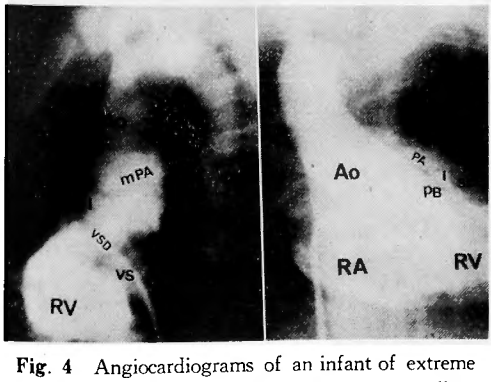

Fig. 4 Angiocardiograms of an infant of extreme teralogy of Fallot who had no cyanotic spell.

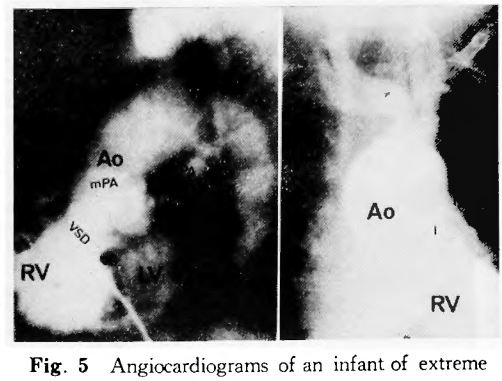

Fig. 5 Angiocardiograms of an infant of extreme tetralogy of Fallot who had no cyanotic spell.

have never showed cyanotic spells despite their anatomical features (Fig. 4, 5). These are regarded functionally as a truncus arteriosus, and extensive collateralization develops. This type of tetralogy of Fallot—extreme tetralogy of Fallot—must be treated by heart transplantation if possible.

As reported previously, we have indicated total correction of tetralogy of Fallot to those patients whose diameter of pulmonary artery is more than 30 per cent of that of the aorta so that we think that patients with tetralogy rarely require palliative shunt operation even in infancy or early childhood. If life-endangering symptoms appear, we perform total correction to the defects irrespective of age.

One of the adventages of early operation exist in the fact that postoperative right ventricular pressure in infants seemed to fall more rapidly and more readily than in the adults and older children.

In addition, tetralogy of Fallot is the disease which is progressive in its natural course, and an appropriate example is presented in Fig. 6 and 7. Angiocardiograms of a patient

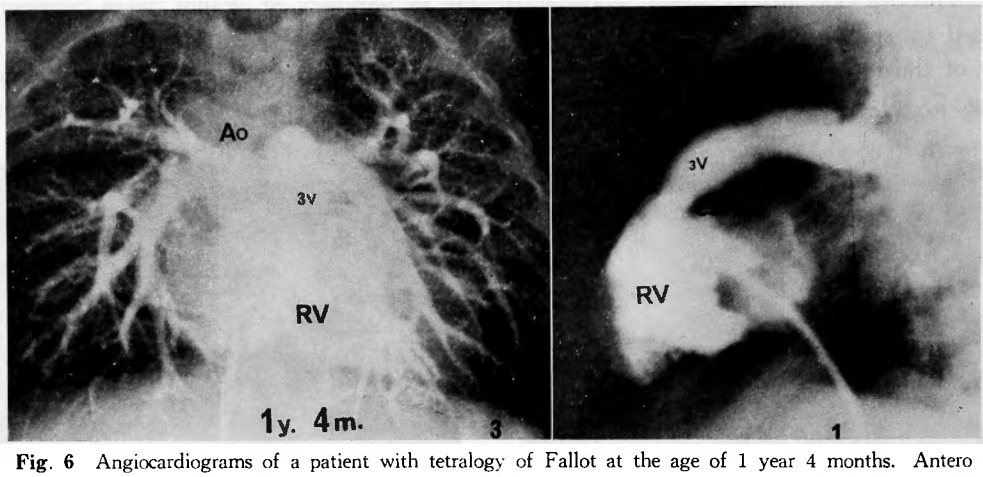

Fig. 6 Angiocardiograms of a patient with tetralogy of Fallot at the age of 1 year 4 months. Antero posterior and lateral views demonstrating moderate degree of infundibular stenosis and slightly opacified aorta.

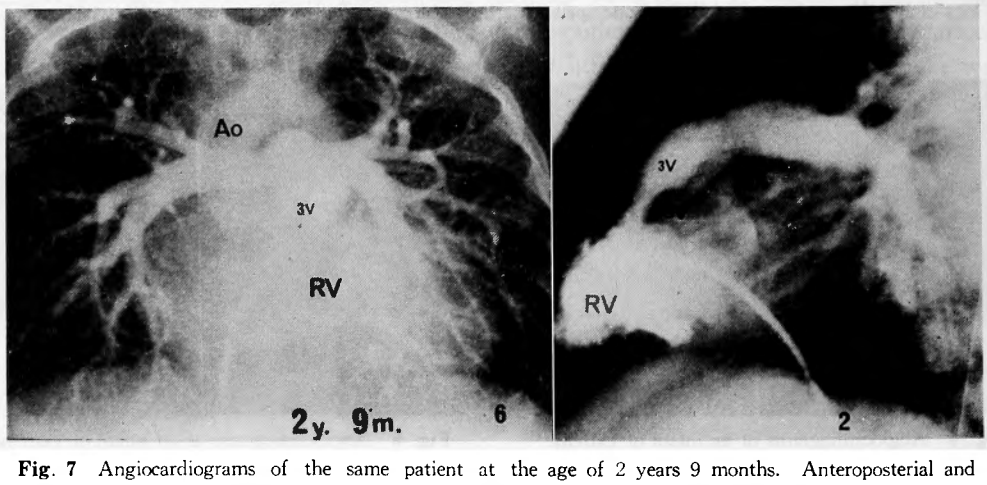

Fig. 7 Angiocardiograms of the same patient at the age of 2 years 9 months. Anteroposterial and lateral views demonstrating increased infundibular stenosis and opacification of the aorta.

with tetralogy of Fallot at the age of 2 years 9 months show more intensely stenotic outflow tract and more distinctly opacified aorta as compared with the angiocardiograms at 1 year 4 months of the same patient. It is apparent that in tetralogy of Fallot, obstruction of the right ventricular outflow tract, hypoplasia of the pulmonary valve annulus and pulmonary artery, increase in right to left shunt become progressively more severe with age.

Accordingly, it is desirable to perform toral correction early in infancy when a surgical intervention at this time is necessary to maintain life if operative mortality is acceptably low.

SUMMARY

Nine infants weighing between 4.9 and 9.8 kg. and aging 3 months to 1 year 9 months with tetralogy of Fallot underwent total correction, using the deep hypothermia combined with partial extracorporeal perfusion which was used for resuscitation and rewarming.

Most of these infants had fequent cyanotic spells and necessitated surgical intervention as a life-saving procedure. Eight of 9 patients totally corrected survived and their improvement was dramatic, immediate and persisted. The youngest infant who survived the operation was 3 months old weighing 4.9 kg., and he might be the youngest in the world who survived the total correction of tetralogy of Fallot.

The postoperative right ventricular pressure in infants seemed to fall more rapidly and readily than in adults and older children.

ADDENDUM

Since this manuscript was submitted, 4 additional infants, 5 months to 22 months of age, weighing from 5.2 to 8.7 kg., have successfully undergone total correction of tetralogy of Fallot.

All four patients had cyanosis at rest and three of them had frequent cyanotic attacks. Their improvement after surgery was dramatic and satisfactory.

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和文抄録

ファロー氏四徴症根治術（第3報）

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4～5才以下のファロー氏四徴症で，それまでに何らかの外科的処置を必要とするものに対しては，従来その根治手術の危険率が甚だ高いために主として姑息的な短絡手術が行なわれて来た．最近になつて，ようやく根治手術の年令を下げる努力がなされるようになつて来たが，未だに2～3才以上とか，15kg以上とかに限られているようである．10kg以下なししは2才未満のファロー氏四徴症に関しては世界的にみてもごく少数の施設に於て，ごく少数の症例に根治手術が行なわれ極めて高い死亡率をみている現状である．しかるに我々は，表面冷却による低余温法と復温時部分体外循環を併用する独自の開心補助手段と，従来より我々が主張して来た右室機能を温存する手術々式により，現在までに9例の生徒3ヵ月から1才9ヵ月，体重4.9kgから9.8kgの患児に根治手術を施行し，その8例に成功を納めた．これらの症例の大部分が頻回の無酸素発作のため乳児期手術を余儀なくされたものである．我々の根治術成功最年少例は生後3ヵ月，4.9kgの男児で，本例はおそらく世界的にみても根治手術に成功した最年少例であろうと思われる．

ファロー氏四徴症自体が年令と共に漏斗部狭窄が増強し，右→左短絡が助長され，益々肺動脈の発育が障害されるという進行的病像を呈する疾患である事実と，乳児期に頻回の無酸素症発作を起す例では漏斗部狭窄が主体で肺動脈自体は根治手術に耐え得るだけの大きさを有していること及び術後の右室圧の正常化が成人や年長小児に比して迅速且確実で，しかも臨床症状の改善がまさに劇的で術後めざましい発育を示すことから，根治手術の成績さえ良好ならば乳児期に於ても短絡手術でなく根治手術を遂行すべきであると考える．