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Surgical Repair of Tetralogy of Fallot associated with Unilateral Anomaly of the Pulmonary Artery

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Introduction

In a few cases of tetralogy of Fallot associated with unilateral dysplasia of the pulmonary artery, problems in surgical management arise. Unilateral dysplasia of the pulmonary artery occurs more frequently in the left pulmonary artery than in the right pulmonary artery. In these cases, operative mortality is high and there is a possibility of postoperative hypertension, severe right heart failure and pulmonary regurgitation. POOL³⁾ and KIRKLIN⁴⁾ reported poor surgical repair and etiology of tetralogy of Fallot with unilateral dysplasia of the pulmonary artery. Furthermore, in cases in which one side of the pulmonary artery is obstructed following Blalock-Taussig's operation, it is advisable to reconstruct the obstructed pulmonary artery.

In this paper, two cases of tetralogy of Fallot associated with unilateral dysplasia of the left pulmonary artery will be described.

Case reports

Case 1. Y.M., a 3-year-old boy, was born after normal full-term pregnancy and normal delivery; his weight was 3000 gm at birth. A heart murmur was noted at birth, and when crying, cyanosis first became evident at 2 months of age. Over the next few months intermittent cyanosis when crying was noted. At 8 months of age, cardiac catheterization (Table 1) revealed tetralogy of Fallot associated with atresia of the left pulmonary artery which was connected to a small patent ductus arteriosus (PDA). After the cardiac catheterization, Inderal was administered in order to increase blood flow into the right pulmonary artery through a narrow outflow tract of the right ventricle. A second cardiac catheterization was performed at the age of 2 yr 8 mo and the

Key words: Unilateral anomaly of the pulmonary artery, Tetralogy of Fallot, Postoperative pulmonary hypertension, Postoperative pulmonary regurgitation, Hancock's conduit.

索引語: 一側肺動脈異常, フェロー四徴症, 術后肺高血圧症, 術后肺動脈閉鎖不全症. Hancock's conduit
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Table 1. Preoperative cardiac catheterization data (Case 1)

	8 mo (6-III-'75)		2 yr 10 mo(6-V-'77)	
	Pressure (mmHg)	Oxygen (Vol%)	Pressure (mmHg)	Oxygen (Vol%)
SVC	(6)	10.21	(4)	7.24
RAmiddle	(6)	9.81	(3)	7.39
IVC	(7)	8.82	(3)	8.03
(SIVC)		9.38		7.71
RV inflow	86/6, 8(46)		82/2, 6(36)	
RV outflow	26~10/4, 6(11)	10.61	16/6, 8(9)	7.27
m-PA	17/10 (12)		13/7 (9)	7.17
r-PA	16/10 (13)	10.76	12/8 (9)	
Ao.	74/48 (62)	13.11 (65.8%)	78/50 (60)	9.35 (54.9%)

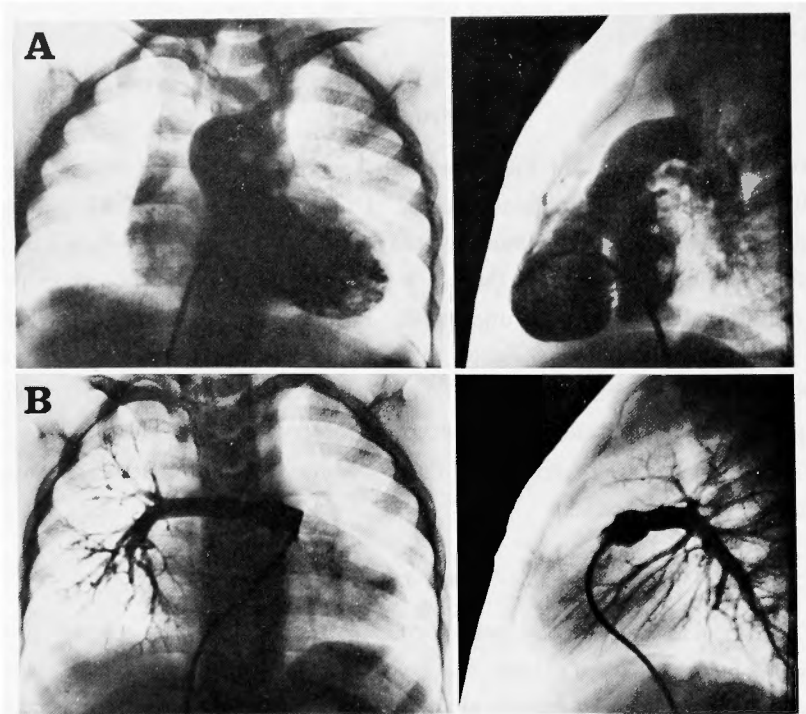


Fig. 1. Preoperative right ventriculography (A) and Selective pulmonary arteriography (B) (Case 1) Right ventriculography reveals the absence of the left pulmonary artery. Selective angiography injected into the main pulmonary artery reveals the presence of only a right pulmonary artery.

previous diagnosis was reconfirmed. Right ventriculography revealed the absence of a left pulmonary artery (Fig. 1-A) but aortography revealed a small PDA, which was connected to the left pulmonary artery (Fig. 2).

Selective angiography injected into the main pulmonary artery revealed the presence of only a right pulmonary artery (Fig. 1-B).

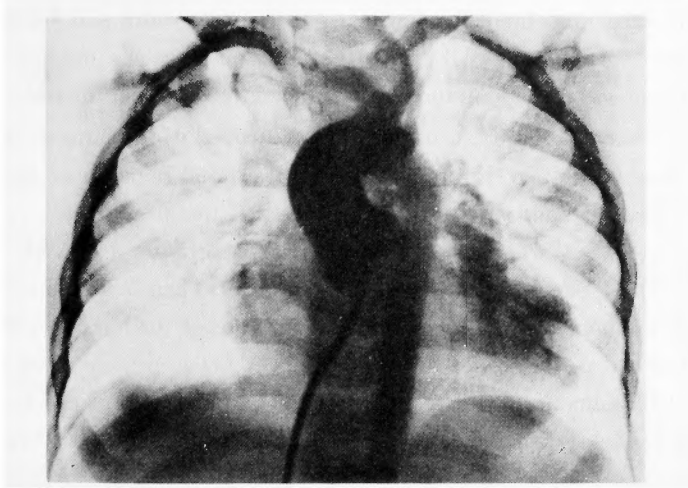


Fig. 2. Preoperative aortography (Case 1)
Aortography reveals a small PDA, which is connected to the left pulmonary artery.

Upon admission, the patient's height and body weight were 92.5 cm and 14.0 kg, respectively. Physical examination revealed severe cyanosis and clubbing. There was a Grade 2/6 continuous murmur in the pulmonary area. Thrill was not palpable. Electrocardiogram revealed marked right ventricular hypertrophy. Chest roentgenogram demonstrated a concave pulmonary artery segment, indicating a decrease in pulmonary vasculatures in the lungs.

On June 28, 1977, total correction was performed in Tenri Hospital (Fig. 3). The diameters of the aorta and main pulmonary artery were 18 and 6 mm, respectively. In the portion of the left pulmonary artery, a ligamentum, such as Lig. ductus arteriosus, was found. The left lung

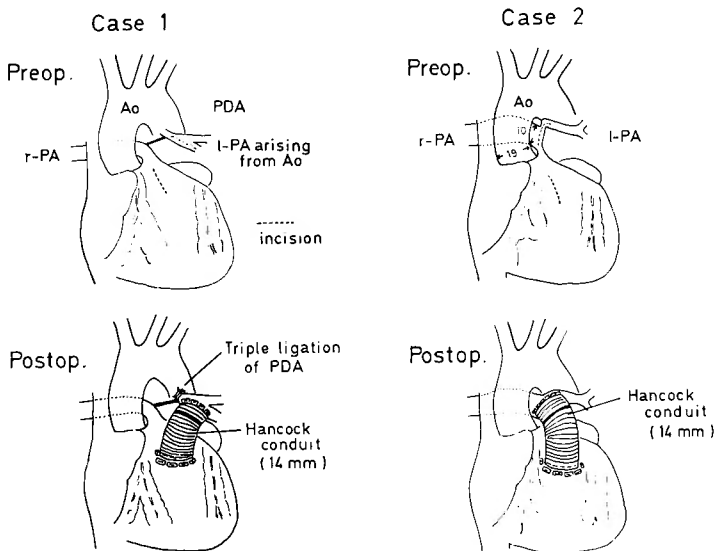


Fig. 3. Surgical procedures performed in Cases 1 and 2.

received blood flow through a small PDA which was 3 mm in width and 5 mm in length. The left pulmonary artery was almost equal to the right pulmonary artery in diameter at the hilus of the lung. An incision was made longitudinally on the outflow tract of the right ventricle.

The pulmonary valve had two cusps. As slight fusions were found at both commissures, commissurotomy was performed. Infundibular stenosis was remarkable due to hypertrophy of the septal and parietal bands. Infundibular resection was performed. A 14×14 mm ventricular septal defect (VSD) was found in the subcrystal portion. The VSD was closed using a Dacron patch. The PDA was closed by triple ligation. Then an incision was made longitudinally on the left pulmonary artery, extending to the hilus of the lung. A Hancock's conduit (14 mm in diameter) was used in order to make a bypass between the outflow tract of the right ventricle and the left pulmonary artery. The Hancock's valve was placed as near the left pulmonary artery as possible, in order to avoid pressure by the sternum. Weaning from H-L bypass was easy, except for slight intraalveolar bleeding. Intracardiac pressures during the operation were measured immediately after H-L bypass. The pressures of the right and left ventricles were 70 and 80 mmHg, respectively.

The intraalveolar bleeding continued for 7 days postoperatively, and was controlled by IPPB and frequent intratracheal suction. Morphine hydrochloride was effective in decreasing the intraalveolar bleeding. Systemic blood pressure was stable and the central venous pressure was less than 18 cm of water.

After the 10th postoperative day, the patient's recovery was relatively uneventful. Postoperative cardiac catheterization was performed on August 31, 1977 (Table 2 and Fig. 4). The pressures (mmHg) of the right ventricle and the femoral artery were 54/0,6 and 108/60, respectively. As the catheter could not be passed through the Hancock's valve, the pressure of the left pulmonary artery was not measured.

Case 2. T. I., a 4-yr and 1 mo-old boy, was born after full-term pregnancy and normal delivery. Body weight at birth was 3200 gm. Cyanosis was noted at birth. A heart murmur was first detected at the age of 1 month. The first cardiac catheterization was performed in a hospital in Hyogo Prefecture at the age of 10 months, on October 7, 1973, and the patient was diagnosed as tetralogy of Fallot. After the first cardiac catheterization, Inderal was administered.

On June 21, 1977, the second cardiac catheterization was performed in Tenri Hospital, reconfirming the previous diagnosis (Table 3). An angiography injected in the outflow tract of the right ventricle demonstrated a stenosis of the left pulmonary artery and a hypoplastic main pulmonary artery (Fig. 5). The size of the right pulmonary artery was normal. He underwent total repair on November 29, 1977 (Fig. 3). The diameters of the aorta and the main pulmonary artery were 19 and 5 mm, respectively. The pulmonary valve was bicuspid and hypoplastic. Furthermore, the left pulmonary artery was small in size and its origin was very stenotic. Therefore, reconstruction of the main and left pulmonary artery was necessary. A Hancock's conduit (14 mm in diameter) was used in order to make a bypass between the pulmonary artery and the right ventricle. An incision was made longitudinally on the main pulmonary artery extending

Table 2. Postoperative cardiac catheterization data (Case 1)
—64 days postoperatively—

	Pressure (mmHg)	Oxygen (Vol%)
SVC	(4)	10.07
RA middle	(4)	9.47
IVC	(4)	9.83
(SIVC)		9.93
RV inflow	54/0, 6 (19)	
RV outflow	50/-2, 5(18)	9.23
m-PA		
FA	108/60 (77)	13.01 (90.8%)

Table 3. Preoperative cardiac catheterization data (Case 2)

	Pressure (mmHg)	Oxygen (Vol%)
SVC	(2)	8.34
RA middle	(2)	7.48
IVC	(2)	8.05
RV inflow	80/0.4 (36)	
RV outflow	70/0.2 (32)	7.43
m-PA	8/4 (4)	7.07
γ -PA	10/6	7.05
Ao.	80/50	10.30 (65.7%)



Fig. 4. Postoperative right ventriculography (Case 1)

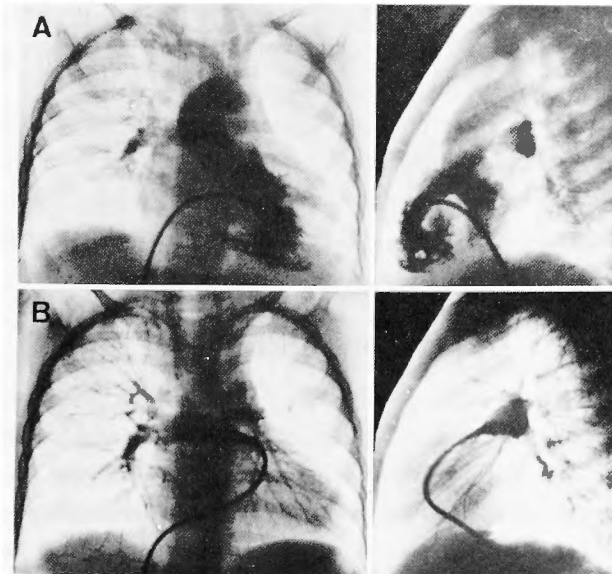


Fig. 5. An angiography injected in the outflow tract of the right ventricle (B) demonstrates stenosis of the left pulmonary artery and a hypoplastic main pulmonary artery (Case 2).

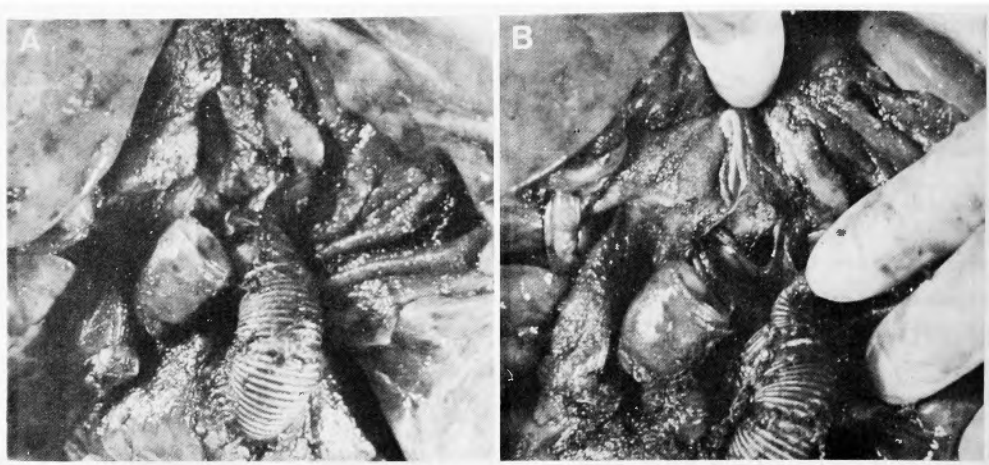


Fig. 6. Autopsy revealed that the main pulmonary artery and the stenotic portion of the left pulmonary artery had been pulled anteriorly by the Hancock's conduit, and thus the left pulmonary artery was kinked and obstructed at the bifurcation (Case 2).

to the stenotic portion of the left pulmonary artery. An another incision was made longitudinally on the outflow tract of the right ventricle. The intracardiac pressures of the right ventricle and the left ventricle were the same. Intraalveolar bleeding occurred on the 2nd postoperative day and continued to the 15th postoperative day.

On the 6th postoperative day, massive bleeding occurred from the operative skin incision. Immediately, resternotomy was performed and the bleeding point on the right ventricular wall was found. This was a 21G needle hole from which the intracardiac pressures were measured during the previous operation. The hole may have reopened due to increasing pressure of the right ventricle following greater pulmonary arterial resistance. After the resternotomy, right heart failure increased and intraalveolar bleeding became severe. The patient's condition was complicated by pneumonia and he died on the 15th postoperative day.

Autopsy revealed that the main pulmonary artery and the stenotic portion of the left pulmonary artery had been pulled anteriorly by the Hancock's conduit, and thus the left pulmonary artery was kinked and obstructed at the bifurcation (Fig. 6). Therefore the right lung received all the blood that returned to the right side of the heart and subsequently, intraalveolar bleeding occurred. No residual VSD was found. Bronchopneumonia was found in both lungs.

Discussion

Some problems still remain in the use of Hancock's conduit for infants and children, namely, the development of thrombosis, the degeneration of porcine valve and lack of space in placing a suitable size Hancock's conduit. However, there are some advantages, i.e. Hancock's conduit with various size can be easily obtained and they have a lower incidence of thrombosis than mechanical valves, and thus do not need anticoagulant therapy. Thus in tetralogy of Fallot with hypoplasia of the pulmonary artery or an abnormal origin of unilateral pulmonary artery, it is advisable to use a Hancock's conduit in order to reconstruct the outflow tract of the right

ventricle and the abnormal pulmonary artery. Surgical repair of tetralogy of Fallot with unilateral absence of pulmonary artery has a high operative mortality rate because of postoperative pulmonary hypertension, intractable heart failure and severe pulmonary regurgitation. In tetralogy of Fallot in which right and left pulmonary arteries are of different diameters (Case 2)⁵⁾, the results of surgical repair may be poor, as in the case with unilateral absence of pulmonary artery, unless the blood flow of right and left pulmonary artery is suitably separated. When the pulmonary artery is considerably small in size, Blalock-Taussig's operation may be advisable in order to develop a peripheral pulmonary capillary bed.

In fact, as described by GOLDSMITH et al.²⁾, palliative operations for these cases also have a high mortality rate.

KITO et al.⁵⁾ reported a successful surgical correction of tetralogy of Fallot with congenital absence of the left pulmonary artery without postoperative pulmonary hypertension. In this case, a lung scintigram revealed that left pulmonary flow was very small through a PDA. However surgical repair of the tetralogy of Fallot with unilateral absence of pulmonary artery usually results in pulmonary hypertension. Reports of follow-up results in patients with pulmonary hypertension after repair of tetralogy of Fallot are not in agreement.

MALM et al.⁶⁾ described 2 patients who had a single functioning major peripheral pulmonary artery after repair.

BIRCKS et al.¹⁾ noted that pulmonary arterial pressure frequently remained unchanged or actually increased after repair of tetralogy of Fallot, especially when associated with pulmonary regurgitation. On the other hand, KINSLEY et al.³⁾ noted that pulmonary regurgitation was of little prognostic significance despite pulmonary hypertension after repair of tetralogy of Fallot. The presence of pulmonary regurgitation after the repair of tetralogy of Fallot with unilateral absence of one pulmonary artery may be primary or secondary. In fact we have usually found that severe pulmonary regurgitation shortly after operation results in a low cardiac output and an increased mortality rate. Therefore, we introduced a bypass method using a Hancock's conduit in order to completely prevent subsequent pulmonary regurgitation. In Case 2, subsequent pulmonary hypertension and severe heart failure should have been managed by severe medication until pulmonary capillary beds could develop. A Hancock's conduit is too difficult to suture to the pulmonary artery without bleeding; occasionally uncontrollable bleeding continues at the site of the anastomosis of the conduit in the left pulmonary artery.

In addition, as in Case 2, the pulmonary artery sutured to a Hancock's conduit had been pulled anteriorly and kinked, interrupting the blood flow. Retrospectively, a suitable pericardium should have been placed between the pulmonary artery and conduit. When a bypass (Hancock's conduit) between the right ventricle and the pulmonary artery is established parallel to the main pulmonary artery, pulmonary regurgitation should be minimized to decrease the overload on the right ventricle. Otherwise, fatal right heart failure may occur postoperatively due to pulmonary regurgitation and pulmonary hypertension.

Preoperative selective angiography of the main pulmonary artery is essential for estimating the prognosis and selecting the surgical procedures.

Summary

Tetralogy of Fallot associated with unilateral dysplasia of the pulmonary artery is a rare form of congenital heart disease and its surgical management is often difficult. There are considerable variations in approach and prognosis.

The two cases in this report were managed by using a valve-retaining xenograft (Hancock's conduit) to completely prevent subsequent pulmonary regurgitation. In the first case, the left pulmonary artery arose from the aorta through a PDA. The PDA was ligated and the Hancock's conduit was placed between the left pulmonary artery and the outflow tract of the right ventricle. Postoperative intraalveolar bleeding was under control after 7 days. He is well-doing now.

In the second case, the left pulmonary artery was hypoplastic and smaller than the right pulmonary artery. A Hancock's conduit was sutured to the site between the left and the main pulmonary arteries. However, the left pulmonary artery was kinked interrupting the blood flow. The intraalveolar bleeding, possibly from the right lung, was uncontrolled.

In tetralogy of Fallot associated with unilateral dysplasia, pulmonary hypertension and pulmonary regurgitation are anticipated. Thus, it is advisable that a Hancock's conduit is used to completely prevent subsequent pulmonary regurgitation and to overcome right heart failure.

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和文抄録

一側肺動脈異常を伴うファロー四徴症の外科治療

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左肺動脈異常を伴う T/F 2 例を経験した。症例 1: 左肺動脈は大動脈より分枝, 本来の左肺動脈は索状物と化し完全に閉塞していた。右室流出路と左肺動脈の間を Hancock conduit で bypass を作製した。術後 7 日間肺出血を来した。症例 2: 左肺動脈は発育不良で, 太さは右側の 1/4 程度であった。右室流出路と肺動脈分枝部の間に Hancock conduit で bypass を作製した。しかし, 術後左肺動脈は屈曲し, 右肺出血を来

した。術後 6 日目右室壁より出血あり再開胸止血, しかし術後 15 日目, 左心不全で死亡した。

一側肺動脈異常のある T/F では, 術後, 高度肺高血圧症を残し, 肺動脈弁逆流の増強又は続発を来し, 致命的右室不全に到ることが予測される。従って, こうした症例では, 肺動脈弁逆流を防止し, 右室不全軽減の為, Hancock conduit を使用することが望ましい。