Tetralogy of Fallot with Anomalous Origin of Left Pulmonary Artery

MASATO ENDO, KIYOSHI HANEDA, HITOSHI MOHRI and SHIGEO YAMAKI*

Department of Thoracic and Cardiovascular Surgery, Tohoku University School of Medicine, Sendai 980 and *Katta Hospital, Shiroishi 989-02

Between 1971 and 1990, 7 patients of tetralogy of Fallot with anomalous origin of left pulmonary artery underwent intracardiac repairs at Tohoku University Hospital. They were 2 males and 5 females with ages ranging from 4 to 26 years old. The right pulmonary artery connected to right ventricle in all cases, whereas no communications between right ventricle and the left pulmonary artery were found. The left pulmonary artery directly originated from the ascending aorta in 2 patients (group I) and connected to the ductus arteriosus in 5 patients (groups II and III). In 2 patients (group II), the left pulmonary artery was separated from the pulmonary arterial trunk by the intraluminal membrane, receiving blood supply through the ductus. In the remaining 3 patients (group III), there were no continuations between both pulmonary arteries. At the correction, communication between the left pulmonary artery and the pulmonary arterial trunk could be reconstructed in groups I and II. However, it was not possible in group III, because the ductus arteriosus and the left pulmonary artery had already been occluded before the intracardiac repair. In group III, ventricular septal defect was closed using a one-way valved patch or a perforated patch to decrease supersystemic right ventricular pressure. Postoperative right ventricular aortic pressure ratio was between 0.5 and 0.8 in groups I and II, and between 0.8 and 1.0 in group III. Three patients (one in each group) died after the operation. Severe pulmonary vascular obstructive disease was found in the left lung of group I. Tetralogy of Fallot with anomalous origin of unilateral pulmonary artery should be corrected before the advance of pulmonary vascular obstructive disease in case pulmonary artery originates from the ascending aorta, and before the occlusion of the ductus arteriosus in case pulmonary artery originates from it. ——— Tetralogy of Fallot; anomalous origin of pulmonary artery; correction

The results of the correction of Tetralogy of Fallot have been improved. However, Tetralogy of Fallot complicated with other anomalies still has some difficulties for total correction. Among various anomalies, the anomalous origin of left pulmonary artery was encountered 7 (3%) out of 222 cases of Tetralogy of Fallot during past 20 years of our experience. These cases were classified into 3
groups by the morphology of the affected pulmonary artery. Each group demonstrated a characteristic clinical course and different surgical problems. In this article, our classification of the anomaly and the surgical problems were described.

**Materials**

From 1971 to 1990, 222 cases of Tetralogy of Fallot underwent the intracardiac repair at Tohoku University Hospital. Among them the anomalous origin of unilateral pulmonary artery was found in 7 cases. Although the right pulmonary artery connected to the pulmonary arterial trunk originated from right ventricle in all cases, the left pulmonary artery either originated from the ascending aorta or connected to the ductus arteriosus. These 7 cases were classified into 3 groups (Fig. 1, Table 1).

In group I (2 cases), the left pulmonary artery originated directly from the ascending aorta. In group II and III, the left pulmonary artery connected to the ductus arteriosus. In group II (2 cases), there was morphological continuation between the pulmonary arterial trunk and the left pulmonary artery, although they were separated by the intraluminal membrane. Blood supply to the left pulmonary artery was through the ductus. In group

**Fig. 1.** Tetralogy of Fallot with anomalous origin of left pulmonary artery. 7 cases of this anomaly were classified into 3 groups. Ao, ascending aorta; RPA, right pulmonary artery; LPA, left pulmonary artery; PDA, patent ductus arteriosus; RV, right ventricle; LV, left ventricle.

**Table 1. Patients' data and associated cardiac anomalies**

<table>
<thead>
<tr>
<th>Case</th>
<th>Age, (years)</th>
<th>Sex</th>
<th>Group</th>
<th>Other cardiovascular anomalies</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>13</td>
<td>F</td>
<td>I</td>
<td>Right aortic arch</td>
</tr>
<tr>
<td>2</td>
<td>26</td>
<td>F</td>
<td>I</td>
<td>Right aortic arch</td>
</tr>
<tr>
<td>3</td>
<td>4</td>
<td>F</td>
<td>II</td>
<td>PDA, PFO</td>
</tr>
<tr>
<td>4</td>
<td>5</td>
<td>M</td>
<td>II</td>
<td>PDA</td>
</tr>
<tr>
<td>5</td>
<td>4</td>
<td>F</td>
<td>III</td>
<td>PDA, PFO</td>
</tr>
<tr>
<td>6</td>
<td>14</td>
<td>M</td>
<td>III</td>
<td>PDA</td>
</tr>
<tr>
<td>7</td>
<td>16</td>
<td>F</td>
<td>III</td>
<td>PDA, Right aortic arch</td>
</tr>
</tbody>
</table>

PDA, patent ductus arteriosus, PFO, patent foramen ovale.
Tetralogy of Fallot with Anomalous Left Pulmonary Artery

III (3 cases), there were no anatomical continuations between both pulmonary arteries.

Surgical procedures

Surgical procedures are summarised in Table 2. In group I, the left pulmonary artery was resected from the ascending aorta and connected to the pulmonary arterial trunk by direct anastomosis (case 1) (Fig. 2) or interposing a prosthetic graft (case 2). Ventricular septal defect was closed by Dacron patch. In group II, the confluence between the left

<table>
<thead>
<tr>
<th>Case</th>
<th>Group</th>
<th>VSD patch</th>
<th>RVOT-PA reconstruction</th>
<th>Post-op RV/Ao</th>
<th>Result</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>I</td>
<td></td>
<td></td>
<td>0.7</td>
<td>Died</td>
</tr>
<tr>
<td>2</td>
<td>I</td>
<td></td>
<td></td>
<td>0.8</td>
<td>Alive</td>
</tr>
<tr>
<td>3</td>
<td>II</td>
<td></td>
<td></td>
<td>0.6</td>
<td>Alive</td>
</tr>
<tr>
<td>4</td>
<td>II</td>
<td></td>
<td></td>
<td>0.5</td>
<td>Died</td>
</tr>
<tr>
<td>5</td>
<td>III</td>
<td></td>
<td></td>
<td>1.0</td>
<td>Alive</td>
</tr>
<tr>
<td>6</td>
<td>III</td>
<td></td>
<td></td>
<td>1.0</td>
<td>Alive</td>
</tr>
<tr>
<td>7</td>
<td>III</td>
<td></td>
<td></td>
<td>0.8</td>
<td>Died</td>
</tr>
</tbody>
</table>

VSD, ventricular septal defect; RVOT, right ventricular outflow tract.

Fig. 2. Surgical findings of case 1.
pulmonary artery and the pulmonary arterial trunk was created by resection of the membranous tissue and patch angioplasty. In group III, complete occlusion of the left pulmonary artery from the ductus to the hilar portion was found by preoperative angiography, although the blood flow of the left pulmonary artery from the ductus had been revealed at 2 to 9 years before the operation. Consequently, the reconstruction of the left pulmonary artery could not be achieved. The diameter of the right pulmonary artery was small in these cases. Therefore, a perforated patch or an one-way valved patch with the perforation of 1 cm in diameter, which allowed blood flow from right ventricle to left ventricle, was used for the closure of ventricular septal defect in order to abate postoperative right ventricular pressure overload (Fig. 3).

**RESULTS**

Postoperative right ventricular/aortic pressure ratio (RV/Ao) ranged from 0.5 to 0.8 in all patients of group I and II, whereas it ranged from 0.8 to 1.0 in group III. The right to left interventricular shunt through the perforated or the one-way valved patch was detected by echocardiography in case 5 and case 6 (group III) with RV/Ao of 1.0. Although peak right ventricular pressure was slightly lower than left ventricular pressure in case 7 (RV/Ao = 0.8), a right to left shunt through the one-way valved patch was also revealed. It suggested that right ventricular pressure exceeded left ventricular pressure in certain points of the contracting cycle. In case 5, RV/Ao had decreased from 1.0 to 0.8 in a month.

**Fig. 3. Surgical findings of case 5.**
after the operation. Case 6, in which the perforated patch was used, demonstrated RV/Ao of 1.0 even at one year after the operation.

Case 1 died due to multiple organ failure on the 38th postoperative day. Histological examination of the right lung showed abnormally thin media of pulmonary arterioles suggesting reduced pulmonary blood flow (Fig. 4). On the other hand, pulmonary vascular obstructive disease including medial thickening and intimal proliferation was observed in the left lung (Fig. 5).

Case 4 died of low cardiac output syndrome on the 2nd postoperative day despite successful decrease in RV/Ao (0.5).

In case 7, stable hemodynamics were maintained after the operation. A right to left shunt through the one-way valved patch, which was detected on the first postoperative day, disappeared on the next day. However, grandmal epileptic seizure, which had been caused by previous brain abscess and controlled by anticonvulsants preoperatively, reappeared after extubation of the endotracheal

Fig. 4. Pulmonary arteriole with thin media in the right lung of case 1.
tube. The patient fell into uncontrollable status epilepticus and died on the 8th postoperative day. Moderate medial hypertrophy of pulmonary arterioles were found in the left lung. Remaining 4 patients took uneventful recoveries after the operation. At discharge, their physical conditions were NYHA class 1 for 3 patients and class 2 for 1 patient.

**DISCUSSION**

Anomalous origin of unilateral pulmonary artery associated with Tetralogy of Fallot was found more often at the left pulmonary artery than at the right pulmonary artery. Pool et al. (1962) reported that 16 cases had the anomalous origin of the left pulmonary artery out of 17 cases with this anomaly. Kutsche and Van Mielop (1988) also reported that 12 cases had the anomalous origin of the left pulmonary artery out of 15 cases of Tetralogy of Fallot with the unilateral
Tetralogy of Fallot with Anomalous Left Pulmonary Artery

There have been several reports dealing with the pathogenesis of the anomalous origin of a pulmonary artery. Concerning the anomalous origin of a pulmonary artery from the ascending aorta such as group I, Cucci et al. (1964) explained that the dislocation of the trunco-conal ridge in early embryo resulted in the abnormal aorticopulmonary septation and, eventually, the anomalously originated pulmonary artery from the ascending aorta. However, Kutsche and Van Mielop (1988) mentioned that its pathogenesis involved the failure of normal development of the fifth or sixth aortic arch.

The membranous obstruction of the left pulmonary artery found in group II is likely to be the congenital lesion. According to some papers (Coles et al. 1966; Momma et al. 1986), however, this lesion can be caused by contraction of the ductal smooth muscle which invades into the pulmonary artery, and also by the wall thickening following endothelial damage of jet lesion usually found at the branching sites of vessels.

With regard to the pulmonary artery originated from the aortic arch by way of the ductus, Pool et al. (1962) explained that the aortic side (ventral bud) of the sixth aortic arch failed to develop and the ductal side (dorsal bud), which would form the ductus arteriosus, communicated with the pulmonary post-branchial plexus, resulting in the ductus supplying the affected lung.

Surgical treatment of Tetralogy of Fallot with anomalous origin of unilateral pulmonary artery was first reported by Pool in 1962. Since then, there have been several reports (Kirklin et al. 1965; Kawada et al. 1971; Imai et al. 1974; Calder et al. 1980; Robita 1985) of surgical correction of this anomaly. But, the operative results had not been satisfactory not only because complicated operations were required but also because this anomaly had some inherent problems. Our experiences also demonstrated several problems with this anomaly.

In the case of the unilateral pulmonary artery originated from the ascending aorta, there is strong possibility of pulmonary vascular obstructive disease in the affected lung as seen in case 1, which showed medial thickening, cellular and fibrous intimal proliferation of pulmonary arterioles. Kawada et al. (1971) and Robita (1985) reported the same findings. The difference of the pulmonary vascular disease between both lungs is the problem influencing the pulmonary circulation as well as the respiratory function.

The unilateral pulmonary artery originated by way of the ductus arteriosus has the inborn problem of future occlusion of the ductus and resultant occlusion of the pulmonary artery. In our case 7, the ductus and the pulmonary artery were already closed by the age of 4. Then, this type of anomaly should be corrected as early as possible to avoid the occlusion of the affected pulmonary artery. Direct anastomosis between the pulmonary arterial trunk and the anomalous pulmonary artery were feasible in the cases reported by Kirklin et al. (1965) and Kawada et al. (1971). However, it should be also considered that a prosthetic...
graft may be required to reconstruct the extrapericardial pulmonary artery for the cases with a long distance between the pulmonary arterial trunk and the anomalous pulmonary artery.

For the cases of occluded pulmonary artery, we could not reconstruct the pulmonary artery because it occluded all the way to the pulmonary hilum. But, Presbitero et al. (1984) reported that pulmonary artery was patent at the pulmonary hilum in most cases of absent pulmonary artery, which included the cases of the occlusion of ductus and pulmonary artery. For such cases, the reconstruction of pulmonary artery might be possible by further dissection of pulmonary artery and prosthetic grafting.

It is important to decompress the right ventricular pressure overload for the cases in which the affected pulmonary artery can't be reconstructed and the contralateral pulmonary artery is small. The one-way valved patch and the perforated patch for the closure of ventricular septal defect were effective in our cases to prevent supersystemic right ventricular pressure at acute postoperative period. One-way valved patch is considered to be better, because it allows right to left shunt at the period of right ventricular overload just after the operation and prevents left to right shunt when right ventricular pressure decreases lower than left ventricular pressure. The adequate size of perforation and the configuration of the valve should be studied furthermore, although we have not experienced uncontrollable desaturation caused by right to left shunt through one-way valved patch with perforation of 1 cm in diameter.

In conclusion, 7 cases of Tetralogy of Fallot with anomalous origin of unilateral pulmonary artery were morphologically classified into 3 groups. These anomalies should be corrected before either the advance of pulmonary vascular obstructive disease or the occlusion of the ductus arteriosus, when the unilateral pulmonary artery originates from the ascending aorta or from the ductus arteriosus, respectively.

References


