Diagnosis and Surgical Treatment of “Angiographically Absent Pulmonary Artery Syndrome”

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Treatment of “Angiographically Absent Pulmonary Artery Syndrome”*. Tohoku J.
exp. Med., 1978, 125 (1), 1-9 — Among the heart diseases, there are some
cases in which pulmonary arteries cannot be visualized by standard right
ventriculography and aortography. We have categorized such a case as
“angiographically absent pulmonary artery syndrome”, which may include
congenital or acquired absent pulmonary artery, and anatomically present but
non-visualized pulmonary artery. It has been extremely difficult to achieve an
accurate diagnosis in such cases and there has been no report of successful surgical
treatment for them. Recently, we developed a method of retrograde pulmonary
arteriography, with which the pulmonary artery can be successfully visualized
in cases of angiographically absent pulmonary artery. Subsequently, re-
establishment of the blood flow to the non-visualized pulmonary artery has
been successfully performed in 7 of 8 cases of this syndrome. — angiographi-
cally absent pulmonary artery syndrome; retrograde pulmonary arteriography;
surgical repair

Unilateral absence of the pulmonary artery is a well documented lesion which
has been encountered most frequently in association with tetralogy of Fallot.
Blood flow to the affected lung is possibly established from collateral routes such as
the ascending aorta, patent ductus arteriosus (either through the aortic arch or via
the brachiocephalic artery), and bronchial artery.

On the other hand, there are many cases in which pulmonary arteries cannot
be visualized by standard angiographic procedures despite anatomical presence of
the pulmonary artery. It is thus worthwhile to categorize such cases as “angio-
graphically absent pulmonary artery syndrome”. Recently one of us (M.T.) acci-
dentally discovered a method of retrograde pulmonary arteriography. Using this
new technique the pulmonary artery can be successfully visualized in cases of
angiographically absent pulmonary artery. Consequently, surgical repairs were

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performed and the blood flow to such "non-visualized pulmonary artery" was successfully re-established in 7 of 8 patients.

The aim of the this paper is to present this new technique and the results of surgical repair for the so-called "angiographically absent pulmonary artery."

METHOD OF THE RETROGRADE PULMONARY ARTERIOGRAPHY

The details of the method are as follows: An endhole catheter is introduced from the femoral vein through the atrial septum into the pulmonary vein. This method is technically simple, especially in cases associated with atrial septal defect or patent foramen ovale. The catheter is wedged slightly into the pulmonary vein and contrast medium is injected at a relatively low flow rate to avoid recoiling of the catheter. Manual or automatic injection of 0.3 ml/kg of Angiographin or Angioconray is delivered at a rate of about 10 ml/sec and rapid serial rentgeno-graphy at a rate of 6/sec for 2 sec are made. Immediately after the injection of contrast medium, the catheter is flushed with normal saline and then pulled back into the left atrium. After the procedure, a few patients had bouts of bloody sputum, which disappeared rapidly without any special treatment.

TABLE 1. *Cases of retrograde*

Patient No.	Age (years)	Basic lesion	Findings of rt. ventriculo-graphy or aortography	Etiology	Previous surgery
1	21	Pentalogy of Fallot	No opacification of rt. PA	Acquired	Modified Blalock
2	11	Tetralogy of Fallot	No opacification of lt. PA	Acquired	Exploratory thoracotomy
3	5	Pulmonary atresia, asplenia syndrome	No opacification of both PA	Acquired	Intrapericardial aorto-pulmonary artery anastomosis
4	6	Tetralogy of Fallot	No opacification of lt. PA	Congenital	None
5	1	Complete TGA	No opacification of lt. PA	Congenital	None
6	16	Tetralogy of Fallot	No opacification of rt. PA	Acquired	Rt. Blalock
	38	Tricuspid regurgitation, peripheral pulmonary stenosis	No opacification of lt. PA	Acquired	None
8	4	Pulmonary atresia, asplenia syndrome	No opacification of both PA	Congenital	None

Complete TGA, complete transposition of the great arteries.

CLINICAL MATERIALS

The method was applied to 8 patients from April, 1970 to September, 1974 at the Tohoku University Hospital. The cases included tetralogy of Fallot, pentalogy of Fallot, complete transposition of the great arteries, cor biloculare with asplenic syndrome and tricuspid regurgitation (Table 1). In 7 of these 8 patients, basic lesions were considered as congenital, and as acquired in the remainder. Selective right ventriculography and aortography could not demonstrate the right pulmonary artery in 2 cases (Cases 1 and 6), the left pulmonary artery in 4 (Cases 2, 4, 5 and 7), and both pulmonary arteries in 2 (Cases 3 and 8).

The retrograde pulmonary arteriographies were performed 10 times in these 8 patients and the pulmonary artery was successfully visualized 9 times (all cases except Case 7). The diameter of the affected pulmonary artery could be calculated preoperatively with high accuracy, and thus reconstructive surgery could be performed as planned in the majority of cases.

Case 1. This 21-year-old male had undergone an end-to-end anastomosis of the left subclavian artery to the left pulmonary artery at the age of 11 with some clinical

pulmonary arteriography

Findings of retrograde pulmonary arteriography	Proposed operation	Surgery	Remarks
Opacification of rt. PA (9 mm in diameter)	Intracardiac repair, widening of obstructed rt. PA	Intracardiac repair	Improved, peak RV/AO pressure ratio 0.25, residual L-R shunt of 42%
Opacification of lt. PA (4 mm in diameter)	Intracardiac repair, angioplasty of lt. PA	Intracardiac repair with valved patch, patch angioplasty for lt. PA	Improved, peak RV/AO pressure ratio 0.44
Opacification of both PA (6 & 5 mm in diameter)	Blalock for rt. PA	Rt. Blalock	Improved
Opacification of lt. PA (4 mm in diameter)	Blalock for lt. PA	Lt. Blalock	Improved
Opacification of lt. PA (4 mm in diameter)	Mustard procedure, Blalock for lt. PA	Mustard procedure	Expired 2 hours p.o. (low cardiac output syndrome)
Opacification of rt. PA (10 mm diameter)	Intracardiac repair, angioplasty for rt. PA	Intracardiac repair, angioplasty for rt. PA	Improved, peak RV/AO pressure ratio 0.35
Failed to opacify lt. PA	TVR, angioplasty for both PA	TVR, widening of rt. PA	Expired on 5th day p.o. (pulmonary edema)
Opacification of both PA	Blalock for rt. PA	Lt. Blalock	Improved

TVR, tricuspid valve replacement.

Fig. 1. Retrograde right pulmonary arteriogram in Case 1. The right pulmonary artery of 9 mm in diameter was visualized.

improvements. However, cyanosis had gradually increased and he was referred to our clinic for further evaluation. A diagnosis of pentalogy of Fallot with severe right ventricular outflow obstruction was established by cardiac catheterization. The right pulmonary artery could not be visualized by means of right ventriculography and aortography. The retrograde pulmonary arteriography, however, revealed a well developed right pulmonary artery of 9 mm in diameter (Fig. 1). Intracardiac repair was performed successfully using an autologous pericardial valved patch for widening of the right ventricular outflow tract. The obstructed right main pulmonary artery was widened with a pericardial patch. The left subclavian artery to the left pulmonary artery anastomosis was left intact because of severe adhesion. His postoperative course was uneventful, although about 42% of the left to right shunt remained.

Case 2. This markedly cyanosed 11-year-old female had an exploratory thoracotomy at the age of 3 at another institution to attempt a Blalock-Taussig operation, which was not successful due to a technical difficulty. The patient was later referred to our clinic for possible intracardiac repair of her cardiac lesion. A diagnosis of tetralogy of Fallot with severe hypoplasia of the main pulmonary artery was established, but standard right ventriculography and aortography could not demonstrate the left pulmonary artery (Fig. 2A). Finally, retrograde pulmonary arteriography revealed the presence of hypoplastic left pulmonary artery of 4 mm in diameter (Fig. 2B). At surgery, her left pulmonary artery was completely occluded with an organized thrombus at a point 2 cm from bifurcation. The obstruction was successfully removed and the left pulmonary artery was repaired using saphenous vein patch graft, and the blood flow to the left lung was restored. The intracardiac repair was also carried out using the autologous pericardial patch (Fig. 2C).

Her postoperative course was uneventful, and the postoperative cardiac catheterization disclosed satisfactory hemodynamics with a peak right ventricular-aortic pressure ratio of 0.44 and the absence of any significant residual pulmonary stenosis or intracardiac shunt. Postoperative serial angiography showed patency of the left pulmonary artery (Fig. 2D).

Case 4. This 6-year-old male with pulmonary atresia and ventricular septal defect, who received a Waterston shunt procedure at the age of 4 months with remarkable clinical improvement, was referred to our clinic for the possible intracardiac repair. An aortography revealed only opacification of the right pulmonary artery (Fig. 3A), and possible kinking of the right main pulmonary artery at the point of Waterston anastomosis. Retrograde

Fig. 2. Pre- and postoperative pulmonary arteriograms and diagram of operative procedures in Case 2.

The left pulmonary artery was not visualized by conventional right ventriculography (A). Retrograde left pulmonary arteriography revealed the existence of the left main pulmonary artery (B). The left main pulmonary artery was repaired by a saphenous vein patch angioplasty and intracardiac repair was done with a pericardial valved patch (C). Postoperative pulmonary arteriogram demonstrated patency of both pulmonary arteries (D).

pulmonary arteriography demonstrated the hypoplastic left pulmonary artery of 4 mm in diameter, but failed to ensure the existence of the main pulmonary artery and the junction of the left and right pulmonary arteries (Fig. 3B). From these angiographic findings, definitive surgery for this patient has been suspended for the time being, but a left Blalock-Taussig anastomosis was performed expecting a further development of the left pulmonary artery.

Case 6. This 16-year-old male received the left Blalock-Taussig anastomosis at the age of 4 years with reasonable clinical improvement. He was referred to our clinic for the possible intracardiac repair. Right ventriculography failed to show any portion of the pulmonary artery (Fig. 4A), and the aortography revealed severe obstruction of the left Blalock-Taussig anastomosis (Fig. 4B). Retrograde pulmonary arteriography revealed a well developed right pulmonary artery and the junction of both pulmonary arteries (Fig. 4C). After simple ligation of the left subclavian artery, the intracardiac repair was successfully performed using an autologous pericardial valved patch to widen the right ventricular outflow tract, and also using the distal portion of this valved patch the obstructed place of the left pulmonary artery was widened. His postoperative course was uneventful except

Fig. 3. Aortogram and retrograde left pulmonary arteriogram in Case 4. Aortography revealed only opacification of the right pulmonary artery (A). Retrograde left pulmonary arteriography demonstrated the existence of the left pulmonary artery (B).

moderately severe congestive heart failure, which was easily controlled with an anti-congestive regimen. Postoperative serial angiography disclosed well established blood supply to both lungs (Fig. 4D), and the peak RV-AO pressure ratio remained around 0.35.

DISCUSSION

Congenital unilateral absence of a pulmonary artery was first described by Frentzel in 1868. Pool et al. (1962) reviewed 98 cases including 4 of their own, and Kawada et al. (1972) reported their experience of 8 cases. Their reports showed that the lesion was most frequently associated with tetralogy of Fallot, followed by the patent ductus arteriosus and the atrial septal defect. Of particular interest is a trend that the absence of the left pulmonary artery was associated

Fig. 4. Pre- and postoperative angiograms in Case 6. No opacification was obtained by conventional right ventriculography (A). Selective left subclavian arteriography revealed obstructed Blalock-Taussig anastomosis and the existence of the left pulmonary artery (B). Retrograde right pulmonary arteriography revealed a well developed right pulmonary artery (C). Postoperative right ventriculography showed both pulmonary arteries of adequate size (D).

predominantly with tetralogy of Fallot, while absence of the right pulmonary artery with the patent ductus arteriosus.

Terminology used in previous reports for anomalies similar to our "angiographically absent pulmonary artery" includes agenesis of pulmonary artery, unilateral absence of a pulmonary artery, unilateral single pulmonary artery, unilateral pulmonary atresia, and major peripheral pulmonary artery obstruction. It may be worthwhile to review the reported cases individually to clarify our classification. In the report of Pool et al. (1962), it was shown that corrective surgery for tetralogy of Fallot with absent pulmonary artery was highly hazardous because increased pulmonary vascular resistance with decreased pulmonary vascular beds tended to produce pulmonary hypertension and subsequent congestive heart failure postoperatively.

Bjork et al. (1963) reported a case of tetralogy of Fallot with congenital absence of the pulmonary artery, which could have survived through intracardiac repair, but his postoperative course was troublesome because of sequential right ventricular hypertension and finally the patient succumbed to pulmonary edema.

Kirklin et al. (1965) also described congenital absence of the left pulmonary artery as one of the crucial problems in total correction of tetralogy of Fallot. One of the three operated cases of this lesion was lost shortly after the surgery and the other two survived two to three years with severe congestive heart failure and significantly restricted physical activity due to the residual pulmonary hypertension.

It is noteworthy that a majority of surgically treated cases of absent pulmonary artery in the literature (Malm et al. 1966; Koyamada et al. 1969; Williams et al. 1972; Donahoo et al. 1973; Yamashita et al. 1974), except those recently reported by the Mayo group (Kinsley et al. 1974), had high operative risk and sequential pulmonary hypertension and congestive heart failure as nearly universal findings when the affected pulmonary artery was not repaired.

As far as we know, there are no reports of the successful reconstruction of "absent pulmonary artery", except one equivocal case of absent left pulmonary artery with severe bifurcation stenosis reported by Wallsh et al. (1968). This case was successfully repaired by means of a pericardial patch angioplasty to relieve severe stenosis of the bifurcation without any sequel. We are convinced, through detailed reviews of angiographic findings in previous reports, that in the majority of these cases including the above particular case more accurate diagnoses could have been established preoperatively and reconstructive surgery for the so-called absent pulmonary artery could have been performed successfully if the retrograde pulmonary arteriography were applied.

Fortunately, "angiographically absent pulmonary artery" in three of our four patients with tetralogy of Fallot has been successfully repaired as planned preoperatively according to the findings from retrograde pulmonary arteriography. In the remaining case, however, severe adhesion at the site of previous end-to-end anastomosis of the Blalock shunt prevented reconstruction of the obstructed left pulmonary artery. Fortunately, in this case the right ventricular systolic pressure was normalized postoperatively, but 42% of the left to right diversion through the previous shunt persisted. We suspect that we should have ligated the left subclavian artery at the time of corrective surgery in this case, as Kirklin (1965) had reported. Blood flow to the affected lung through the bronchial artery, which was about 25% of the total cardiac output, is thought to be less than through the shunt, and the remaining shunt is thought not to be of much burden to the left ventricle in this particular case.

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