Bilateral Coronary-Pulmonary Artery Fistulas in Pulmonary Atresia With Ventricular Septal Defect

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SUMMARY

We present a very rare case of bilateral coronary to pulmonary artery fistulas associated with pulmonary atresia with ventricular septal defect. The courses of coronary to pulmonary artery fistulas have to be clearly delineated by detailed angiography prior to corrective surgery. (Int Heart J 2017; 58: 463-465)

Key words: Cyanosis, Rastelli procedure, Coronary angiography, Blalock-Taussig shunt

In patients with pulmonary atresia with ventricular septal defect (PA/VSD), pulmonary blood flow can be usually derived from different segments of the aorta and its branches, such as a patent ductus arteriosus or major aortopulmonary collateral arteries. A coronary-pulmonary artery fistula is also known to be a rare source of pulmonary blood flow. We encountered a notable case of bilateral coronary-pulmonary artery fistulas associated with PA/VSD in whom two fistulous tracts arising from both the left and right coronary arteries to the right pulmonary artery.

CASE REPORT

A male newborn delivered at 37 weeks of gestation with a weight of 2,513 g was admitted to our hospital because of cyanosis and heart murmur. Echocardiography demonstrated a large ventricular septal defect beneath the overriding aorta and an absence of antegrade pulmonary blood flow from the right ventricle, ie, pulmonary atresia with ventricular septal defect (PA/VSD). The left main coronary artery trunk was dilated. Aortography and selective coronary angiography were performed to evaluate the precise anatomy of the coronary arteries, which revealed two fistulous communications arising from coronary arteries to the right pulmonary artery (RPA); one was a communication between the dilated left coronary artery and the RPA (Figure 1A and 1B), and the other was a communication between the right coronary artery and the RPA (Figure 1C and 1D). These findings suggested bilateral coronary-pulmonary artery fistulas. The left pulmonary artery (LPA) was disconnected to the RPA and supplied from a large patent ductus arteriosus (PDA) (Figure 2). Cardiac catheterization demonstrated that the LPA and RPA pressures were 52/29 mmHg (systolic/diastolic pressure) and 35/26 mmHg, respectively. The LPA pressure was increased due to the large PDA. At 3 months of age, we performed a modified Blalock-Taussig shunt using a 4-mm Gore-Tex graft through left thoracotomy. A pericardial roll was made and sutured to the hilar LPA after ligation of the ductus. The roll was fixed to the aortic arch for the later reconstruction of the pulmonary arterial tree. The oxygen saturation was changed from 85% to 80%. Follow-up catheterization was performed at 1 year of age, which demonstrated that the LPA and RPA pressures were 16/10 mmHg and 45/32 mmHg, respectively. The pulmonary artery bed had developed to the pulmonary artery index of 334 cm²/m². Therefore, we decided to proceed with the corrective surgery at 16 months of age. After a median sternotomy using cardiopulmonary bypass, we divided the previous Blalock-Taussig shunt graft and connected the pericardial roll on the LPA to the RPA using a 8-mm Gore-Tex graft. Both coronary-pulmonary artery fistulas from the right coronary artery and left coronary artery were divided at the sides of the RPA. The ventricular septal defect was closed with a patch via a right atriotomy, and a 14-mm Gore-Tex conduit with trileaflet bulging sinuses was interposed between the right ventricle and the main pulmonary artery to obtain an antegrade pulmonary blood flow, ie, Rastelli procedure (Supplemental Figure 1). Subsequent electrocardiograms showed no ischemic changes, and the postoperative course was uneventful. The oxygen saturation level was elevated to 100% with room air. At 19 months of age, cardiac catheterization demonstrated that the right ventricular pressure and RPA pressure were as low as 38/8 mmHg and 26/9 mmHg, respectively. Selective coronary angiography revealed that the left main coronary arterial trunk still remained dilated but the contrast media runoff adequately (Figure 3). The right coronary artery was intact. He is being treated with oral aspirin and has been doing well without any symptoms or myocardial ischemia.

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Figure 1. Selective coronary angiography revealed bilateral coronary-pulmonary artery fistulas. Left coronary angiograms (A and B) showed that the dilated left coronary artery (black arrow) was connected to the right pulmonary artery, whereas right coronary angiograms (C and D) showed that a fistulous tract arising from the right coronary artery (white arrow) was connected to the right pulmonary artery, suggesting bilateral coronary pulmonary arterial fistulas.

Figure 2. Scheme of the angiograms shown in Figure 1: the black arrow indicates a CPAF arising from the left coronary artery (LCA) to the right pulmonary artery (RPA), and the white arrow indicates another CPAF arising from the right coronary artery (RCA) to the RPA. AO indicates aorta; PDA, patent ductus arteriosus; LCx, left circumflex artery; LAD, left anterior descending artery; and LMT, left main coronary trunk.

Figure 3. Selective coronary angiogram after the corrective surgery showing that the proximal left main coronary artery remained dilated.
**DISCUSSION**

To the best of our knowledge, this is the first case of bilateral coronary-pulmonary artery fistulas arising from the left and right coronary arteries as rare sources of pulmonary blood flow in a patient with PA/VSD. The incidence of coronary-pulmonary artery fistulas is 7-10% among patients with PA/VSD, and the incidence of PA/VSD is 4.2 per 100,000 live births. A coronary-pulmonary artery fistula arises more commonly from the left coronary artery, especially the left circumflex artery, than from the right coronary artery, and occasionally arises from a solitary coronary system. Coronary-pulmonary artery fistulas associated with PA/VSD usually coexist with major aortopulmonary collaterals. Two reports have described the coexistence of a coronary-pulmonary artery fistula and a nonconfluent left pulmonary artery connected through a patent ductus arteriosus, and both cases were similar to the present case. This association seems to be characteristic in patients with coronary-pulmonary artery fistula and PA/VSD.

When early maldevelopment of the central pulmonary artery leads to improper continuity with the embryonic lung, the remnants of the intersegmental vascular system, such as aortopulmonary collaterals, consequently become the major source of supply of pulmonary blood flow. During development of the coronary arterial system, protruded proepicardial cells migrate over the surface of the heart and the coronary vascular plexus, and grow inward to penetrate the aorta. As proepicardial cells are closely related to the intersegmental vascular system, it is possible that both the left and right coronary arteries become the fistulous tracts that are the major source of pulmonary blood flow in patients with PA/VSD. Another embryological assessment in the present case was that the coronary-pulmonary artery fistula was connected through Vieussens' collateral pathway coursing from the right coronary artery across the right ventricular outflow to the left coronary artery (Supplemental Figure 2). Since the incidence of Vieussens' collateral is 32%, it is possible that a coronary-pulmonary artery fistula through a Vieussens' collateral can be the major source of pulmonary blood flow in PA/VSD. Although the occurrence of a fistula between a Vieussens' collateral and a pulmonary artery has been previously reported, there is no report of a patient with PA/VSD in whom a Vieussens' collateral mainly supplies pulmonary blood flow.

In all reported cases, the coronary artery proximal to the fistula was enlarged, supplying blood to both pulmonary and coronary arteries. The risk of development of pulmonary arterial hypertension cannot be excluded. A review of the literature suggests a good prognosis after surgical repair before progression of a pulmonary vascular occlusive disease. Contrary to expectations, coronary insufficiency has not been reported either before or after repair surgery. However, the ectasia of LMT that remained after surgery in the present case was resembled a coronary aneurysmal lesion, so we must carefully monitor any myocardial ischemic symptoms.

A coronary-pulmonary artery fistula with PA/VSD should be managed by any technique that allows preservation of the coronary circulation and connection of the right ventricle to the pulmonary artery. Therefore, it is essential to determine the precise anatomy of a coronary-pulmonary artery fistula before corrective surgery. In spite of the efficacy of cardiac computed tomography or magnetic resonance imaging in the current management practices of congenital heart disease, these imaging modalities cannot establish without a doubt a conclusive diagnosis of coronary-pulmonary artery fistula. We believe that selective angiography is warranted among these patients.

In conclusion, a coronary-pulmonary artery fistula should be considered as a possible source of pulmonary supply in patients with PA/VSD. Furthermore, fistulous communications can rarely arise from bilateral coronary arteries. The courses of coronary-pulmonary artery fistulas have to be clearly delineated using detailed angiography prior to surgery.

**DISCLOSURE**

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**REFERENCES**


**SUPPLEMENTAL FILES**

Supplemental Figure 1, 2
Please see supplemental files: https://www.jstage.jst.co.jp/article/ihj/58/3/58_16-324/_article/supplement