Huge right ventricle–right coronary artery fistula compromising right ventricular function in a patient with pulmonary atresia and intact ventricular septum: A case report

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Pulmonary atresia with intact ventricular septum (PA/IVS) is a morphologic heterogeneous congenital cardiac anomaly and shows numerous physiologic conditions. In patients with a fistula between the small hypertensive right ventricle (RV) and the coronary arteries, decompression of the hypertensive RV continues to be controversial,^{1,2} whereas leaving the hypertensive RV may cause progressive coronary stenosis and dilation.³ Recently, we encountered a boy with PA/IVS in whom a huge fistula developed between the RV and right coronary artery (RCA) late after a modified Blalock-Taussig shunt and bidirectional cavopulmonary shunt operation.

Clinical Summary

A 10-year-old boy with PA/IVS associated with a huge RV-RCA fistula was admitted to relieve RV suprasystemic hypertension and

dilatation. He was born at term and noted to be cyanotic a few hours after birth. Cardiac ultrasonography revealed PA/IVS, patent ductus arteriosus, RV-RCA fistula, and severe hypoplasia of the RV. He underwent modified right and left Blalock-Taussig shunts at 3 and 14 months of age, respectively. At 3 years of age, he underwent a bidirectional cavopulmonary shunt during which the Blalock-Taussig shunts were taken down, because preoperative catheterization, showing a hypoplastic left pulmonary artery and an RV-RCA fistula (6 mm in diameter), suggested that the risk of a Fontan operation would be great. He underwent left pulmonary balloon dilatation and stent placement in the left pulmonary artery to increase left pulmonary blood flow at 6 and 8 years of age, respectively. However, severe cyanosis and chest pain developed when he was 9 years of age.

Cardiac catheterization showed a huge RV-RCA fistula, 12 mm in diameter, with stenosis at the middle portion of the RCA (Figure 1). Right ventriculography showed to-and-fro blood flow between the aorta and RV through a huge RV-RCA fistula, which seemed to increase the volume load of the RV and decrease the diastolic blood flow of the left coronary artery (LCA) as in aortic regurgitation. Most of the RV wall was supplied by blood flow from the LCAs (Figure 2). There was no tricuspid regurgitation. RV systolic pressure and end-diastolic volume (162 mm Hg and 42 mL, respectively) were higher than those at 9 years (139 mm Hg and 19 mL, respectively). The electrocardiogram showed apparent LCA lesion ischemia. A balloon occlusion test of the fistula did not cause ischemic changes in the electrocardiogram. These data suggested that ligation of the RV-RCA fistula might reduce the RV volume load and improve coronary perfusion, resulting in an

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Figure 1. Right ventriculogram in the posteroanterior projection showing a diminutive RV ejecting blood to the ascending aorta in systole.

improvement of left ventricular performance. To improve arterial blood oxygenation, reestablishment of the systemic-pulmonary shunt was planned in addition to fistula ligation.

At 10 years of age, he underwent fistula ligation and systemic-pulmonary shunt through a median sternotomy without the use of cardiopulmonary bypass. The distal portion of the fistulous RCA was ligated to save other blood supply to the RV from this artery. Ligation caused no ischemic change in the electrocardiogram, and RV systolic pressure decreased from 160 to 110 mm Hg. Then, a systemic-pulmonary artery shunt was constructed between the brachiocephalic artery and the left pulmonary artery with an 8-mm polytetrafluoroethylene graft. Arterial oxygen saturation increased from 82% to 90%. Cardiac catheterization 2 weeks after the operation showed decreases in RV systolic pressure and end-diastolic volume (110 mm Hg and 32 mL, respectively) (Figure 3). Left ventricular ejection fraction was improved from 41% to 55%. His chest pain disappeared and cyanosis was improved. The patient was discharged uneventfully with an arterial oxygen saturation of 86%.

Comment

Because huge fistulas between the RV and coronary arteries create diverse conditions, a surgical strategy for the fistula should be determined on a case-by-case basis. If a large RV-RCA fistula supplied blood flow to the RV wall, decompression of the RV would be contraindicated and the patient would be considered a candidate for heart transplantation. In the present case, the blood flow to the RV wall was mainly supplied by the LCA, and a huge fistula seemed to be the only

Figure 2. Left coronary angiogram showing collateral arteries to segment 4, the posterior descending branch and atrioventricular node branch at the RCA. There was no communicating artery between the LCA and RV.

Figure 3. Postoperative right ventriculogram in the posteroanterior projection showing an occluded RCA and reduced RV cavity.

cause of increased RV pressure and volume, because there was no tricuspid regurgitation. Therefore, ligation of the fistula was chosen, although aorta-RV shunt,⁴ tricuspid closure,⁵ and thromboexclusion of the RV⁶ have been reported to decompress the RV.

The patient underwent the ligation of the fistula and left aortopulmonary shunt uneventfully with a decrease in RV pressure. Postoperative cardiac catheterization also revealed decreased RV pressure and volume, although RV hypertrophy did not regress, probably because RV systolic pressure was still equal to left ventricular systolic pressure. Clinically, his chest pain decreased. These data suggested that ligation of the fistula decompressed RV cavity and increased LCA blood flow, resulting in improvement of left ventricular performance.

Because the follow-up period was short, the fate of the RV cavity, which has no blood outflow, is uncertain. If the RV cavity should redilate, other surgical options such as tricuspid closure or thromboexclusion of the RV should be considered. Therefore, the case should be closely monitored.

In summary, ligation of the fistula is one surgical option to decompress the RV cavity for PA/IVS associated with a large RV-LCA fistula if blood flow to RV wall is mainly supplied by the LCA.

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