Anomalous origin of the left coronary artery from the main pulmonary artery associated with Berry syndrome

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Berry syndrome, which consists of distal aortopulmonary septal defect (APSD), aortic origin of the right pulmonary artery, interrupted aortic arch, intact ventricular septum, and patent ductus arteriosus, is an extremely uncommon congenital cardiac malformation, with only 25 cases having been reported since the first report in 1982.[1,2] Here we report the first case of Berry syndrome complicated by anomalous origin of the left coronary artery from the main pulmonary artery. The possibility of coronary anomaly should be seriously considered in all cases of Berry syndrome.

Clinical Summary

A 2-day-old male infant weighing 3.2 kg was referred to our hospital because of congestive heart failure. Electrocardiography showed right ventricular hypertrophy, and chest radiography showed cardiomegaly with increased pulmonary vascular markings. Echocardiography revealed a type A interrupted aortic arch, a confluent APSD, aortic origin of the right pulmonary artery, an intact ventricular septum, and a patent ductus arteriosus. Cardiac catheterization and angiography confirmed the above diagnosis (Figure 1). However, at that point, we had not detected anomalous origin of the left coronary artery from the main pulmonary artery. At 6 days of age, the patient underwent surgical intervention for total repair through a median sternotomy. An end-to-side anastomosis of the descending aorta to the undersurface of the aortic arch was performed during circulatory arrest. Low-flow bypass was resumed, and during cardioplegic arrest, inspection of the internal anatomy confirmed the existence of APSD and a right pulmonary artery orifice adjacent to the APSD (Figure 2). We also detected a small hole immediately below the APSD (Figure 2). Because a conductor inserted into this hole appeared to enter the pulmonary artery, we assumed that the hole was part of the APSD. The ascending aorta was transected, leaving its posterior wall to form the posterior wall of the confluence between the right and left pulmonary arteries. The anterior wall of the pulmonary artery was reconstructed by means of anastomosis of an autologous pericardial patch; the small defect was therefore in the pulmonary side. The divided segments of the ascending aorta were directly anastomosed.

Soon after reperfusion, the patient's heartbeat spontaneously reverted to sinus rhythm, and cardiopulmonary bypass was successfully discontinued. After discontinuation of cardiopulmonary

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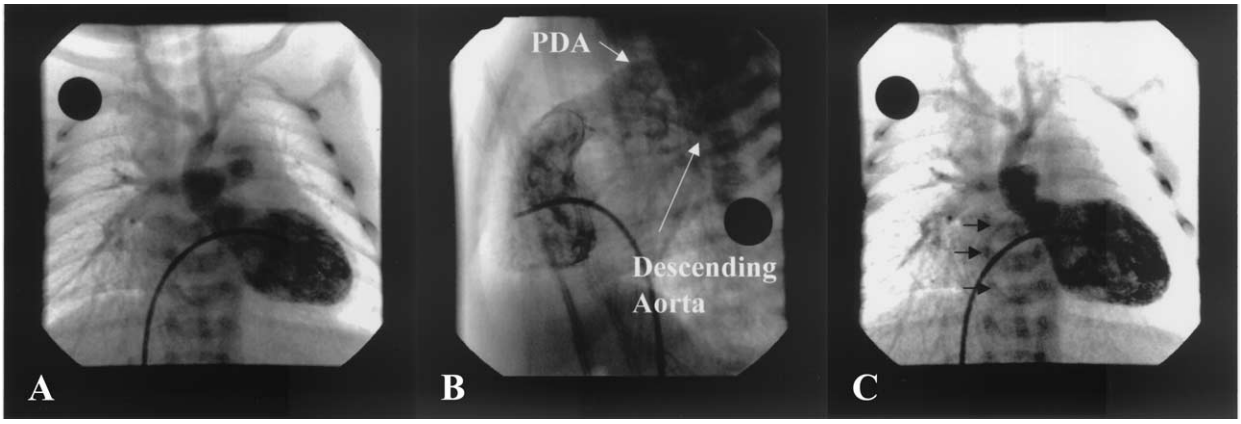

Figure 1. A, Left ventriculogram showing anomalous origin of the right pulmonary artery and interrupted aortic arch (type A) with intact ventricular septum. Contrast enters the left pulmonary artery through the APSD. B, Lateral view of right ventriculogram showing patent ductus arteriosus *(PDA)* and descending aorta. C, Left ventriculogram showing right (indicated by *arrows*), but not left, coronary artery.

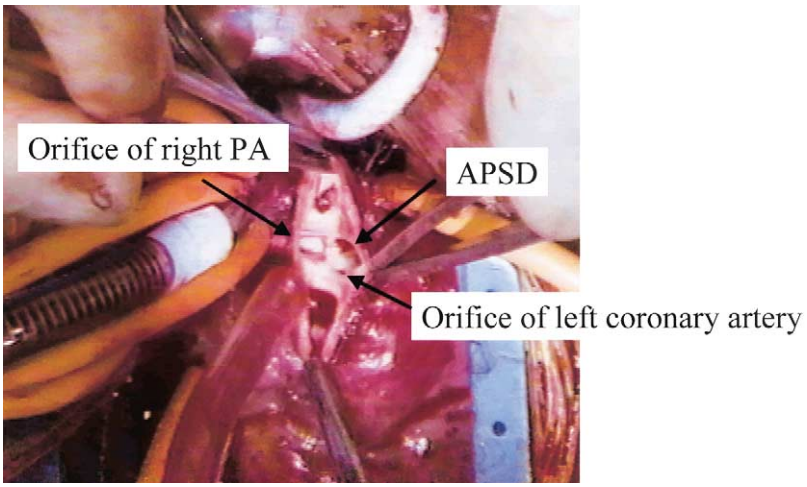

Figure 2. Intraoperative view of APSD and orifice of right pulmonary artery *(PA)*, with ascending aorta incised longitudinally. Note the small defect-like hole just below (caudal to) the APSD. Postoperatively, this was revealed to be the orifice of the left coronary artery.

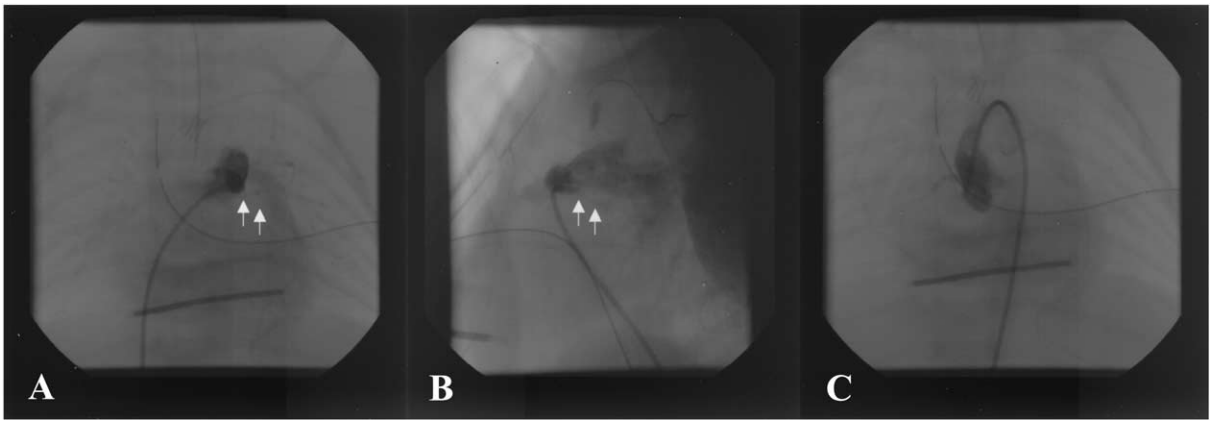

Figure 3. Postoperative pulmonary arteriogram (A, anteroposterior view; B, lateral view) showing anomalous origin of the left coronary artery from the main pulmonary artery (indicated by *arrows*). Because a simple pulmonary arteriogram did not provide a clear view of the coronary artery, a balloon-tipped catheter was used to better visualize the coronary artery. Ascending aortography shows only the right coronary artery (C).

bypass, pulmonary artery pressure and aortic pressure were 45 and 55 mm Hg, respectively. Although postoperative hemodynamics were stable during the first several days, left ventricular function gradually deteriorated, coinciding with a decrease in pulmonary artery pressure.

On the seventh postoperative day, electrocardiography showed depression of the ST segment in lead III and elevation of the ST segment in leads II and aV_F. At that point, we reviewed ventriculography and found that left ventriculography showed the right, but not left, coronary artery, suggesting coronary abnormality (Figure 1, *C*). However, right ventriculography did not show the left coronary artery arising from the main pulmonary artery (Figure 1, *B*). Therefore, we again performed angiography and confirmed the anomalous origin of the left coronary artery from the main pulmonary artery (Figure 3). The coronary artery orifice was hollowed out into a button shape and was transferred to the ascending aorta. The pulmonary artery was reconstructed with an autologous pericardial patch, and the ascending aorta was again directly anastomosed. Left ventricular function gradually improved after this operation, and the patient was discharged on the 40th postoperative day.

Discussion

To the best of our knowledge, this is the first report of Berry syndrome complicated by anomalous origin of the left coronary artery from the pulmonary artery. Richardson and colleagues[3] have classified APSD into 3 types according to the anatomy of the defect. Type I defects (classic APSD) are between the posteromedial wall of the ascending aorta just above the sinus of Valsalva

and the main pulmonary artery; thus the left coronary orifice is very near the defect. Anomalous origin of the left coronary artery has been detected in patients with type I APSD, although rarely.[3] There have been no reports of anomalous origin of the coronary artery in patients with type II or III APSD, in which the defect is located more cephalad on the ascending aorta and far from the sinus of Valsalva.[3,4] The anomalous origin of the coronary artery in the present case is therefore very unusual because the patient's Berry syndrome was classified as a combination of types II and III.

When anomalous coronary circulation coexists with other congenital malformations, particularly those associated with pulmonary hypertension, the initial presentation can be quite confusing and is often misinterpreted.[5] Surgical correction of associated malformations can unmask the coronary anomaly, which is then revealed by unexpected cardiac decompensation.[5] Failure to anticipate coronary anomaly can result in cardiogenic shock and death.[5] In the present case, relatively high pulmonary artery pressure persisting immediately after the first operation helped avoid acute manifestation of cardiac failure, but subsequent amelioration of pulmonary hypertension revealed the coronary anomaly. If we had anticipated the coronary anomaly in the present case, preoperative left ventriculography (Figure 1, *C*) showing only the right coronary artery would have caused us to further evaluate the coronary artery before or during the operation. Because in Berry syndrome it is difficult (for anatomic reasons) to perform aortography with a catheter positioned above the aortic valve, visualization of the coronary arteries can be limited. Main pulmonary angiography with an inflated balloon (which we performed at the second catheterization in the present patient) can help improve the clarity of identification of the coronary abnormality.

In summary, anomalous origin of the coronary artery from the pulmonary artery should be considered a possible serious complication of Berry syndrome and should be evaluated preoperatively, intraoperatively, or both.

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