Case Reports

Anomalous Origin of the Right Coronary Artery from the Pulmonary Artery

in Association with a Ventricular Septal Defect

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Origin of the right coronary artery from the pulmonary artery is a rare lesion occasionally found at angiography or autopsy. We report the rare preoperative diagnosis, in a child, of anomalous origin of the right coronary artery from the pulmonary artery, in association with a ventricular septal defect. The chest radiograph was normal, but auscultation revealed a continuous murmur at the left sternal border and electrocardiography showed right and left ventricular hypertrophy. A transthoracic echocardiogram depicted anomalous origin of the right coronary artery from the pulmonary artery. Color-flow Doppler echocardiography indicated possible right-coronary-artery–to–right-ventricle fistulae. Diagnosis was made by selective left coronary arteriography, which showed retrograde filling of the right coronary artery from collateral vessels. Selective left coronary arteriography depicted intercoronary flow, with no fistulae. Operative repair consisted of moving the proximal right coronary artery from its origin at the pulmonary trunk to the aorta. An associated procedure for correction of the ventricular septal defect was performed. The postoperative cardiac angiogram showed that the ventricular septal defect was closed and that flow through the right coronary artery was normal. Preoperative diagnosis of anomalous origin of the right coronary artery from the pulmonary artery is important, because this condition is surgically correctable. ***(Tex Heart Inst J 1997;24:226-9)***

Key words: *Child; collateral circulation; coronary circulation; coronary vessel anomalies/diagnosis; heart defects, congenital/diagnosis; pulmonary artery/abnormalities*

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When the left coronary artery (LCA) arises from the pulmonary artery,[1-3] the anomaly is highly symptomatic: ischemia of the left ventricle induces heart failure in infancy. However, when the right coronary artery (RCA) arises from the pulmonary artery, this apparently much rarer malformation[4-13] can escape detection if it is present as an isolated congenital defect (a few cases have been diagnosed by means of angiography[14,15]). Infants so affected can survive to adulthood without symptoms. Although lethal cardiac arrhythmia and RCA thrombosis have been reported as sequelae,[10] these events seldom occur in the absence of other congenital heart disease. Indeed there have been isolated reports of patients with anomalous origin of the RCA from the pulmonary artery, found in association with atrial septal defect, tetralogy of Fallot with patent ductus arteriosus, pulmonary atresia with intact ventricular septum, aortopulmonary window with and without a ventricular septal defect, and aneurysm of the sinus of Valsalva.[16-19]

The following report describes 1 additional case involving a patient who had anomalous origin of the RCA in combination with ventricular septal defect (VSD). He was diagnosed prior to operation and underwent a successful surgical repair.

Case Report

In September of 1995, a 4-year-old boy was referred for evaluation of a change in the pattern of a heart murmur. A systolic murmur, noted shortly after birth, had led to the diagnosis of a VSD when the patient was 2 months old. At this present (1995) admission, the boy had a few symptoms and a grade 2/6 continuous murmur along the left sternal border. The electrocardiogram (ECG) showed right and left ventricular hypertrophy, and the chest radiograph was normal. Transthoracic echocardiography showed anomalous origin of the RCA from the main pulmonary artery (Fig. 1), and color-flow Doppler echocardiography detected possible

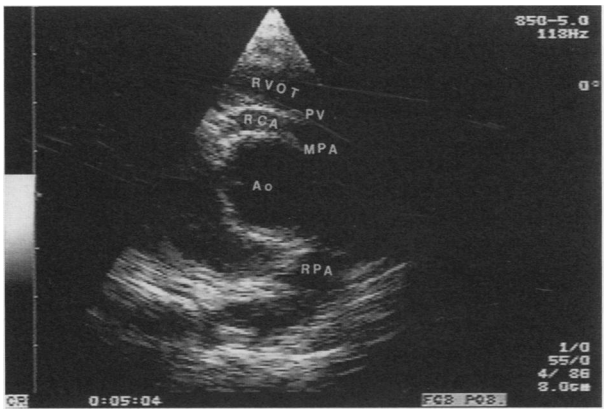

Fig. 1 *Two-dimensional echocardiogram, short-axis view, showing the origin of the right coronary artery (RCA) at the main pulmonary artery (MPA).*

Ao = aorta; PV = pulmonary valve; RPA = right pulmonary artery; RVOT = right ventricular outflow tract

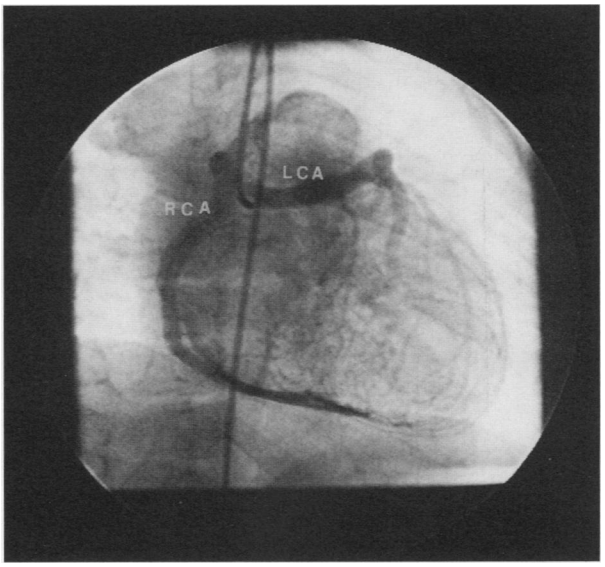

Fig. 3 *Selective left coronary angiogram in the right lateral view, showing the large right coronary artery (RCA) filling in a retrograde fashion through a network of vessels from the left coronary artery (LCA).*

RCA–to–right ventricle fistulae. When cardiac catheterization was performed, intracardiac pressures were normal.

The aortic root cineangiogram depicted a single left coronary artery arising from the aorta (Fig. 2). A selective LCA cineangiogram showed a well-developed network of large intercoronary collateral vessels and retrograde filling of a large RCA (Fig. 3), which drained into the main pulmonary artery, thus excluding the suspicion of fistulae raised by the echocardiogram. Right coronary artery cineangiography performed through the main pulmonary artery showed the contrast-material washout in the distal flow of the RCA (Fig. 4).

Early in 1996, the patient was admitted to the hospital for surgical correction of this malformation. The approach was through a median sternotomy. We used hypothermic cardiopulmonary bypass and achieved myocardial protection with cold blood cardioplegia. The VSD was closed with a patch of porcine pericardium through a right atriotomy. The large serpentine RCA, arising from the right side of the pulmonary artery, was dissected to a distance of 2 to 3 cm from its origin and was transposed to the ascending aorta, 2 cm above the sinuses of Valsalva. A button of tissue, including the orifice of the RCA, was excised from the main pulmonary artery, and a patch of porcine pericardium was used to reconstruct that vessel.

The postoperative course was uneventful and no heart murmurs were heard. Mechanical respiratory assistance was discontinued on the 1st postoperative day. The ECG showed normal sinus rhythm and a right bundle branch block. The patient was dis-

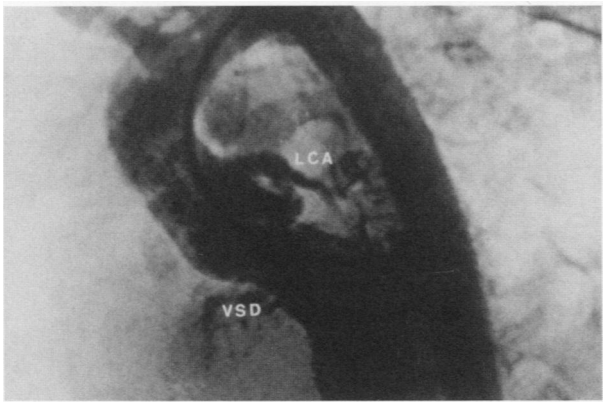

Fig. 2 *Left ventricular angiogram in the left lateral projection, showing a ventricular septal defect (VSD) and the origin of the left coronary artery (LCA) at the aorta.*

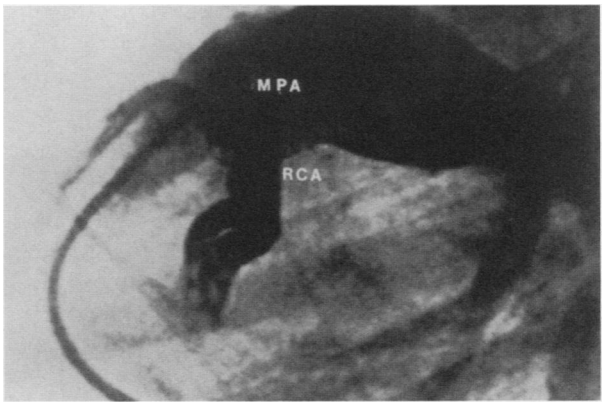

Fig. 4 *Pulmonary artery angiogram in the left lateral projection, showing the origin of the right coronary artery (RCA) at the main pulmonary artery (MPA).*

charged on the 7th postoperative day. One week later, cardiac angiography, performed to check the effectiveness of the surgical procedure, showed a closed VSD and a normal filling pattern of the RCA through its anastomosis to the aorta (Fig. 5).

Discussion

Anomalous origin of the LCA from the pulmonary artery is a well-known abnormality and progresses, in many cases, to severe ischemia in infancy. Anomalous origin of the RCA is less common, and those who have it are usually free of symptoms, with no angina or unusual ECG changes.[20] It is an occasional finding at necropsy.

The development of a large network of intercoronary collateral vessels between the LCA and the RCA meets the metabolic needs of the myocardium fed by the anomalous artery, and the direction of flow within the RCA depends on the pulmonary vascular resistance and the degree of development of collateral circulation.

Persistence of a left–to–right shunt might not be perceived if the patient does not have clinical symptoms, such as those caused by myocardial ischemia.

The existence of associated defects helps in making an earlier diagnosis,[16-18] thus permitting surgical correction in a more timely fashion, as occurred in our patient. Despite having a VSD, our patient had no clinical symptoms; it was the change in the pattern of the murmur (from systolic to continuous) that was critical in prompting further diagnostic exploration. The results of color-flow Doppler echocardiographic mapping, which at first were thought to indicate coronary–to–right-ventricle fistulae, were later explained by cineangiography, which confirmed the anomalous origin of the RCA and disclosed a network of LCA–to–RCA vessels.

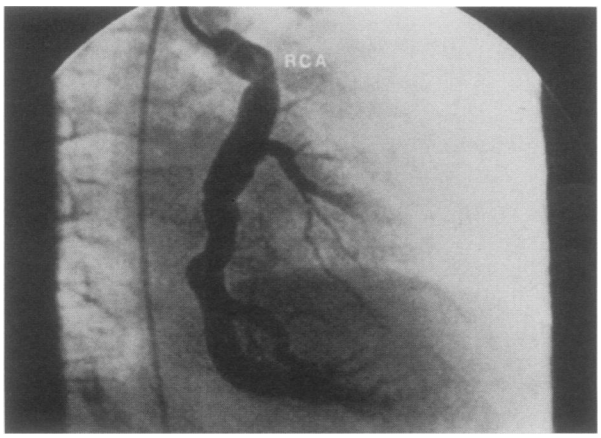

Fig. 5 *Postoperative selective aortogram showing the anastomosis of the right coronary artery (RCA) at the aorta and its normal filling pattern.*

To the best of our knowledge, anomalous origin of the RCA from the pulmonary artery has never before been reported in association with a VSD in a child.

Correction of the abnormality can be performed by simple ligation of the RCA at its point of origin at the main pulmonary artery, which enables the well-developed intercoronary blood flow to maintain perfusion of the RCA territory. Alternatively, if the anomaly is associated with aortopulmonary window, the surgeon can anastomose a flap of the pulmonary wall (including the ostrium of the RCA) to the aorta.[21] In our patient, however, reimplantation of the proximal RCA was undertaken on the anterolateral aspect of the ascending aorta. We rotated the proximal segment of the vessel counterclockwise, and no prosthetic interposition was necessary.[8,11,13] This procedure was chosen because it was anatomically feasible and provided the most physiologic form of repair.

Normalization of coronary blood flow after RCA reimplantation created a pressure balance between both coronary territories, producing involution of the LCA–to–RCA vascular network, as shown by postoperative echocardiograms and coronary angiography.

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