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Anomalous Origin of the Right Coronary Artery From the Pulmonary Artery With Aortopulmonary Window

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Abstract

This report describes a rare case of anomalous origin of the right coronary artery from the pulmonary artery associated with a large aortopulmonary window in a 2-month-old boy. The right coronary artery was exposed to systemic pressure and carried fairly well-oxygenated blood to the myocardium. Closure of the aortopulmonary window alone could have caused acute myocardial ischemia. The purpose of this case report is to describe successful diagnosis and management of anomalous origin of the right coronary artery associated with an aortopulmonary window. The pathological findings and the physiological effects, clinical importance, and method of correction used are discussed.

Keywords

aortopulmonary window, anomalous origin of right coronary artery from pulmonary artery

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Anomalous origin of the right coronary artery from the pulmonary artery (ARCAPA) associated with an aortopulmonary window (APW) is a rare and potentially dangerous combination of congenital heart defects. Because of a large left-to-right shunt at the APW level and increased pulmonary artery (PA) pressure, myocardial ischemia is not pronounced, and the presence of ARCAPA might be overlooked at the diagnostic stage. Closure of the APW alone might result in myocardial ischemia due to coronary artery steal in the presence of good collateral circulation. Alternatively, it could result in acute ischemia in the distribution of the right coronary artery (RCA) if intercoronary collaterals are not well developed.

The majority of reported cases[1-8] have been diagnosed by angiocardiography or discovered at the time of surgical repair of an associated defect. We report a case of a 2-month-old male infant with APW and ARCAPA in whom the diagnosis was made preoperatively by transthoracic echocardiography followed by a successful one-stage surgical repair.

A 2-month-old male infant weighting 3.8 kg was referred to Bakulev Cardiovascular Research Center with a history of congestive cardiac failure and persistent atrial tachycardia. Respiratory rate was 60 breaths/min with marked intercostal retractions and sternal bulging. His pulse rate was 170 beats/min. There was a systolic thrill at the left sternal edge in the third intercostal space and a grade 3/4 ejection systolic murmur at the upper left sternal border. The liver was enlarged. Electrocardiogram showed atrial tachycardia with a mean frontal QRS vector of +700 and biventricular hypertrophy (Figure 1). Chest radiography showed a dilated main PA, a prominent left ventricle, and plethoric lung fields. Cardiothoracic ratio was greater than 72% (Figure 2).

The echocardiographic evaluation showed dilatation of the left heart chambers. Left ventricular end-systolic volume was 92 mL/m^2. Left ventricular ejection fraction was 42%, with moderate mitral regurgitation. A large (approximately 8 mm) aortopulmonary defect could be seen in an anterosuperior location. Systolic pulmonary arterial pressure was 70 mm Hg. The RCA was identified arising between the posterior and the right sinus of the pulmonary valve, and color Doppler demonstrated retrograde flow into the main PA (Figure 3).

Cardiac catheterization and angiocardiography were performed prior to surgical intervention and demonstrated a large left-to-right shunt at the arterial level with systemic level

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Abbreviations and Acronyms

APW	aortopulmonary window
ARCAPA	anomalous origin of right coronary artery from the pulmonary artery
PA	pulmonary artery
RCA	right coronary artery

pressure in the PA (Table 1). There was an 8-mm APW between the inner curvature of the ascending aorta and the pulmonary trunk (Figure 5) The left coronary artery was normal with respect to origin and course (Figure 6), but the RCA originated anomalously from the right anterior aspect of the main PA proximal to the window (Figures 7 and 8). A small atrial septal defect and patient ductus arteriosus were also present. The angiographic diagnosis of ARCAPA in patients with APW can be difficult to establish because both great arteries fill simultaneously.

The operation was performed through a trans-sternal approach, with cardiopulmonary bypass, deep hypothermia, and cold crystalloid cardioplegia. The aorta was cannulated

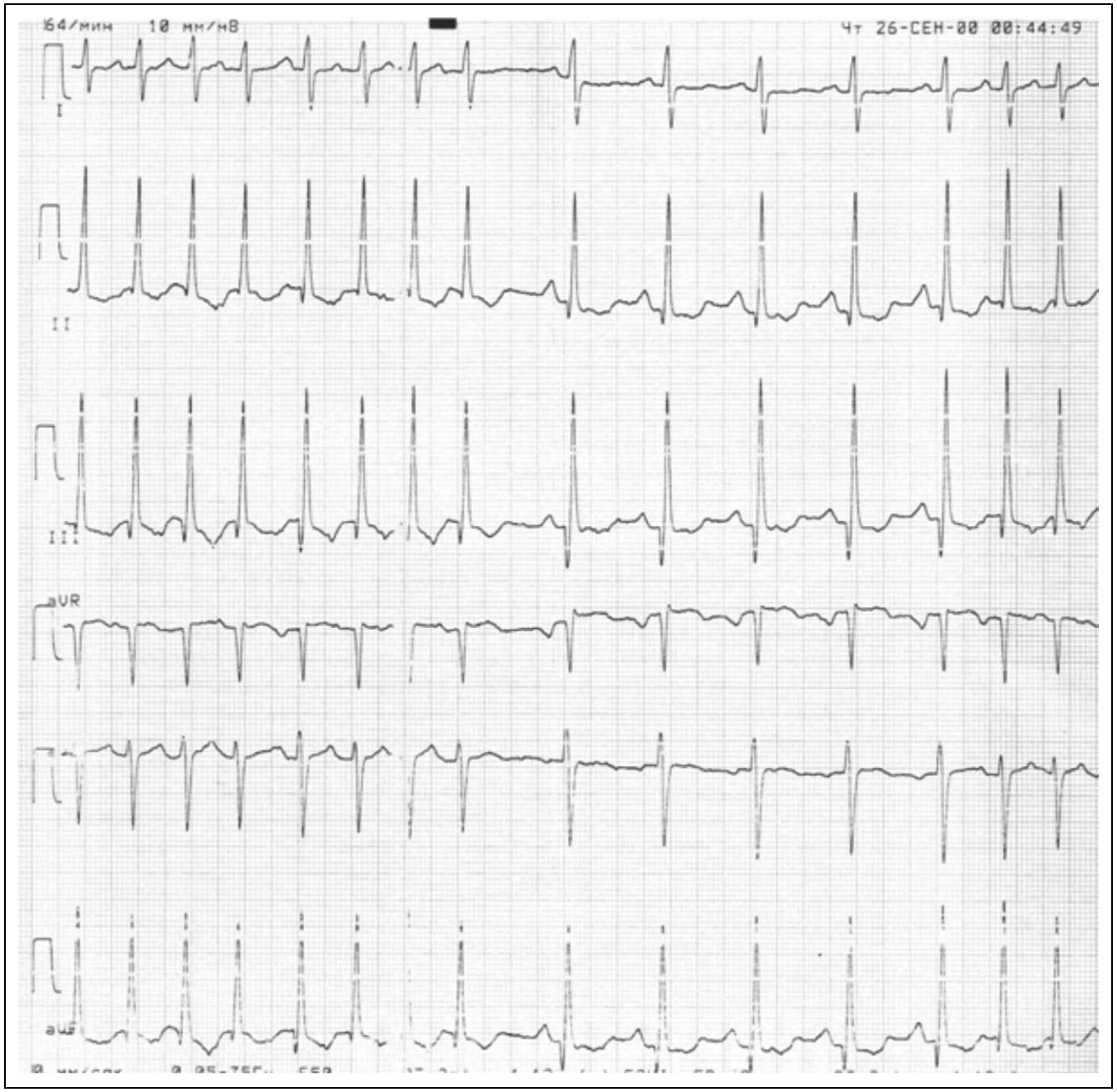

Figure 1. Electrocardiogram.

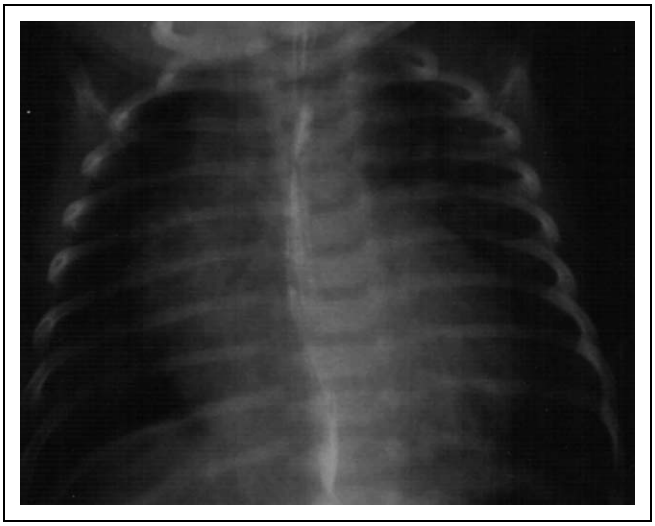

Figure 2. Chest radiography.

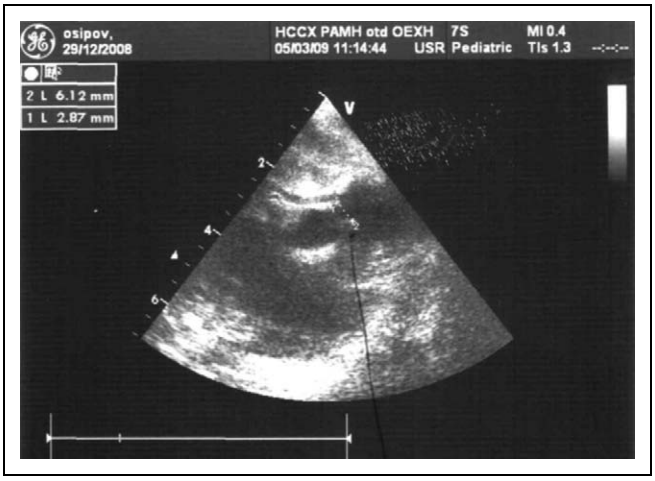

Figure 3. The white dotted line points toward the right coronary artery arising between the right and posterior sinus of the pulmonary valve; the black line shows the aortopulmonary window.

Table 1. Cardiac Catheterization Data

	Pressure, mm Hg
Left ventricle	107/12
Aorta	107/41/78
Right ventricle	78/0/6
Main pulmonary artery	78/37/52
Right atrium	6/2/4

distally, well beyond the level of the APW. Bicaval venous drainage was established. A venting catheter was inserted into the left atrium. After initiating cardiopulmonary bypass, tourniquets were placed and snugged around the right and left branch pulmonary arteries to ensure satisfactory systemic perfusion and delivery of cardioplegia solution to both coronary arteries. Following cardioplegic arrest, an oblique incision was made in the main PA (Figure 9). An APW (6 mm in diameter) was

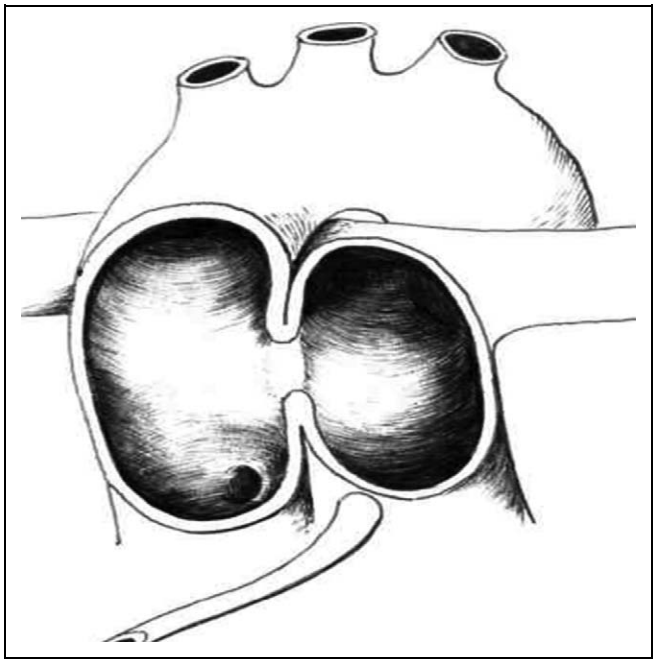

Figure 4. Illustration of the aortopulmonary window and the anomalous origin of the right coronary artery from the pulmonary trunk.

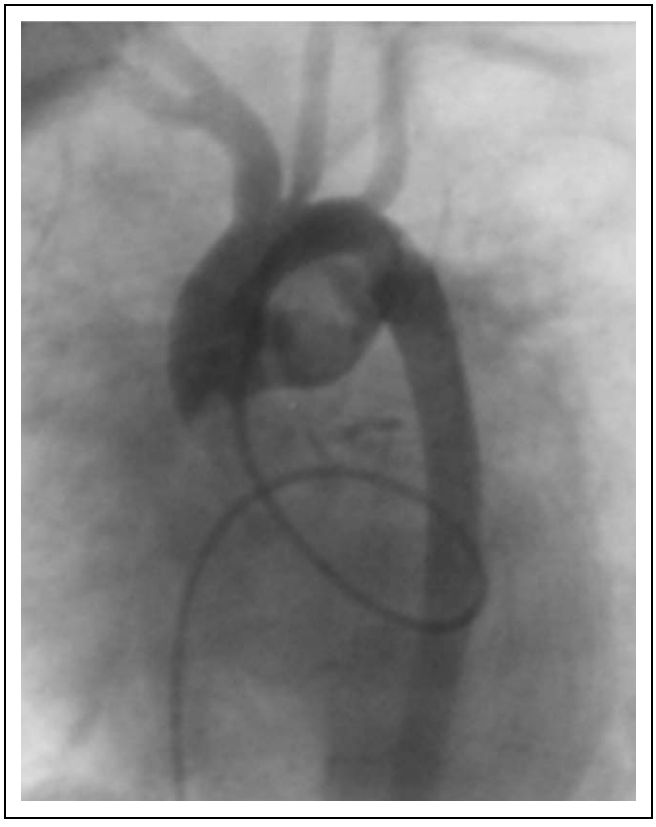

Figure 5. Angiogram demonstrating the aortopulmonary window between the inner curvature of the ascending aorta and the pulmonary trunk.

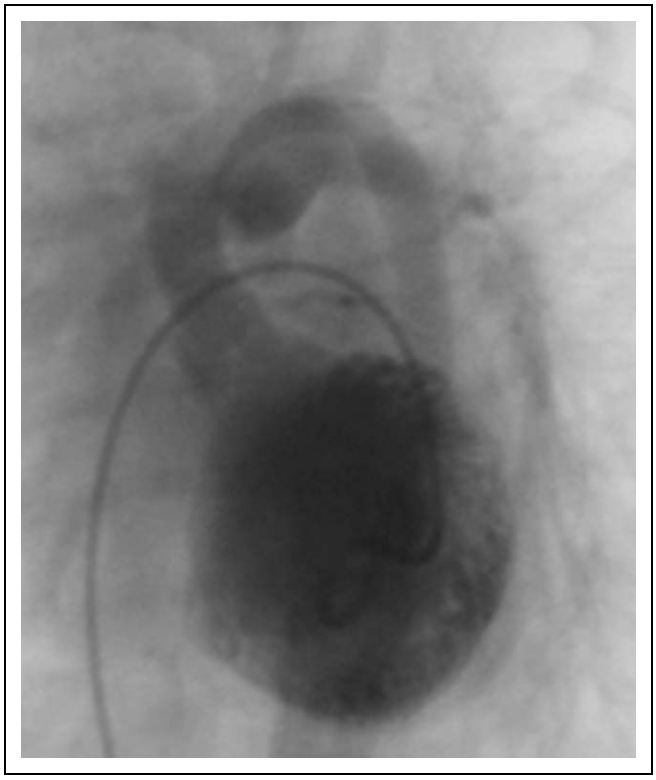

Figure 6. Left ventriculogram demonstrating normal aortic origin and course of the left coronary artery.

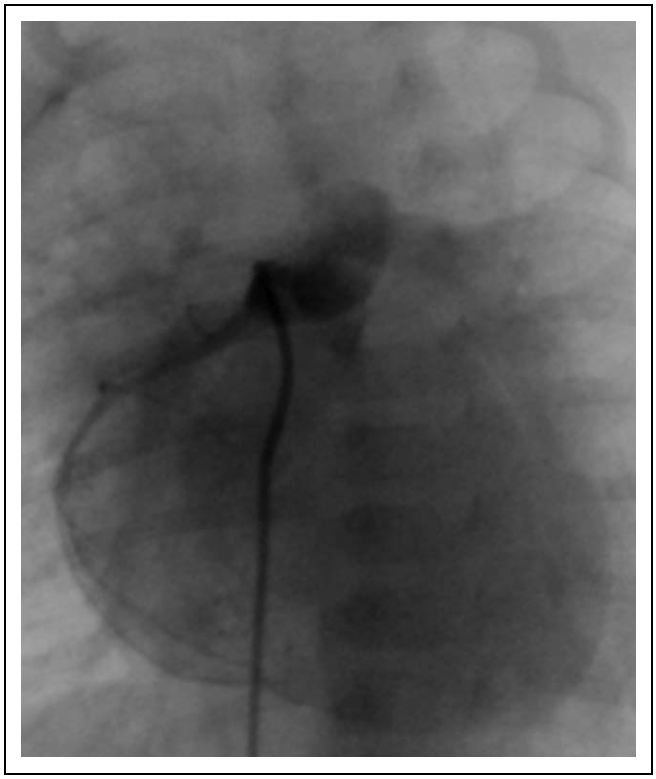

Figure 7. Selective angiogram demonstrating the right coronary artery originating anomalously from the right anterior aspect of the main pulmonary artery.

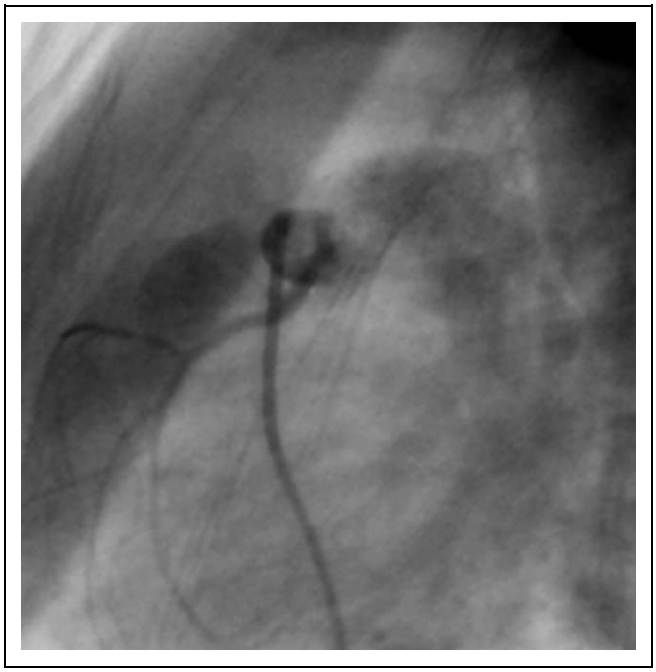

Figure 8. Lateral view of angiogram showing the right coronary artery originating from the main pulmonary artery.

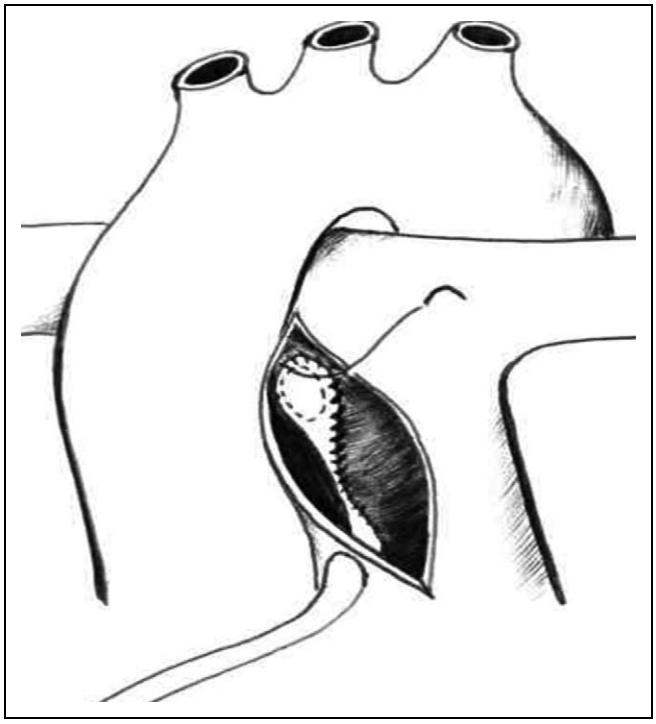

Figure 9. Illustration of the method of "transposing" the anomalous right coronary artery from the pulmonary trunk to the aorta with simultaneous closure of the aortopulmonary window.

found just above the pulmonary valve. There was anomalous origin of the RCA from the right anterior aspect of the main pulmonary trunk with the ostium situated 0.5 cm inferior to the lower margin of the window (Figure 4). The vessel was of

normal size. The left coronary artery was normal. The position of the RCA ostium was considered unfavorable for direct transfer to the aorta. A polytetrafluoroethylene baffle was sewn into the pulmonary trunk, incorporating the window superiorly and the RCA orifice inferiorly, thus creating a channel for blood to flow from the aortic root, beneath the baffle, and into the RCA. This simultaneously achieved closure of the window and association of the orifice of RCA with the aortic root (Figure 9). The patent ductus arteriosus was ligated, and the atrial septal defect was sutured trough the right atrium. Aortic cross-clamp time was 58 minutes, and total cardiopulmonary bypass time was 155 minutes with minimal temperature 22°C. After correction, the left ventricular pressure was 85/4-5 mm Hg and the right ventricular pressure 42/4 mm Hg. The postoperative course was uneventful, and the patient was discharged 12 days later.

Discussion

The association of anomalous origin of the RCA from the pulmonary trunk with APW is very rare. To our knowledge, only 7 cases have been reported. In our case, the anomalous vessel originated from the PA in close proximity to the APW, with a proximal course in front of the ascending aorta before reaching the right atrioventricular groove. Both coronary arteries were of normal caliber, with no significant collateral vessels. Because of the presence of a large APW, the anomalous RCA was exposed to systemic pressure and carried fairly well-oxygenated blood from the pulmonary trunk to the myocardium. This situation is quite different from that seen in patients in whom anomalous origin of the RCA from the PA is an isolated lesion. The left coronary artery in those patients is dilated and tortuous and, in most cases, gives several large interarterial anastomotic channels to the RCA system. The presence of a left-to-right shunt through a low-resistance coronary artery system can cause a pulmonary steal from the myocardium. The clinical picture in patients with an additional APW is dominated by the signs of the left-to-right shunt through the window. Prognosis depends mainly on the magnitude of the left-to-right shunt and pulmonary vascular resistance. Early diagnosis and surgical correction are essential to prevent the onset of intractable congestive heart failure and irreversible pulmonary vascular disease. The anomalous RCA must be identified and dealt with at operation, because closure of the window without addressing the coronary anomaly can cause acute ischemic changes or myocardial infarction from a sudden decrease in right coronary pressure in the absence of well-developed anastomotic channels from the left coronary artery. Furthermore, late complications from myocardial ischemia may occur. The anomaly can be corrected by a plastic procedure which should allow normal growth.

Accurate preoperative diagnosis and full understanding of the pathology are essential for an optimal surgical correction, which should be achieved early to prevent development of severe pulmonary vascular changes.

Declaration of Conflicting Interests

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