CASE REPORTS

Pulmonary Atresia with Intact Ventricular Septum and Ventriculocoronary Communications: Surgical Significance

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SUMMARY  The first stage of a repair of pulmonary atresia with intact ventricular septum (type I) was attempted in a 2-day-old infant. At surgery, decompression of the hypertensive small right ventricle was followed by a sudden loss of myocardial contractility and death. Postmortem examination revealed a fistula with a large orifice in the right ventricular infundibulum that communicated directly with the left main coronary artery. Severe hypertensive changes indicative of abnormally high perfusion pressure were noted in the distal left coronary artery branches. The clinical course suggests that the effect of relieving right ventricular outflow obstruction was a reduction of left main coronary artery blood flow, resulting in fatal intraoperative myocardial ischemia. This unusual case draws attention to the anomalous ventriculocoronary communications often present in pulmonary atresia and their potential for limiting a successful surgical repair.

IN SURGICAL CORRECTION of pulmonary atresia with intact ventricular septum, several factors appear to limit a successful outcome. These include the site of atresia (valvular or infundibular or both), the anatomic size of the right ventricular chamber, the competence and size of the tricuspid valve orifice and the adequacy of the opening between the right and left atrium to allow unrestricted right-to-left shunting. Right ventricular myocardial sinusoids that communicate directly with the coronary system are usually identified in this malformation of the right heart. Although Freedom and Harrington speculated that they may result in postoperative myocardial ischemia, the significance of their role in surgical outcome is not well defined.

In this report, we present a case of pulmonary atresia with intact septum and discuss the potential role of ventriculocoronary connections on the unfavorable surgical outcome.

Case Report

BF, a 3700-g full-term baby, was referred at 40 hours of age for evaluation of cyanotic congenital heart disease. He was the first-born infant of a 22-year-old mother, delivered by cesarean section because of cephalopelvic disproportion. Cyanosis was initially noted at 2 hours and a heart murmur was heard at 12 hours of age. Physical examination revealed a well-developed but deeply cyanotic infant without respiratory distress. His pulse was 120 beats/min, his respiratory rate 60 breaths/min, and his systolic blood pressure was 64 mm Hg. He had a quiet precordium. A grade 3/6 systolic regurgitant murmur was audible at the xiphoid and along the lower left sternal border. A diastolic rumble was also heard at the xiphoid, and his second heart sound was single. The lungs were clear and the liver was palpable 2 cm below the right costal margin, but otherwise the abdomen was normal. The peripheral pulses were easily palpable and symmetrical. An x-ray film of the chest revealed a large heart with decreased pulmonary flow. The ECG showed an axis of +120° with left ventricular predominance and little expression of right ventricular forces. By blood gas analysis, pH was 7.35, PCO₂ 19 mm Hg, and PO₂ 24 mm Hg with 42% saturation in room air; these values did not change in an oxygen environment. The echocardiogram was compatible with the clinical diagnosis of pulmonary atresia with intact ventricular septum; emergency cardiac catheterization (fig. 1) and right ventricular angiography confirmed the diagnosis. Contrast medium passed from the right ventricular chamber to the aorta, in retrospect via the left coronary system (fig. 2). The ductus arteriosus was not imaged. Oxygen saturation decreased from 56% in the left ventricle to 35% in the descending aorta. Because of previous unsatisfactory results with systemic-to-pulmonary shunting alone, the patient was taken directly to the operating room and placed on cardiopulmonary bypass with systemic hypothermia to 24°C and myocardial protection with cardioplegia solution. The right atrium was opened and the interatrial septum resected. The pulmonary artery was then opened and the valve was found to be completely closed, with approximately 1 cm of infundibular muscle separating it from the small right ventricle. A sized cork bore was used to resect the muscle and valve tissue, thereby connecting the pulmonary artery with the right ventricular chamber. A 4-mm side-to-side shunt from the aorta to the right pulmonary artery was also completed and the patent

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ductus arteriosus was ligated. The aortic cross-clamp time was only 18 minutes, but myocardial contractility after the procedure was inadequate to maintain the circulation and the child died.

**Autopsy Findings**

Postmortem examination of the heart confirmed the diagnosis of pulmonary atresia with intact ventricular septum. The right ventricular cavity measured $11 \times 9$ mm and the myocardium $13$ mm in its sinus portion; the left ventricular free wall was $6$ mm. The tricuspid valve was normally formed, competent and had a stenotic annulus $15$ mm in circumference. The comparable mitral annular circumference was $23$ mm. The thin-walled hypoplastic pulmonary trunk contained remnants of commissural ridges above the site of exci-

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**Figure 1.** Cardiac catheterization data and its relationship to the central circulation. Preoperatively, a step-down in oxygen saturation from left ventricle (LV) ($56\%$) to aorta (AO) ($35\%$) was present. Solid arrows show the presumed effect of surgical right ventricular decompression on perfusion of the left coronary system. LA = left atrium; RA = right atrium; ART = arterial; PULM = pulmonary; SYST = systemic; LMCA = left main coronary artery.

**Figure 2.** Anteroposterior right ventriculogram showing contrast entering the left main coronary artery (LMCA) through the fistula and subsequently appearing in the aorta (arrows). Artist's sketch with hatched internal image depicts relationships of the fistula (F) to the left anterior descending coronary artery (LAD) and to the small main pulmonary artery (MPA). Arrow denotes direction of flow from the right ventricle (RV) into the fistula. RA = right atrium; RPA = right pulmonary artery; LPA = left pulmonary artery; PDA = patent ductus arteriosus.
sion of the atretic valve and infundibular muscle. A silk ligature occluded the ductus arteriosus, which was 20 mm long and 7 mm in circumference. There was a striking external bulge on the anterosuperior surface of the heart (fig. 3A). Upon dissection, this prominence overlay a large fistula connecting the right ventricle with the left main coronary artery. The ostium of the fistula in the right ventricular infundibulum was

![Image of heart](http://circ.ahajournals.org/)

**FIGURE 3.** Left anterior views of heart at autopsy. (A) Central bulge (arrows) and prominent coronary artery network over the left ventricle. (B) Unroofed fistula connecting the right ventricular infundibulum with the left main coronary artery. The tip of the white pointer is at the distal end of the fistula.

11 mm in circumference and led directly into a single channel, which gradually tapered and emerged on the epicardium. Here, the lumen (fig. 3B) was 5 mm in circumference and surrounded by a dense, fibrous wall that merged with the enlarged left main coronary artery at its bifurcation into the anterior descending and circumflex branches. The ostium of the left coronary artery in the aorta was enlarged to 11 mm in circumference, and the left main coronary artery was three times larger in caliber than the proximal right coronary artery, which suggests that the blood flow in the left main coronary artery was greater than normal. Distally, the left coronary artery formed a prominent network over the entire left ventricular surface. Microscopically, both extra- and intramural branches showed adventitial fibrosis, medial muscular hypertrophy and intimal fibrous thickening with luminal narrowing (fig. 4). These changes were not identified in the right coronary artery branches. An area of dimpling over the distal left anterior descending coronary artery suggested another communication between the coronary vascular bed and the right ventricular chamber. The underlying myocardium contained an especially striking pattern of sinusoidal spaces, smaller numbers of which were present in all sections of the right ventricle. A second single channel connecting to the coronary vascular bed to correspond with the communication in the area of the cardiac apex on the preoperative angiogram (fig. 2) could not be documented at autopsy.

Upon gross inspection of the left ventricle, the entire cut surface of each papillary muscle belly was yellow; microscopically, there was a confluent zone of coagulative necrosis with patchy dystrophic calcification. However, there was no inflammatory infiltrate or evidence of resolution or fibrosis. The finding of established infarction 24-48 hours old suggests that the ischemic insult occurred immediately after birth. No other zones of infarction could be identified in representative sections of left and right ventricular myocardium.

**Discussion**

The right ventricle is a small, thick-walled chamber in the majority of patients with pulmonary atresia and intact ventricular septum. More than 50% of the patients in this subgroup (type I) have a competent tricuspid valve, and radiopaque dye can usually be shown to enter the coronary vascular bed from a closed, obstructed right ventricle at angiography. The generally accepted mechanism for this anomalous communication is that blood at abnormally high pressure forces channels normally present in the developing heart to remain patent. These channels have been well documented in autopsy sections of the right ventricular myocardium where compressed, slit-like spaces are commonly identified. The channels represent persistence of the intertrabecular spaces and sinusoids of the embryonic blood bed, which connects with the coronary vascular bed and form an anastomosis between it and the ventricular lumen. The most common anatomic pattern of these connections is a confluence of right ventricular sinusoids that
coalesce into a single vessel and subsequently connect with the left coronary system, often by the left anterior descending branch. At the site of connection there may be dimpling of the cardiac surface.

Cord-like thickening of the coronary vessels on the surface of the heart has also been observed, although its significance has been uncertain. In the case reported here we found thickening of extramural coronary artery branches involving only the divisions of the left main coronary artery. The right coronary artery and its external ramifications, which did not connect directly with the right ventricle, were normal. It seems reasonable to assume that the changes in the left coronary system reflect longstanding intrauterine perfusion of these vessels at high pressure from the right ventricle. This perfusion probably derived not only from the large single noncompressible fistula, but also from multiple myocardial sinusoids connecting the apical region of the right ventricle to the distal left anterior descending coronary artery. Further, the resistance provided by intramural narrowing in the left coronary system favored retrograde flow in the left main coronary artery toward the aorta; this finding was demonstrated angiographically and is supported by the greatly increased size of this artery at autopsy. These observations suggest that blood flow in the left coronary system was dependent on high right ventricular pressure. The preoperative cardiac catheterization and angiography support this flow pattern (fig. 1).

Apart from functioning as an egress for blood from the right ventricle and forming the anatomic basis for a right-sided shunt, the anomalous connections between the right ventricular lumen and coronary arteries may contribute to myocardial ischemia by impeding normal diastolic filling of the coronary arteries from the aorta in the unoperated state. We have morphologic evidence of this in the finding of recent infarction involving the left ventricular papillary muscles. Microscopic findings are consistent with the infarction having occurred soon after birth, probably resulting from the postnatal decrease in oxygen content of the blood in the right ventricle.

The effects of the ductus arteriosus on diastolic coronary perfusion should also be considered. When patency persists after birth, it may provide a steal from the aorta once the pressure and resistance in the pulmonary circulation decrease, and thus might contribute to a reduction in coronary flow. Although anatomic and angiographic findings in the case described, the ductus arteriosus was not seen during angiography, which suggests that it was not the source of a significant coronary steal.

The existence of a significant right ventricular-to-coronary arterial shunt was of major surgical importance. The pathologic and hemodynamic data indicate that coronary perfusion was at least partly maintained by high pressure in the right ventricle. The surgical approach resulted in decompression of this chamber. Although not proved, it would be expected that the left main coronary arterial perfusion deprived of a portion of right ventricular supply would be compromised and could have led to global myocardial ischemia. The more effective operation was in reducing the right ventricular pressure by extensive valvulotomy and infundibulotomy, the more likely it was to decrease left coronary perfusion. Although the possibility of a postoperative steal from the left coronary system to the right ventricle could also be raised in this case, it cannot be proved.

Considering the anatomy, one could suggest that the fistula should also be closed at surgery. However, two factors would have made this approach impractical in the case presented here. First, the extensive hypertensive narrowing of the left ventricular vascular bed suggests that after closure of the fistula, normal coronary perfusion pressures from the aorta would have been inadequate to maintain myocardial coronary perfusion. Second, the usually diffuse fistulous communications in the newborn infant might preclude their surgical closure. In addition, there is
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evidence that the intramyocardial connections in time close spontaneously by progressive sclerosis. 20

The unusual sequence of events in this case suggests that the surgical approach in patients with pulmonary atresia who have fistulas should be different from that in patients who do not. In patients with significant sinusoidal-coronary shunts, a hypertensive right ventricle and an intact ventricular septum, a staged approach may be indicated. After successful balloon atrial septostomy, a systemic-to-pulmonary shunt procedure should be considered (stage I). Before opening of the right ventricular outflow tract (stage II), serious consideration should be given to occluding the significant left coronary artery–right ventricular fistulas. By preventing a reduction in left coronary perfusion, left ventricular ischemia or infarction may be avoided.

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