Coronary Artery-Right Ventricular Communication Associated with Pulmonary Atresia and Ventricular Septal Defect


SUMMARY
A case of pulmonary atresia and right ventricular-left coronary artery fistulae with an associated ventricular septal defect is described. Hemodynamic findings like those reported in similar cases, but with an intact ventricular septum are also described. The implications of these observations in the genesis of right ventricular-coronary artery fistulae are discussed.

Additional Indexing Words:
Pulmonary stenosis Congenital heart disease Malformations

The genesis of communications between the right ventricle and coronary arteries commonly associated with pulmonary atresia remains uncertain. Edwards\(^1\) attributed the persistence of these communicating sinusoids to increased right ventricular pressure and emphasized the importance of right ventricular outflow obstruction, intact ventricular septum, and competence of the tricuspid valve. Other authors,\(^2\)\(^-\)\(^5\) based on the earlier work of Grant,\(^6\) have suggested that inadequate early right ventricular maturation fails to occlude the embryonic sinusoids which then persist as right ventricular to coronary artery fistulae. This report concerns a case in which pulmonary atresia, right ventricular-coronary artery fistulae, ventricular septal defect, and malformation of the tricuspid valve were all present.

Report of a Case
B. A. was 4 days old when first seen at the University of Colorado Medical Center. He was the result of a 40-week uncomplicated gestation, weighing 7 lbs. 8 oz. at birth. A heart murmur was first noted about 24 hours following delivery. Tachypnea and fatigue during feeding occurred on the third postpartum day.

Physical examination revealed a Negroid male infant who weighed 3,450 g. Respiratory rate was 120 and heart rate 136 per minute. The pulses were normal. There was no lift or thrust referable to enlargement of either ventricle and no thrill was palpable over the precordium or neck. A grade II/VI to-and-fro murmur was present, loudest in the third intercostal space, 1 cm lateral to the left sternal margin. The second heart sound was single and normal in intensity. The lungs were clear to auscultation, liver and spleen were not enlarged, and there was no edema.

Chest x-ray demonstrated prominence of the cardiothymic silhouette. The great vessels were not demonstrated, and the pulmonary vascularity appeared diminished. There was electrocardiographic evidence of right atrial and right ventricular enlargement (fig. 1).

The patient was digitalized with digoxin and maintained in an atmosphere of humidified oxygen. Tachypnea without objective evidence of congestive heart failure persisted.

Cardiac catheterization was performed at 5 days of age. The catheter passed from the right ventricle through a ventricular septal defect into the aorta. The pulmonary artery could not be
entered. Pressures were 63/48 mm Hg in aorta, 96/0 to 14 mm Hg in the right ventricle, and a mean of 5 with an a wave of 9 and a v wave of 7 mm Hg in the right atrium. Blood oxygen saturations were 59% in the superior vena cava, 67% in the inferior vena cava, 76% in the right atrium, 71% in the right ventricle, 79% in the left atrium, and 82% in the aorta.

Cineangiograms of the right ventricle (fig. 2) revealed a small right ventricular chamber with sinusoidal communications to the left coronary artery. The aorta was opacified from this source. The pulmonary artery was not visualized. A small jet of contrast media directed posteriorly through a ventricular septal defect was seen in the lateral projection.

Persistent tachypnea and progressive fatigue complicated the patient’s course and at age 15 days, he died during an apneic spell.

Postmortem examination revealed pulmonary atresia, a right ventricular-left coronary artery fistula, ventricular septal defect, downward displacement of the tricuspid valve, and congestion of the lungs, liver, and spleen (figs. 3 and 4). The combined weight of the heart and lungs was

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Figure 1

Electrocardiogram at age 5 days demonstrating right atrial enlargement and right ventricular hypertrophy.

Figure 2

The left lateral projection of a right ventricular cineangiocardiogram demonstrating a small right ventricular (RV) chamber from which the large anterior coronary fistula originated and subsequently opacified the aorta (A). A small posteriorly directed jet from right ventricle through the intraventricular septal defect (VSD) is also demonstrated. A blind infundibulum (I) may be noted below the coronary artery (CA).
The heart with the right ventricle incised demonstrating the rather large ventricular septal defect (VSD) and adjacent tricuspid valve (TV). The atretic pulmonary artery (PA) and the area of epicardial sclerosis overlying the fistulous anterior descending branch of the left coronary artery (CA) and orifice of the fistula (F) are also demonstrated.

130 g. The right atrium was dilated, measuring 3 to 4 mm in thickness. The foramen ovale measured 5 mm in greatest diameter. The tricuspid valve was displaced downward into the right ventricle, the chamber of which measured 1 cm in greatest dimension. The right ventricular wall was markedly hypertrophied to 1.5 to 2 cm in greatest thickness. There was a 4 to 5 mm defect in the membranous ventricular septum. The pulmonic valve was not patent and there was a fibrous, atretic pulmonary artery. The pulmonary veins and left atrium were normal. The mitral valve was 4 cm in circumference and the cusps were thin and delicate. The left ventricle measured 1 cm in greatest thickness. There was a 2.5 by 3 cm tricuspid aortic valve, with normal leaflets. The coronary arteries arose normally from the aorta. The left coronary ostium and left main coronary artery were markedly dilated. This dilated vessel continued beyond the origin of a normal circumflex branch as a fistula to the apical portion of the right ventricle. A wide linear zone of fibrosis overlying this fistula was present in the anterior interventricular sulcus. There was a right aortic arch, and a left innominate and a right subclavian artery. Prominent bronchial communications to both lungs were present.

Discussion
This case illustrates the rather common association of coronary artery-right ventricular communication with pulmonary atresia. The presence of a ventricular septal defect, in addition, is uncommon and presents an interesting point concerning the genesis of
coronary artery-right ventricular fistulae in pulmonary atresia.

If Edwards' concept concerning the role of elevated right ventricular pressure in preserving ventricular-coronary artery communications is correct, it would appear that a ventricular septal defect could sufficiently decompress the right ventricle to make this combination of lesions improbable. Kaufman and Anderson have collected 10 cases and have recently added an eleventh case of coronary arterial-right ventricular fistulae associated with pulmonary atresia or severe pulmonary stenosis. The ventricular septum was intact in eight of the 11 cases. Thus, while the statistical likelihood of an intact ventricular septum is much increased by the presence of coronary-ventricular fistulae, the cases of Grant, Guidici and Becu, Muir, and the case presented here are notable exceptions.

Few hemodynamic observations have been made in patients with an intact ventricular septum, and the authors are not aware of any such studies among the small group of patients with an associated ventricular septal defect, as reported here. Anselmi and associates reported angiographic evidence of flow from the right ventricle through an anomalous coronary artery to the aorta and diastolic regurgitation from the aorta to the right ventricle in a patient with pulmonary atresia, right ventricular-coronary artery communication, and an intact ventricular septum. Sissman and Abrams have published angiographic documentation of a similar flow in a patient with pulmonary atresia and a coronary artery-right ventricular fistula.

Hemodynamic observations reported here document systolic and diastolic pressure gradients between the aorta and right ventricle consistent with similar bidirectional blood flow through the right ventricular-coronary arterial communication. An angiocardiogram from the right ventricle clearly documented flow from the right ventricle through the left coronary artery to the aorta during systole and regurgitant diastolic flow.

From these observations, it is apparent that the ventricular septal defect in this patient did not alter the pattern of right ventricular-coronary arterial blood flow as observed in cases with an intact ventricular septum. The functional size of the ventricular septal defect was sufficiently small to preserve a systolic pressure gradient between the right ventricle and the aorta which resulted in this pattern of flow and perhaps caused preservation of the right ventricular-coronary arterial communication. While the ventricular septal defect was moderate by anatomic measurement, it was small angiographically. In this case, the disparity may be secondary to apposition of the septal leaflet of the downwardly displaced tricuspid valve and the ventricular septal defect. Neufeld and associates have postulated a similar mechanism in three cases of Fallot's tetralogy with anomalous tricuspid valves, which clinically and hemodynamically simulated pulmonary stenosis with an intact ventricular septum. No mention was made of right ventricular-coronary artery fistulae in these patients.

The presence of right ventricular-coronary artery fistulae in association with pulmonary atresia thus does not exclude a coexisting ventricular septal defect. Neither does the presence of a ventricular septal defect invalidate Edwards' concept of pathogenesis of right ventricular coronary artery communication, provided the intraventricular septal defect is sufficiently small or is rendered functionally small by the tricuspid valve and a high pressure gradient between the right ventricle and aorta is preserved as in this case.

Acknowledgment

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References


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**Foetal Circulation—Harvey, 1628**

For the right receiving the blood from the ear, thrusts it forth through the *vena arteriosa,* and its branch called *canalis arteriosus,* into the great arterie. Likewise, the left at the same time by the mediation of the motion of the ear, receives that blood, which is brought into the left ear through that oval hole from the *vena cava,* and by its tension and constriction thrusts it through the root of the Aorta into the great arterie likewise. So in Embryons whilst the lungs are idle, and have no action nor motion (as if there were none at all) Nature makes use of both the ventricles of the heart, as of one for transmission of blood. And so the condition of Embryons that have lungs and make no use of them, is like to the condition of those creatures which have none at all.—*The Anatomical Exercises of Dr. William Harvey: De Motu Cordis 1628; De Circulatione Sanquinis 1649* (first English text). Edited by Geoffrey Keynes. London, The Nonesuch Press, 1653, p. 46.
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