Bidirectional Shunting in a Coronary Artery-Right Ventricular Fistula Associated with Pulmonary Atresia and an Intact Ventricular Septum

By Norman J. Sissman, M.D., and Herbert L. Abrams, M.D.

DESCRIPTIONS of anomalous arteriovenous-like communications between the right ventricle and coronary arteries in the presence of pulmonary atresia or severe pulmonary stenosis have been published with increasing frequency since the case report and review of the literature by Williams et al.1 in 1951. The majority of patients with this pathologic complex die in early infancy and are diagnosed at autopsy.

Edwards' group1 speculated on the physiology of blood flow through the anomalous communicating vessels but lacked the opportunity to confirm their theories. Anselmi et al.2 described the direction of blood flow during systole and diastole through the coronary artery in their case, but did not illustrate it.

The following case of a 6-year-old boy with this combination of defects is reported to demonstrate, by means of cineangiography, the dynamics of flow through an anomalous right coronary-right ventricular communicating vessel, and to describe the clinical features that led to the correct diagnosis during life.

Case Report

J.W.S. (P.A.S. no. 9-67-53) was born on August 1, 1958. On the first day of life, he was noted to be cyanotic and to have systolic and diastolic murmurs over the precordium. During the first 4 months of life, he had increasingly severe cyanosis and dyspnea. He was diagnosed, at another hospital, as having a tetralogy of Fallot and on December 19, 1958, an end-to-end anastomosis between the left subclavian and the left pulmonary arteries was performed. Although the anastomosis remained patent and the patient was symptomatically improved, he remained cyanotic. From the age of 1 year to 3½ years he prospered, but, thereafter, gradually increasing cyanosis, dyspnea, and fatigue on exertion were noted again. On physical examination, on May 30, 1963, the cardiac findings were as follows: There were moderate cyanosis and clubbing. Vital signs were normal: pulses were full except in the left arm. There was no chest deformity. The apical impulse was displaced to the left and was overactive. There was a faint systolic thrill over the fourth and fifth interspaces at the left sternal border. “P₂” was louder than “A₂”; both sounds were single. There was a grade II-III/VI continuous machinery-like murmur over the entire left upper thorax. In addition, at the lower left sternal border, radiating to the right over the sternum and to the left-toward the apex, there was a rough, medium-pitched, grade II-III/VI systolic murmur with late systolic accentuation, followed by a low-pitched, rumbling grade II/VI mid-diastolic murmur. The auscultatory findings were confirmed by phonocardiography.

Laboratory data were as follows: Hemoglobin, 21 Gm. per cent; hematocrit value, 64 vol. per cent; and red blood cells, 6.53 million per mm.³. The electrocardiogram revealed right axis deviation in the frontal plane and marked right ventricular hypertrophy.

Chest roentgenograms demonstrated no evidence of gross cardiomegaly. The apex of the heart was elevated. Slight left atrial and left ventricular enlargement were present. There was a lack of normal filling-in of the retrosternal space in the lateral view at the usual site of the main pulmonary artery, reflecting the small pulmonary artery in the presence of an atretic valve. In the frontal view (fig. 1), between the border of the right atrium and the diaphragm,
to-right shunt into the right ventricle and moderate desaturation of the peripheral arterial blood. There was marked systolic hypertension, which greatly exceeded the systemic systolic pressure, in the right ventricle. A prominent “a” wave was seen in the right atrial pressure tracing, and there was a moderate degree of widening of the systemic pulse pressure.

Three injections of contrast material for cineangiocardiography were made, one each into the right ventricle, the left ventricle, and the right atrium. Biplane filming at a speed of 48 frames per second was carried out. The films demonstrated the following: (1) The right ventricular chamber was small, oval-shaped, and displaced to the right. It was grossly trabeculated. The right ventricular outflow tract consisted of a thin finger-like projection, approximately 2 cm. long and 4-5 mm. wide, which pointed anteriorly and superiorly and ended blindly at the site of pulmonary atresia. (2) Projecting from the left inferior wall of the right ventricle there was a large, saccular, vermiform dilatation from which arose a single anomalous vessel which was continuous with the right coronary artery. This vessel coursed inferiorly and posteriorly and then curved anteriorly in the right atrioventricular groove to terminate in the right coronary sinus of the aorta. Viewed from the left lateral aspect, the course of the vessel described the shape of an “S.” Contrast material flowed through this vessel from the right ventricle into the aorta during systole and in the opposite direction during diastole (see figures 2 and 3 for details of the anatomy of the vessel and of the dynamics of flow through it). (3) No ventricular septal defect was present. (4) Injection into the right atrium showed a large right-to-left shunt across the atrial septum. (5) Left ventricular injection showed that the left-sided end-to-end subclavian-pulmonary artery anastomosis was patent but small. Filling of the arteries to the right lung was poor, and no definite right or main

Table 1

<table>
<thead>
<tr>
<th>Location</th>
<th>Per cent O2 saturation</th>
<th>Pressures (mm. Hg above mideast)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Superior vena cava</td>
<td>66</td>
<td></td>
</tr>
<tr>
<td>Inferior vena cava</td>
<td>63</td>
<td></td>
</tr>
<tr>
<td>Right atrium (high)</td>
<td>65</td>
<td></td>
</tr>
<tr>
<td>Right atrium (low)</td>
<td>60</td>
<td></td>
</tr>
<tr>
<td>Right ventricle—mid (1)</td>
<td>68</td>
<td>7—mean</td>
</tr>
<tr>
<td>Right ventricle—mid (2)</td>
<td>71</td>
<td>15—“a” wave</td>
</tr>
<tr>
<td>Right femoral artery</td>
<td>74</td>
<td>190/7</td>
</tr>
<tr>
<td>Capacity (calculated)</td>
<td>27.5 Vol. %</td>
<td>119/65 mean—87</td>
</tr>
</tbody>
</table>

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Figure 3
Cineangiocardiogram in the frontal plane with injection of contrast material into the left ventricle. The catheter passed into the left ventricle through a patent foramen ovale and the mitral valve. Contrast material flows into the coronary artery-right ventricular fistula from the aorta during diastole. In frame 201 the fistula is maximally filled just before reversal of flow occurs. The left end-to-end subclavian-pulmonary artery anastomosis is seen in all frames.

Figure 2
Cineangiocardiogram in the frontal plane with injection of contrast material into the right ventricle. The numbers refer to the frames of the film strip which are arranged in sequence vertically. Contrast material flows from the right ventricle through the coronary artery-right ventricular fistula during mid and late systole and is cleared by the flow of nonopaque blood in the reverse direction from the aorta during diastole. (1 = right ventricular outflow tract ending in pulmonary atresia; 2 = veriform sac arising from the right ventricle; RV = right ventricle; and A = aorta. White arrows indicate course of the fistula.)

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pulmonary arteries were visualized. During ventricular diastole, dense opacification of the right coronary artery from the aorta was observed: during systole, the right coronary artery emptied of contrast material as nonopaque blood from the right ventricle entered it. (6) There was no left coronary artery.

The final diagnoses were pulmonary atresia with an intact ventricular septum; anomalous arteriovenous-like communication between the right ventricle and the right coronary artery; patent foramen ovale or atrial septal defect with large right-to-left shunt; patent left end-to-end subclavian-pulmonary artery anastomosis; and absent left coronary artery.

No further surgical treatment was carried out. At his last follow-up examination, in September 1964, he had continued cyanosis, moderate growth retardation, and dyspnea and fatigue on exertion, but he was active and happy. There was no change in the physical signs.

Discussion

Edwards\(^8\) has classified the types of anomalies of the coronary artery system that are encountered clinically into three groups: (1) abnormal numbers, distribution or structure of coronary vessels without communications with the heart chambers or great vessels, (2) isolated coronary artery-heart chamber or great vessel arteriovenous-like communications, and (3) coronary artery-heart chamber or great vessel communications which occur in association with other primary cardiac malformations. Isolated anomalous connections between coronary arteries and any of the cardiac chambers or great vessels are not infrequent.\(^4\) However, those associated with malformations of the right ventricle are rare. Kaufman and Anderson\(^5\) collected 10 cases of the latter type from the literature and added one of their own. Nine of these 11 cases had pulmonary atresia and two had severe stenosis. All had small right ventricular chambers. Eight of the 11 had intact ventricular septa. Four of the anomalous vessels were between the right ventricle and the left coronary artery, three between the right ventricle and the right coronary artery, and four between the right ventricle and both coronary arteries. Only two had single coronary arteries. Davignon et al.\(^6\), in their account of pulmonary atresia with intact ventricular septa, stated that nine of the 13 cases with this anomaly and small right ventricular chambers had "anomalous coronary vessels coursing between the right ventricular chamber and the coronary arteries," but no further details of the pathology were given. In a separate report on the physiologic and angiocardio graphic aspects of one of these cases, Davignon et al.\(^7\) described the presence of a continuous murmur at the upper left sternal border: probably this originated from a patent ductus arteriosus. In this case, "myocardial sinusoids" communicating with an "anomalous coronary vessel" were seen on an angiogram. Anselmi et al.\(^2\) described the oldest case reported up to the present time, a 6-year-old boy with severe pulmonary stenosis and an intact ventricular septum. The anomalous communicating vessel was between the small-sized right ventricular chamber and the anterior descending branch of a single coronary artery which arose from the anterior right sinus of Valsalva. The patient underwent open-heart pulmonary valvotomy and ligation of the anterior descending coronary artery, but died from cardiac standstill within an hour after surgery. Post-mortem injection of the anomalous coronary vessel revealed that its branches arose in a retrograde fashion, forming an acute angle toward the proximal end of the vessel, opposite from the normal branching of arterial vessels.

There are two theories concerning the etiology of the combination of defects under consideration. Edwards\(^8\) believes that the coronary-ventricular communications are secondary to the physiologic effects of the pulmonary atresia with an intact ventricular septum. He has postulated that the high ventricular pressure produced in the blind chamber keeps open communications which are present normally between embryonic sinusoids and coronary arteries. Kaufman and Anderson,\(^6\) Guidici and Becu,\(^8\) and Muir,\(^9\) however, basing their conclusions on the observations and interpretations of coronary vascular system embryogenesis made by Grant,\(^10\) believe that the etiology of the pulmonary atresia and the coronary-ventricular communication is the same. Normally, large inter trabecular spaces, which
CORONARY ARTERY–RIGHT VENTRICULAR FISTULA

...are in free communication with the coronary arteries during early states of cardiac morphogenesis, are obliterated or narrowed to the diameter of capillaries by outward growth of compact myocardium. The failure of expansion of the right ventricular myocardium which accompanies the pulmonary atresia thus, in certain cases, would allow the embryonic connections to persist, without involving pressure relationships as etiologic factors.

Most observers, however, agree on the probable pattern of blood flow through the anomalous vessels once they are established. During systole, right ventricular pressure exceeds that of the aorta and this causes propulsion of blood from the chamber through the anomalous vessel-coronary artery system into the aorta: during diastole, the pressure gradient is reversed and blood flow is in the opposite direction. Anselmi et al.² were the first to report that such a sequence of events had been observed. They stated that the angiocardiogram of their patient showed that "during systole the right ventricle pumped blood through the anomalous coronary artery into the aorta, and, during diastole, blood regurgitated from the aorta into the right ventricle," but they did not illustrate this. The cineangiocardiograms of our patient (figs. 2 and 3) are the first demonstration of bidirectional flow from both the aortic and the right ventricular ends of the fistula.

Comparison of the pattern of blood flow through this coronary-right ventricular fistula with that occurring normally in the major coronary arteries reveals both similarities and differences. Gregg¹¹ and Muller¹² have studied the relation of flow in the left coronary artery to phases of the cardiac cycle. The major portion of flow in this vessel occurs during diastole, although Gregg has observed that from 15 to 60 per cent of the total flow per heart beat may occur during systole under normal circumstances and that this percentage may increase under conditions of stress, such as excitement and exercise. Whether or not the systolic component of the flow enters the intramyocardial portion of the coronary vascular system is unclear. These authors have not observed significant reversal of direction of left coronary flow, except for slight "late systolic backflow" in one dog of Gregg's under conditions of extreme excitement. Also, it seems clear that the magnitude and timing of blood flow in the left coronary artery do not bear a direct relationship to pressures in either the left ventricular cavity or the aorta, although Gregg¹¹ has shown that the contour of the right coronary artery phasic flow curve in the dog parallels the aortic pressure.

By contrast, the direction of blood flow during different phases of the cardiac cycle through the coronary-right ventricular fistula in this patient with pulmonary atresia is apparently directly related to differences in pressure between the right ventricular cavity and the aorta. During mid and late systole, right ventricular pressure exceeds that in the aorta and blood flows through the fistula from the cavity into the aorta. During diastole, right ventricular pressure falls below aortic pressure, and the direction of blood flow is reversed. That the fistula acts thus as a passive conduit is probably due to two main differences between it and normal major coronary arteries: (1) The major length of the vessel lies on the epicardial surface of the myocardium and thus is not subjected to the effects of intramyocardial pressure. There does appear to be some narrowing of the channel at its site of exit from the right ventricle, but this was not marked in degree and was observed not to change its diameter significantly during systole and diastole. (2) The histologic observations of others¹,⁹,¹⁰ show that the sinusoidal portion of the anomalous vessel has neither the caliber nor the structure of an arteriole and therefore cannot offer resistance to flow either because of an intrinsically small cross-sectional area or through contraction of any medial muscular layer. Since there is only a single coronary artery in our case, blood flow into the capillaries of the myocardium must be derived from branches of the anomalous system. Morphologic details of the branches could not be determined from the cineangiocardiogram, and any account of the dynamics.

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of flow through the arteriolar vessels and capillaries would be entirely speculative.

Our patient presented a clinical picture consistent with a diagnosis of severe pulmonary stenosis or atresia with a ventricular septal defect. Indeed, this diagnosis had been made in infancy, and he appears to have benefited from a systemic-pulmonary artery anastomosis performed to palliate this condition. The main clue that the right ventricular-aortic communication was through a coronary artery system was the presence of systolic and diastolic murmurs at the lower left sternal border. The accentuation of these murmurs in late systole and late diastole corresponded to the periods of maximal flow through the anomalous vessel. Selective cineangiocardiology was the method through which the definitive diagnosis was established. The decision against a further surgical approach in this case was made because of the unlikelihood of obtaining adequate relief of the pulmonary atresia and the unpredictable effect on the myocardial blood supply of ligation of the anomalous coronary artery-right ventricular communication.

Summary

A case of an anomalous arteriovenous-like communication between the right ventricle and a single right coronary artery with pulmonary atresia and an intact ventricular septum in a 6-year-old boy is presented. The dynamics of blood flow through the anomalous vessel are demonstrated for the first time in the literature by means of cineangiocardiology. Factors affecting the flow of blood through the vessel, theories of the embryologic etiology of the condition, and clinical features allowing diagnosis during life and determining therapeutic management are discussed.

References

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