Single Coronary Artery with Fistulous Communication

Report of Two Cases

By Raymond H. Murray, M.D.

Congenital coronary artery fistulas communicating with a cardiac chamber, the coronary venous system or the pulmonary artery, are rare anomalies usually discovered at autopsy. In recent years they have been reported with increasing frequency, and the diagnosis has often been established in the living patient. The distinguishing characteristics of this condition are especially important because most of these fistulas can be repaired successfully by generally available surgical techniques.

The discovery of a single coronary artery is in itself remarkable. The coexistence of these two anomalies is extremely rare; only two unequivocal cases have been reported previously. In the past 6 years two examples of this condition have been found among 818 cases evaluated by the Cardiac Study Group of Blodgett Memorial Hospital.

Case Reports

Case 1

S.P., an 11-year-old white girl was admitted to Blodgett Memorial Hospital four times between January 1959 and April 1960 for mild respiratory infections and the evaluation of a heart murmur heard for the first time in September 1958. The product of a normal, full-term pregnancy, she had enjoyed excellent health, had suffered only the usual childhood illnesses, occasional mild respiratory infections, and had grown and developed well. Except for slight fatigue with great exertion she was asymptomatic. There was no history of heart disease in members of her family.

Physical examination was completely normal except for the heart; the blood pressure was 106/65 mm. Hg. The heart size seemed normal and the sounds were of good quality; the pulmonary second sound was finely split and varied normally with respiration. A grade III/VI holosystolic murmur and louder early-and-mid-diastolic murmur were heard best at the third left interspace anteriously, but the murmurs seemed continuous over the lower right sternal border. A “venous hum” was heard in the lower neck in the sitting position.

The electrocardiogram exhibited a very small mean electrical axis in the frontal plane, with slight right axis deviation and RSR’ pattern at V1 to V3R position consistent with a minor right ventricular conduction delay or “diastolic over-loading” of the right ventricle. Routine laboratory data were normal. Fluoroscopy and films of the heart revealed minimal right ventricular enlargement and slight increase of pulsation in the main pulmonary artery.

Right heart catheterization in January 1959 demonstrated normal pressures in the venae cavae, right atrium and ventricle, and pulmonary and brachial arteries, but a definite rise in the oxygen saturation in the right ventricle. Cardiac output was 4.1 and the cardiac index 3.4 liters per minute. Pulmonary blood flow was 5.0 and left-to-right shunt was 0.9 liters per minute. Evans-blue dye curve with injection into right atrium and sampling from an ear oximeter revealed the pattern of a small left-to-right shunt.

The catheterization was repeated in January 1960 and similar normal pressures were found. No distinct evidence of a shunt was found by blood oxygen saturation data although the dye curve again demonstrated evidence of a small shunt. Aortography was carried out with injection of 40 ml. of diatrizoate (Hypaque) just above the aortic valve under pressure. Serial films (six per second) in the posteroanterior projection were interpreted as showing only a single (left) coronary artery arising from the aorta and the right coronary artery arising from this trunk artery. There was strong evidence of a right coronary artery-right ventricular fistula.

On April 28, 1960, at thoracotomy, slight enlargement of the right atrium, ventricle, and pulmonary artery and enormous enlargement (2 to 3 times the expected size) and extensive tortuosity of all visible coronary arterial vessels were demonstrated. A single coronary artery arose from the usual site of the left coronary artery, and its distribution and branching were normal. A “right coronary artery” arose 2 to 2½ cm. from its origin, traversed the AV groove, and had the usual distribution and branching of the right coronary artery, including the posterior descending branch.

From the Cardiac Study Group, Blodgett Memorial Hospital, Grand Rapids, Michigan.
A, upper. Coronary arteries; basal view of the heart. Case 1. A single left coronary artery gives rise to the anterior descending coronary artery (a.d.CA), circumflex coronary artery (c.CA), and the anomalous right coronary artery (r.CA). A fistula connects the right coronary artery and the right ventricle. (PV, pulmonary valve; AV, aortic valve; MV, mitral valve; TV, tricuspid valve). B, lower. Angiogram demonstrating the dilated and tortuous coronary arteries and the dye-filled fistula along the right heart border. Posteroanterior projection.

Midway in its course a small branch arose laterally and immediately plunged into the right ventricular musculature, high on the posterolateral surface (fig. 1). A continuous “thrill” was felt over this branch and the adjacent myocardium. A probe passed into an arteriotomy at this site entered the right ventricle, and a direct connection between the right coronary artery and the right ventricle was demonstrated. No myocardial sinusoids were found. The fistula’s internal diameter was 4 to 6 mm. The remainder of the chambers and the adjacent vascular channels were essentially normal. The fistula was closed and the arteriotomy was repaired, whereupon the thrill disappeared. In order to confirm the absence of associated intracardiac abnormalities the patient was placed on the “cardiopulmonary bypass apparatus,” and the right atrium and ventricle were explored through a right atriotomy; no intracardiac abnormalities were detected.

After an uneventful convalescence the patient was discharged and enjoyed an asymptomatic first postoperative year. She returned in May 1961 for re-evaluation. The electrocardiogram was essentially unchanged. Fluoroscopy and films of the heart showed a slight increase in the general size of the heart. Retrograde aortography again demonstrated a single coronary artery but the arterial branches were significantly smaller and less tortuous, apparently less than half their previous size.

Case 2

G.T., a 6-year-old white girl was admitted to Blodgett Memorial Hospital in April and again in August 1961 for evaluation of a heart murmur heard first in the spring of 1961. In recent months the patient had done well, was uncomplaining and active, but occasionally seemed to tire at play. On physical examination the child seemed normally developed and was normal except for the heart; the blood pressure was 100/60 mm. Hg. The size and configuration of the heart were normal. The pulmonary second sound was of normal loudness and quality, and the “splitting” varied normally with respiration. A grade III/VI holosystolic, rather coarse murmur was heard best in the second left interspace parasternally, with moderate general transmission. A faint (grade I/VI) systolic and diastolic (discontinuous) murmur was heard over the third to fourth right interspace at the sternal border.

The electrocardiogram demonstrated left axis deviation but no ventricular hypertrophy. Fluoroscopy and films of the heart showed slight enlargement of the right ventricle but no evidence of increased pulmonary blood flow. Routine laboratory data were normal.

Right heart catheterization in April 1961 demonstrated normal pressures in the venae cavae, right atrium and ventricle, pulmonary artery, and in the “wedged” position. There was a small but distinct rise in oxygen saturation in the proximal pulmonary artery region. Cardiac output was calculated to be 3.8 and cardiac index 3.9 liters per minute; pulmonary blood flow was 4.9 and a left-to-right shunt was 1.1 liters per minute. Retro-
grade aortography was carried out with 40 ml of sodium diatrizoate (Diatrikon) injected under pressure into the proximal ascending aorta. Serial films in the right anterior oblique projection demonstrated a dilated and tortuous right coronary artery as the only vessel arising from the proximal aorta. This vessel seemed to be normal in its distribution. The left circumflex artery filled from this right coronary artery and the dye was seen to enter either the high (outflow area) right ventricle or pulmonary trunk, apparently from the circumflex artery.

At operation on September 20, 1961, normal cardiovascular structures were found but the right coronary artery arose alone from the aorta at its usual site; it was very tortuous and was estimated as four times normal in diameter. Its distribution was essentially normal and the posterior descending artery arose from the usual site. In this posterior region the right coronary artery was found to be continuous with an equally large and tortuous left circumflex artery which passed circumferentially in the left AV sulcus, gave off a "normal" anterior descending branch and ended in the left, posterior sinus of the pulmonary artery just above the valve (fig. 2). A faint continuous thrill was felt over the site of entry of the fistula into the pulmonary artery; it disappeared after ligation of the fistula. The child has remained well.

Discussion

Structural congenital anomalies of the coronary arteries (variation in size, number, origin, and configuration) are very common and if fundamental physiologic conditions are not disturbed, they are of little clinical importance. Even a single coronary artery can satisfy myocardial circulatory demands and is usually compatible with normal cardiac function and longevity, provided the distribution of the arterial branches is nearly normal. The available literature\textsuperscript{4-7} contains 72 cases of single coronary artery. The general subject has been reviewed in detail by Smith,\textsuperscript{4} who described three general types of the single coronary artery anomaly, delimited by distribution of the branch arteries. Case 1 reported here is an example of type two ("single artery present in the distribution of both coronary arteries"), and case 2 represents an example of type one ("single artery in the distribution of only one coronary artery"). In the great majority of cases included in these two types, the single coronary artery occurs as an isolated cardiac anomaly; although the average age at death of the patients in these groups is younger than expected, the cause of death for the most part is not attributable to the coronary abnormality.\textsuperscript{5} Type three cases ("atypical distribution of a single coronary artery") are usually

\textbf{Figure 2}

A, upper. Coronary arteries; basal view of the heart. Case 2. Abbreviations as in figure 1. A single right coronary artery, continuous with the circumflex coronary artery, gives off the anterior descending branch before entering the pulmonary artery. B, lower. Angiocardioogram demonstrating the dilated and tortuous right coronary artery. Behind the aorta a "puff" of dye can be seen filling the proximal pulmonary artery. Right anterior oblique projection.
associated with other serious cardiac anomalies, and most of these patients die in infancy.

Fistulous communication of a coronary artery with another vessel or cardiac chamber is rarely found and is usually of great clinical importance. The basic anatomic and physiologic characteristics have been presented by Edwards. The salient clinical, radiologic, and hemodynamic features have been well described by Gasul et al., who presented five cases and described and summarized the data from 47 cases previously reported in the literature. However, seven acceptable cases10-16 were published prior to this review and 46 additional cases17-22 have appeared since. In all, 107 cases of coronary arterial fistulas have been found in previous publications; the present two cases bring the total reported cases to 109.

Recent studies28-28 give strong support to the earlier theory of Brooks that the "adult type" of "anomalous origin of the coronary artery from the pulmonary artery" is functionally a coronary artery-pulmonary artery fistula, i.e., blood flows from the "normal" coronary artery into the pulmonary artery by way of the "aberrant" coronary artery. Thus, from the point of view of the physiologist, this can be considered as a "single coronary artery with fistulous communication to the pulmonary artery." Because the intercommunication is usually anatomically indirect, by way of many, small intramyocardial vessels, cases of this type are not usually considered as coronary artery fistulas. Because case 2 of this report and the case described by Ruddock and Stehley do satisfy anatomic as well as physiologic criteria for coronary artery fistulas, they have been so categorized here. A somewhat analogous arrangement exists in the syndrome described by Davignon et al., wherein large intramyocardial sinusoids provide the communication between the right ventricular chamber and a coronary artery. The hemodynamic relationships here are such that blood flows from the right ventricle to the coronary artery and they may be considered ventricular-coronary artery fistulas. These cases have been retained in the category of coronary artery fistula because the sinusoids seem to provide a semidirect communication.

In view of the clinical importance of coronary artery fistulas and the surgical correction available for most of them, their diagnostic characteristics bear summary and emphasis. The size of the heart in these cases is variable, and the degree of enlargement and distortion of shape depends in large part upon the size and location of the shunt as well as the presence of associated cardiac or vascular abnormalities. Diatation of the aorta is notable in many cases, and is especially well seen in the aortogram. Generalized dilatation and tortuosity of the coronary artery bearing the fistula is a constant aortographic finding.

In the cases of single coronary artery with fistulous communication, one of the visible coronary arterial branches were dilated and tortuous, since the entire coronary arterial system is confluent.

The hemodynamic characteristics of the fistulous flow, its attendant stress upon the heart, and the clinical manifestations depend basically upon the size of the fistula and where it enters. A coronary artery in communication with the coronary venous system, right atrium or ventricle, or pulmonary artery creates the physiologic and clinical features of left-to-right shunt, whereas entry into the left atrium or ventricle may mimic ruptured sinus of Valsalva or mitral or aortic insufficiency. Entry into low-pressure chambers such as the coronary sinus or one of the atra creates a large fistulous flow in systole and diastole, hence a systolic and diastolic murmur; the systolic murmur is usually louder, since flow is greater in systole. A similar relationship is expected in coronary artery-pulmonary artery fistulas. These murmurs are usually continuous. When entry is into the right ventricle, the flow may be similar in systole and diastole, or diastolic flow may be the greater, in which case the diastolic murmur will be the louder. When the fistula is in communication with the left ven-

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tricle, flow occurs in diastole only and no systolic murmur of significance is heard. In addition to these primary causes of flow variations, auscultatory findings and other clinical features are affected by the presence of pulmonary hypertension, thrombosis in coronary artery aneurysms, and associated cardiac or vascular defects. The effect of the myocardial contraction upon the flow of a fistula that passes through the right or left ventricular wall is unknown.

The electrocardiographic patterns are nonspecific and are often normal. Evidence of ventricular hypertrophy may help in the localization of the fistula and in assaying the resulting strain upon the heart. The same is generally true of conventional fluoroscopy and x-ray of the heart, but estimation of pulmonary blood flow and the size and contour of the aorta and pulmonary artery may also prove helpful. Associated anomalies or pulmonary hypertension can also complicate these findings. Cardiac catheterization has been carried out in many of the cases published in recent years, including the two cases presented in this report. The data are of great assistance in localizing and estimating the degree of shunt in the coronary venous system, right-sided chambers, and pulmonary artery. With small shunts dye-curve patterns have proved helpful, and special foreign-indicator techniques, such as hydrogen and radioactive Krypton, should prove helpful in localization.

The sine qua non for establishing the diagnosis and location of the fistula is angiography, and selective arteriography is the most useful of these techniques. Direct coronary arteriography ought to provide the most elegant and exact method of all, but its use has not been mentioned in any of the published cases. In addition to exhibition of the dilated and tortuous coronary arterial system and its fistulous connections, arteriography should exclude certain conditions that must be differentiated, such as patent ductus arteriosus, ruptured sinus of Valsalva, aortico-pulmonary window, and ventricular septal defect with aortic insufficiency. On occasion, patent ductus arteriosus has been found to coexist with a coronary artery fistula. Davignon et al. have collected several cases of pulmonary atresia, intact ventricular septum, diminutive right ventricular chamber, and right ventricular-coronary artery communication by way of intramyocardial sinusoids. Selective right ventricular arteriography demonstrated these sinusoids and the abnormal communications.

In addition to these lesions, several other clinical conditions present auscultatory features somewhat similar to those of coronary artery fistulas. These include the "venous hum" heard in normal children, the precordial bruit of pregnancy and lactation, fistulas involving the innominate, phrenic, internal mammary, and pericardial vessels, pulmonary and chest wall fistulas, unruptured sinus of Valsalva aneurysms, hepatoma in the left hepatic lobe, and postvalvular pulmonary stenosis.

Coronary artery fistulas must be considered life-threatening conditions. Some afflicted patients with small fistulas live a normal life-span but the majority die young, usually of congestive heart failure, bacterial endocarditis, pulmonary hypertension, or myocardial ischemia. It seems reasonable that each of these fistulas should be closed surgically unless prohibited by serious contraindications. In most circumstances, if the anomaly is uncomplicated, extracoronoreal circulatory support is unnecessary for the satisfactory repair of the fistula.

The medical literature pertaining to coronary artery aneurysms is a confusing collection of interesting articles. Many cases presented in the earlier reports are certainly not primary aneurysms but have developed due to the stress of a congenital coronary artery fistula. Others are apparently primary coronary artery aneurysms with secondary rupture into a cardiac chamber or adjacent vessel. Often of diagnostic importance is the finding that simple aneurysms of any etiology are usually localized, saccular or fusiform di-
latations, even if multiple, whereas those secondary to coronary arterial fistulas affect the involved coronary artery diffusely, with great tortuosity and dilatation.

Summary

Two cases of single coronary artery with fistulous communication (one to the right ventricle and one to the pulmonary artery) are presented as extremely rare combinations of two rare coronary anomalies. There have been two, previous, similar cases reported in the medical literature.

Acknowledgment

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Addendum

Since this article was submitted for publication another article on this subject has been reported from the Mayo Clinic.27

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History of Use of Salicylates

The history of the salicylate treatment of rheumatic disease is not without interest; salicin was used as a substitute for quinine in France (Blainecourt), and a little later in England (Elliotson), a hundred years ago, but had dropped out of the pharmacopoeia before 1876 when T. J. Maclagan, then of Dundee, first brought it before the medical profession as a remedy for acute rheumatism. It is true that salicylic acid had previously been employed for rheumatic fever by Buss in Basle, Stricker in Berlin, and William Broadbent at St. Mary's Hospital. Maclagan, who began to use salicin in 1874, reached this therapeutic triumph by a teleological process now rather unusual. Believing that acute rheumatism was, like malaria, "miasmatic" in origin, he argued that "a remedy for it would be most hopefully looked for among those plants and trees whose favourite habitat presented conditions analogous to those under which the rheumatic miasm seemed most to prevail." He therefore turned to the willow, the bark of which was known to yield salicin, and later found that the Hottentots had long employed a decoction of the shoots of the willow as a traditional remedy for rheumatic fever. The actual use of sodium salicylate was introduced by Germain See (1817-96) in 1877. The success of treatment by salicin and salicylic acid was shown by Hilton Fagge's analysis of 355 patients thus treated in Guy's Hospital, 1876-80. Before this time the treatment of acute rheumatism had been of various kinds, and its value, at least in the time of the otherwise cheerful Richard Warren (1731-97) may be judged by his epigrammatic answer of "six weeks" when asked what was good for acute rheumatism.—Sir Humphrey Davy Rolleston. The Harveian Oration. Great Britain, Cambridge University Press, 1928 p. 31.
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