Bilateral Coronary Artery–Pulmonary Artery Fistulas

Report of Five Cases and Review of the Literature

DONALD S. BAUM, M.D., HOWARD KLINE, M.D., AND JAMES F. SILVERMAN, M.D.

SUMMARY  Bilateral coronary artery–pulmonary artery fistulas—fistulas that originate from both coronary arteries—are an uncommon subgroup of coronary fistulas and may have a distinct embryologic origin. We present five original cases and review the nine previously reported cases of this anomaly.

CORONARY ARTERY FISTULAS, precapillary communications between a coronary artery and a cardiac chamber or vessel, have been described as the most common hemodynamically significant congenital coronary anomaly.1 In a review of 21 original cases and 342 previous cases, Levin et al.1 reported that coronary fistulas originated slightly more often from the right than from the left coronary artery, but that bilateral fistulas—those that originate from both coronary arteries—accounted for only 5% (19 of 363) of the total. These bilateral fistulas are unique in their tendency to terminate in the pulmonary artery. More than half of the bilateral, but only 17% of the unilateral fistulas, terminated in this manner.

In this report, we review the nine previously described cases of bilateral coronary artery–pulmonary artery fistulas and present five additional cases studied at our institutions between December 1976 and March 1981. The anatomic uniqueness of these fistulas and some considerations in their management are discussed.

Case 1

A 68-year-old lifelong professional athlete developed infrequent episodes of exertional chest pain 6 years before evaluation. A severe episode of chest pain associated with the new onset of atrial flutter prompted hospitalization, during which direct cardioversion restored sinus rhythm. Treadmill testing produced chest pain and 5 mm of ST depression at 1 minute of stage II of the Bruce protocol, and the patient was referred for coronary angiography. Physical examination revealed no cardiac murmur. The ECG showed only a borderline left atrial abnormality and early repolarization. Chest x-ray showed neither cardiomegaly nor pulmonary hypercirculation.

Cardiac catheterization revealed normal right- and left-sided pressures. Oximetry revealed a slight increase in oxygen content between the right ventricle and pulmonary artery (0.6 vol%), corresponding to a calculated 1.2:1 left-to-right shunt. Left ventricular angiography was normal. Significant stenoses with associated calcification were present in the mid-left anterior descending (90%), midcircumflex (80%), and the distal right (90%) coronary arteries. Anomalous vessels originated from both the proximal right and proximal left anterior descending arteries, and drained into the main pulmonary artery via a network of vessels (fig. 1).

Aortocoronary saphenous vein bypass grafting was performed to the mid-left anterior descending, second obtuse marginal, and distal right coronary arteries. At surgery, an extensive network of serpiginous vessels was noted covering the base of the pulmonary artery, and a thrill was present in this area. The pulmonary artery was incised, and a heavy inflow of arterial blood was noted through a single orifice. This was closed with a suture from within the pulmonary artery. The postoperative course was uncomplicated and the patient has remained asymptomatic for 2 years.

Case 2

A 55-year-old female with a 20-year history of a cardiac murmur developed intermittent episodes of substernal pressure that occurred at rest or during exertion. Two months before study, chest pain frequently increased and exertional lightheadedness developed, leading to referral for coronary angiography. Her medical history included insulin-dependent adult-onset diabetes, obesity and hypertension treated with methylpenta (Aldomet). Cardiac examination revealed a grand III/VI continuous murmur at the left second intercostal space, with a grade III/VI midsystolic murmur at the left lower sternal border, the latter murmur increasing dramatically with the Valsalva maneuver. The ECG showed nonspecific ST-T-wave flattening without findings of left ventricular hypertrophy, and the chest x-ray showed left ventricular prominence with normal pulmonary vascularity. The echocardiogram (M-mode and sector) showed borderline concentric left ventricular hypertrophy (left ventricular posterior wall diastolic thickness 1.3 cm) with a localized area of upper septal thickening and systolic anterior motion of the anterior mitral leaflet.

Cardiac catheterization showed a normal early diastolic left ventricular pressure with an elevated end-diastolic pressure of 20 mm Hg due to a prominent a-wave. A 10-mm Hg resting subaortic gradient was present, and increased to 100 mm Hg during the Valsalva maneuver and to 150 mm Hg after isoproterenol, 3 μg by i.v. bolus. The Brockenbrough (post-pregnancy ventricular complex) maneuver was con-
sistent with idiopathic hypertrophic subaortic stenosis. Left ventricular angiography showed a small hyperdynamic left ventricular cavity with systolic cavity obliteration at the apex, and hypertrophic papillary muscles. Selective coronary arteriography (fig. 2) showed a right-dominant coronary circulation without significant stenosis, but with anomalous vessels originating from the proximal right and left anterior descending coronary arteries, and draining into the pulmonary artery through a network of small vessels. In the setting of insulin-dependent diabetes, therapy was begun with the semiselective β, blocker, metoprolol, 50 mg twice daily, which was increased to 100 mg three times daily over the 12 months after discharge, with complete control of chest pain and exertional presyncope. Because of this clinical response, surgical therapy (septal myectomy or fistula closure) was withheld. Antibiotic prophylaxis against infective endocarditis was recommended, and she remained asymptomatic despite resumption of normal physical activity.

Case 3

A 29-year-old female had been noted to have a continuous heart murmur at age 12 years. Right-heart catheterization at that time revealed no oximetric evidence of intracardiac left-to-right shunt. She remained free of symptoms until 1 year before study, when she noted several episodes of mild exertional dyspnea. At the time of study, the patient was asymptomatic, but cardiac catheterization was suggested to evaluate the nature and significance of her cardiac disorder in view of her desire to become pregnant. Physical examination revealed an obese female with a grade II/VI continuous murmur at the left second intercostal space. The chest x-ray, ECG, and M-mode two-dimensional echocardiograms were normal.

Cardiac catheterization showed normal resting hemodynamics. Exercise hemodynamics were not evaluated. No increment in oxygen content was detected between the right ventricle and either pulmonary artery. Selective coronary arteriography (fig. 3) showed anomalous vessels originating from the proximal right and left anterior descending coronary arteries, draining into the pulmonary artery through a network of vessels. Antibiotic prophylaxis against infective endocarditis was recommended, and she remained asymptomatic despite resumption of normal physical activity.

Case 4

A 25-year-old female was noted to have a continuous murmur 1 year before evaluation. She was free of cardiovascular symptoms, but was admitted for diagnostic cardiac catheterization at St. Mary’s Hospital. Chest x-ray and ECG were normal.

Cardiac catheterization showed normal resting left- and right-sided hemodynamics, and the left ventricular angiogram was normal. The supravalvular aortogram showed no evidence of a patent ductus arteriosus or aortic root abnormality, but did show the early appearance of contrast in the pulmonary artery. Selective coronary arteriography (fig. 4) showed a left-dominant coronary circulation without significant stenosis, but with anomalous vessels originating from the proximal right and left anterior descending coronary arteries, draining into the pulmonary artery through a plexus of small vessels. Antibiotic prophylaxis against infective endocarditis was prescribed.

Case 5

A 76-year-old retired salesman was physically active until 6 months before evaluation, at which time he noted chest pain and dyspnea on minimal exertion. Despite therapy with digoxin, propranolol, hydrochlorothiazide and nitroglycerin ointment, his exercise tolerance continued to decline, and symptoms of orthopnea and paroxysmal nocturnal dyspnea developed. His medical history revealed long-standing systemic hypertension, stable moderate renal insufficiency (serum creatinine 4 mg/dl), and stable, anemia (hematocrit 30%). Physical examination revealed a blood pressure of 160/70 mm Hg, slowed carotid upstroke with a normal pulse volume, and bibasilar rales. A grade III/VI mid-systolic murmur was present in the right second interspace and radiated to the carotid arteries, and a grade I/VI decrescendo diastolic blow was present along the left lower sternal edge. The second heart sound was single. An atrial gallop was noted. Chest radiogram revealed moderate
cardiomegaly, pulmonary vascular redistribution, and septal lines. The ECG was consistent with left atrial enlargement and left ventricular hypertrophy. Echo-cardiography showed increased reflectance and decreased systolic motion of the aortic leaflets, left atrial enlargement, and diastolic fluttering of the anterior mitral leaflet, suggesting aortic regurgitation. Cardiac catheterization revealed an elevated mean pulmonary capillary wedge pressure of 36 mm Hg, moderate pulmonary arterial hypertension (systolic 60 mm Hg), and a slightly decreased cardiac index of 2.3 l/min/m². No left-to-right shunt was detected by oximetry. The aortic leaflets were moderately calcified, and a mean systolic pressure gradient of 40 mm Hg was present between the left ventricle and aorta, yielding a calculated aortic valve area of 0.7 cm² (uncorrected for aortic insufficiency). Left ventricular
angiography revealed moderately severe global hypokinesia, with somewhat more severe hypokinesia of the inferior wall. In light of marked congestive heart failure and renal dysfunction, no supravalvular aortogram was performed. Selective coronary arteriography showed significant stenoses in the distal left anterior descending and midcircumflex coronary arteries. The dominant right coronary artery was totally occluded just beyond the conus branch, with opacification of the distal right anterior descending coronary branches via collaterals from the circumflex. A single large anomalous vessel originated from the mid-left anterior descending coronary artery, and a pair of small anomalous vessels originated from the very proximal right coronary artery, draining into the main pulmonary artery (figs. 5A and B).

One week after cardiac catheterization, the patient underwent aortic valve replacement with a #25 porcine heterograft and saphenous vein bypass grafting to the right coronary artery. The large fistula from the left anterior descending coronary artery was evident (fig. 5C) and was ligated directly. The small fistulas from the right coronary were not apparent, and because the right coronary itself was occluded just beyond its ostium, no additional exploration was undertaken. The pulmonary artery was not entered. Despite hemodynamic and renal stability in the immediate postoperative period, the patient did not regain consciousness and developed refractory grand mal seizure. A computed tomographic scan revealed hemispheric cerebral infarction. The patient died on the third postoperative day. Postmortem examination of the heart showed no abnormalities of the aortic valve prosthesis or bypass graft. The fistulas were identified, and their insertion into the pulmonary artery was visualized as a single ostium located just above the left posterior pulmonary valve leaflet (fig. 5D).

**Discussion**

Neither clinical nor hemodynamic characteristics distinguish patients with bilateral coronary artery–pulmonary artery fistulas from those with coronary artery fistulas in general.\(^1\)\(^2\) Of the 14 patients listed in table 1,\(^3\)\(^7\) seven had a continuous heart murmur without significant cardiovascular symptoms. Four additional patients manifested symptoms of fatigue, dyspnea, or atypical chest pain. Three patients (cases 1, 2 and 5) had severe anginal symptoms, but in each case significant additional cardiac pathology was present. Oximetry at the time of catheterization disclosed a significant left-to-right shunt in only one patient (case 13). Selective coronary arteriography was required in all cases for specific diagnosis.

Bilateral coronary artery–pulmonary artery fistulas, however, are a distinct entity. In contrast with unilateral coronary artery fistulas, of which only 17% terminate in the pulmonary artery, 56% (14 of 25) of the reported bilateral fistulas have this termination. Most coronary artery fistulas arise by failure of intramyocardial sinusoids to obliterate, so that bilateral involvement requires independent developmental errors at two separate sites; but fistulas to the pulmonary artery probably arise by supernumerary implantation of the developing coronary arteries into the pulmonary arterial portion of the embryonic truncus arteriosus. The proximity of both coronary arteries to the truncus may thus facilitate bilateral involvement. This defect in embryologic coronary implantation may be related to that which causes anomalous origin of the left or right coronary artery from the pulmonary artery.\(^8\) In both bilateral coronary artery–pulmonary artery fistulas\(^7\) (cases 1 and 5 in this report) and anomalous origin of a coronary artery from the pulmonary artery, the single anomalous coronary ostium typically lies within the left posterior pulmonary sinus (fig. 5D).

While patients with coronary artery fistulas frequently remain asymptomatic, about half of these patients develop cardiovascular symptoms with advancing age.\(^4\)\(^9\) Coronary fistulas have been reported to be the primary cause of coronary artery steal and myocardial ischemia in a few patients and in one animal model.\(^10\) When other cardiac diseases, such as...
coronary atherosclerosis, hypertrophic cardiomyopathy or aortic valve disease, are present, fistulas may potentiate myocardial ischemia. The three patients in our series with severe anginal syndromes (cases 1, 2 and 5) are examples of this potentiation. In some patients, chronic left-to-right shunting due to coronary fistulas may lead to dyspnea and fatigue. Only 3–4% of patients with coronary fistulas may develop bacterial endocarditis.

Because of the relative rarity of coronary anomalies, and the absence of a coordinated registry for patients with these disorders, no prospective information is available on their true incidence, natural history or correct management. Operative fistula closure seems indicated in patients with large left-to-right shunts, those who undergo cardiac surgery for other disorders, and those in whom a clear relationship can be demonstrated between the presence of fistulas and cardiac dysfunction (using techniques such as exercise thallium scintigraphy). In our series, cases 1, 2 and 5 had these indications for surgery, but surgery was deferred in case 2 because of adequate control of symptoms on medical therapy. Six additional cases (cases 7–11 and 14) underwent fistula closure, four with mild and two with no cardiac symptoms. Surgery in asymptomatic patients, who constitute almost half of those with coronary fistulas, can be performed at low risk in patients younger than age 20 years and has been recommended to prevent the development of cardiac symptoms in later life. On the other hand, patients with coronary fistulas may remain hemodynamically stable for as long as 13 years (case 13, table 1), and there is no evidence that early surgery effectively decreases cardiac symptoms as other cardiovascular pathology subsequently develops. The physician must decide whether to manage asymptomatic patients with observation and endocarditis prophylaxis alone or to perform elective fistula ligation.

When surgical correction of bilateral fistulas to the
pulmonary artery is undertaken, the fact that drainage into the pulmonary artery is usually through a single or very small number of outlets suggests that ligation from within the pulmonary artery may avoid the technical difficulty of approaching the complex and friable vascular network itself. 2, 9 This approach was used in case 1, and would have been suitable for case 5, but was not used in the interest of minimizing operative ischemic time.

In summary, while bilateral coronary artery–pulmonary artery fistulas are an uncommon coronary anomaly, our experience with five cases over a 5-year period (four of these cases at a single institution) suggests that additional cases have been studied but not reported. A disproportionately high incidence of fistulas to the pulmonary artery among bilateral coronary fistulas in general suggests that these fistulas are a discrete clinical entity that may have a distinct embryologic etiology. Such fistulas may independently cause cardiac symptoms or may serve to potentiate symptoms deriving from other cardiac pathology. Surgical correction is possible at low operative risk, and may be performed most easily from within the pulmonary artery.

Acknowledgment

The authors express their appreciation to Susan Groechel for her superb secretarial assistance and to Dr. Nelson Burton.

Table 1. Bilateral Coronary Artery–Pulmonary Artery Fistulas

<table>
<thead>
<tr>
<th>Source</th>
<th>Age (years)</th>
<th>Sex</th>
<th>Cardiac symptoms</th>
<th>Murmur</th>
<th>Associated lesion</th>
<th>Oximetric shunt</th>
<th>Surgical fistula closure</th>
</tr>
</thead>
<tbody>
<tr>
<td>1 Current</td>
<td>68</td>
<td>M</td>
<td>Angina pectoris</td>
<td>None</td>
<td>Coronary artery disease</td>
<td>1:2:1</td>
<td>Yes (with CABG)</td>
</tr>
<tr>
<td>2 Current</td>
<td>55</td>
<td>F</td>
<td>Angina pectoris</td>
<td>Continuous</td>
<td>IHSS</td>
<td>Not detected</td>
<td>No</td>
</tr>
<tr>
<td>3 Current</td>
<td>29</td>
<td>F</td>
<td>None*</td>
<td>Continuous</td>
<td>None</td>
<td>Not detected</td>
<td>No</td>
</tr>
<tr>
<td>4 Current</td>
<td>25</td>
<td>F</td>
<td>None</td>
<td>Continuous</td>
<td>None</td>
<td>Not detected</td>
<td>No</td>
</tr>
<tr>
<td>5 Current</td>
<td>76</td>
<td>M</td>
<td>Angina pectoris, dyspnea</td>
<td>Systolic and diastolic</td>
<td>Aortic stenosis, coronary artery disease</td>
<td>Not detected</td>
<td>Yes (with CABG and AVR)</td>
</tr>
<tr>
<td>6 Gobel et al.</td>
<td>17</td>
<td>F</td>
<td>None</td>
<td>Continuous</td>
<td>PDA (prev. Rx)</td>
<td>“Small”</td>
<td>No</td>
</tr>
<tr>
<td>7 Gobel et al.</td>
<td>20</td>
<td>M</td>
<td>None</td>
<td>Continuous</td>
<td>None</td>
<td>Not detected</td>
<td>Yes</td>
</tr>
<tr>
<td>8 Gobel et al.</td>
<td>19</td>
<td>M</td>
<td>Dyspnea, chest pain</td>
<td>Continuous</td>
<td>None</td>
<td>Not detected</td>
<td>Yes</td>
</tr>
<tr>
<td>9 Gobel et al.</td>
<td>53</td>
<td>F</td>
<td>Fatigue, dyspnea</td>
<td>Continuous</td>
<td>None</td>
<td>Not detected</td>
<td>Yes</td>
</tr>
<tr>
<td>10 Oldham et al.</td>
<td>34</td>
<td>F</td>
<td>Not known</td>
<td>Not known</td>
<td>None</td>
<td>Not known</td>
<td>Yes</td>
</tr>
<tr>
<td>11 Liberthson et al.</td>
<td>41</td>
<td>F</td>
<td>Dyspnea</td>
<td>Not known</td>
<td>PDA (prev. Rx)</td>
<td>Not detected</td>
<td>Yes</td>
</tr>
<tr>
<td>12 Huang et al.</td>
<td>56</td>
<td>M</td>
<td>None</td>
<td>Continuous</td>
<td>None</td>
<td>1.12:1</td>
<td>No</td>
</tr>
<tr>
<td>13 Francis et al.</td>
<td>50</td>
<td>M</td>
<td>None</td>
<td>Continuous</td>
<td>None</td>
<td>2.08:1</td>
<td>No (13-yr follow-up)</td>
</tr>
<tr>
<td>14 Ogden</td>
<td>26</td>
<td>M</td>
<td>Dyspnea</td>
<td>Not known</td>
<td>None</td>
<td>Not detected</td>
<td>Yes</td>
</tr>
</tbody>
</table>

*This patient experienced several episodes of mild exertional dyspnea, but she has remained asymptomatic after cardiac catheterization.

†Patient followed from age 37 years to age 50 years.

Abbreviations: CABG = coronary artery bypass grafting; IHSS = idiopathic hypertrophic subaortic stenosis (hypertrophic obstructive cardiomyopathy); PDA = patent ductus arteriosus; AVR = aortic valve replacement.

References