Coronary Arterial-Right Heart Fistulae

Long-Term Observations in Seven Patients


SUMMARY
Long-term follow-up is described of seven patients with fistulae between a coronary artery and the right atrium or right ventricle. Left-to-right shunt flow ranged from minimal to 2.2:1. Of six patients followed 3 to 17 years (average 10) without operation, five demonstrated symptomatic, electrocardiographic, hemodynamic, and angiographic stability. In the sixth patient, a second angiographic study, performed 15 years after the first one, showed the right coronary artery to be occluded proximal to its fistulous communication with the right ventricle, and a left-to-right shunt could no longer be detected. Four of the seven patients underwent operative closure of a fistulous opening into the right atrium, and all four have been restudied postoperatively. Right heart pressures and the degree of dilatation of the involved coronary artery were essentially unchanged following operation. One patient, who had a moderate-sized shunt preoperatively, noted alleviation of her fatigue and demonstrated electrocardiographic improvement. Analysis of flow dynamics did not suggest that the shunt predisposed to shear-induced intimal damage of the dilated feeding coronary artery, but did suggest such changes might occur in the narrow fistulous communication. We conclude that little anatomic and functional change occurs in patients with coronary artery fistulae and small-to-moderate shunts over rather prolonged medical follow-up periods, and that operative closure does not reduce the size of the dilated proximal coronary artery. Since it is unclear whether the abnormality predisposes to premature coronary atherosclerosis, a better understanding of the natural history of the disease is necessary before the precise role of operation in patients with small-to-moderate shunts can be defined.

Additional Indexing Words: Reynolds number
Shear stress

Although coronary arterial fistula is an uncommon cause of left-to-right shunt, the clinical, hemodynamic, angiographic, and surgical features of this abnormality have been described in detail. Little is known, however, of the long-term course of these fistulous communications, either in patients treated nonoperatively or in those undergoing operative repair. This report describes seven patients with small-to-moderate left-to-right shunts through coronary arterial-right heart fistulae. In six of them hemodynamic and angiographic studies were repeated after a period of nonoperative management ranging from 3 to 17 years (average 10). In four patients similar studies were repeated 1–14 years after operative closure of the fistula. In addition to the routine hemodynamic and angiographic studies, flow conditions within the coronary vessels were analyzed in five patients to determine whether mechanical stresses on the vessel wall existed that might predispose to the development of coronary atherosclerosis.

Patients Studied and Methods
In each patient the fistula was supplied by a coronary artery originating normally from the aorta and terminating in either the right atrium or right ventricle. In six patients, the fistula was congenital, and in one (R.I.) it developed following trauma.

All patients initially had right heart catheterization, and one patient also underwent left heart catheterization. Follow-up studies included both right and left heart catheterizations in all patients. Intracardiac shunts were identified and quantified by oximetry and by nitrous oxide or krypton inhalation tests. In all patients, biplane aortography with serial roll films and/or selective coronary cineangiography was performed at each study.
To characterize the coronary flow conditions in these patients, Reynolds number (Re) and the intimal shear stress (τ) were estimated in the normal coronary arteries, in the dilated shunt (feeding) vessels, and in the narrow fistulous communication between the shunt vessel and its receiving chamber in five patients (R.I., F.P., E.C., A.A., F.M.).

Reynolds number: \( Re = \frac{\rho V d}{\mu} = 4\rho Q/\pi \mu d \), where \( \rho \) = blood density (approximately 1.0 g/cm\(^3\)); \( V \) = mean velocity of axial blood flow (cm/sec); \( d \) = lumen diameter (cm); \( Q \) = flow of blood (cm\(^3\)/sec); and \( \mu \) = blood viscosity (approximately 0.04 poise). Q was computed in the shunt vessels from measured values of cardiac output and \( Q_p/Q_s \), where \( Q_s/Q_p \) = pulmonary-to-systemic flow ratio, and was estimated in the nonpathologic coronaries from existing data on normal human coronary flow;\(^{19} \) d was measured at the midpoint of each vessel, with the appropriate scaling factor, from coronary arteriograms.

Shear stress: For fully developed flow in pipes, the relation between shear stress and kinetic flow properties is expressed by an experimentally determined coefficient (f); f is a known function of Re.\(^{20} \) Therefore, when Re is known, \( \tau \) can be estimated, assuming fully developed flow in the arteries under consideration.

\( \tau = f \frac{Q^2}{\pi d^4} \times 1 \), where \( f = \frac{64}{Re} \) for \( Re < 2000 \); and \( f = 0.032/Re^\frac{1}{4} \) for \( Re > 2000 \).

Results

The data in these seven patients are summarized in Table 1.

Symptoms

Five of the seven patients were asymptomatic at the time of initial catheterization, during the interval years, and when restudied. The patients who had symptoms were the two with the largest left-to-right shunts. One (R.I.) has mild dyspnea with severe exertion. Another (P.M.) tired easily, but that symptom disappeared after operation. No patient has had chest pain resembling angina pectoris.

Electrocardiogram

In three patients, electrocardiograms were normal at all times during the study. Two patients who initially had normal tracings developed abnormal electrocardiograms. One (R.I.) developed a right ventricular conduction defect with a normal QRS duration, and the other patient (F.P.), in whom spontaneous total occlusion of her proximal right coronary artery occurred with resultant closure of her fistula, developed left anterior hemiblock. Two patients, each with a moderate-sized left-to-right shunt, had abnormal electrocardiograms at the time of initial catheterization. One (A.A.) had QRS voltage suggesting left ventricular hypertrophy; the electrocardiogram remained unchanged for 5 years, including the 14 months after operation. The other patient (P.M.) had evidence of biventricular hypertrophy between the ages of 2 and 9 years. At age 10 years, 10 months after closure of her fistula, only equivocal electrocardiographic evidence of left ventricular hypertrophy remained.

Cardiac Catheterization

Initial catheterization revealed a small (\( Q_p/Q_s < 1.4 \)) left-to-right shunt in four of the seven patients and a moderate-sized shunt in the other three. The pulmonary arterial and right ventricular end-diastolic pressure (RVEDP) were also normal in each patient except one, in whom the RVEDP was elevated.

Six patients underwent repeat catheterization after periods of nonoperative management ranging from 3½ to 17 years. In one patient (F.P.) the right coronary artery, which was dominant, was found to be occluded proximal to its fistulous communication with the right ventricle, and a left-to-right shunt could no longer be detected by oximetry, by platinum electrode with the patient inhaling hydrogen, or by selective coronary cineangiography. In the other five patients there was no significant change in the calculated pulmonary-to-systemic flow ratios. Pulmonary arterial pressure remained within normal limits in all patients. Right ventricular end-diastolic pressure became mildly elevated in two patients (F.P., B.D.). Left heart catheterization revealed normal left ventricular and aortic pressures in five patients; one patient (R.I.) had mildly elevated left ventricular end-diastolic pressure.

Four patients, all with fistulous openings into the right atrium, underwent operation. The fistulous opening into the right atrium was oversewn in all patients; in addition, the dilated branch supplying the fistula of one patient (G.J.) was oversewn back to its origin from the main right coronary artery. In one of the four patients catheterized postoperatively, a minute residual shunt could be demonstrated by selective coronary cineangiography but not by oximetry or indicator dilution curves. No evidence of residual shunt was found in the other three patients. Right heart pressures were not significantly different after operation.

Radiographic Studies

Only one patient (A.A.) had a cardiothoracic ratio consistently greater than 0.50. In no patient...
## Table 1

<table>
<thead>
<tr>
<th>Pt</th>
<th>Age (yr)</th>
<th>Sex</th>
<th>Symptoms</th>
<th>ECG</th>
<th>Catheterisation</th>
<th>Radiology Chamber enlargement</th>
<th>Angiographic anatomy</th>
</tr>
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<tbody>
<tr>
<td>R.I.*</td>
<td>20</td>
<td>M</td>
<td>Mild DOE</td>
<td>Normal</td>
<td>1.9</td>
<td>9</td>
<td>None</td>
</tr>
<tr>
<td></td>
<td>25</td>
<td>M</td>
<td>Mild DOE</td>
<td>Normal</td>
<td>2.1</td>
<td>7</td>
<td>0.46</td>
</tr>
<tr>
<td></td>
<td>37</td>
<td>M</td>
<td>Mild DOE, conduction defect</td>
<td>Minor RV</td>
<td>1.6</td>
<td>10</td>
<td>0.45</td>
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<tr>
<td>R.R.</td>
<td>3</td>
<td>M</td>
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<td>Normal</td>
<td>1.1</td>
<td>2</td>
<td>0.50</td>
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<tr>
<td></td>
<td>11</td>
<td>M</td>
<td>None</td>
<td>Normal</td>
<td>1.1</td>
<td>6</td>
<td>0.50</td>
</tr>
<tr>
<td>F.P.</td>
<td>29 F</td>
<td>Male</td>
<td>None</td>
<td>Normal</td>
<td>1.1</td>
<td>3</td>
<td>0.41</td>
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<tr>
<td></td>
<td>44</td>
<td>M</td>
<td>None</td>
<td>Left anterior hemiblock</td>
<td>1.0</td>
<td>8</td>
<td>0.41</td>
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<td>B.D.</td>
<td>8 F</td>
<td>M</td>
<td>None</td>
<td>Normal</td>
<td>1.2</td>
<td>3</td>
<td>0.48</td>
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<tr>
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<td>15</td>
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<td>Normal</td>
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<td>10</td>
<td>0.47</td>
</tr>
<tr>
<td></td>
<td>16</td>
<td>M</td>
<td>Closure of fistula opening into right atrium</td>
<td>Normal</td>
<td>1.0</td>
<td>5</td>
<td>0.47</td>
</tr>
<tr>
<td></td>
<td>17</td>
<td>M</td>
<td>None</td>
<td>Normal</td>
<td>1.0</td>
<td>5</td>
<td>0.47</td>
</tr>
<tr>
<td>A.A.</td>
<td>7</td>
<td>M</td>
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<td>LVH LAE</td>
<td>1.4</td>
<td>5</td>
<td>0.59</td>
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<td>10½</td>
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<td>None</td>
<td>LVH</td>
<td>1.5</td>
<td>6</td>
<td>0.54</td>
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<tr>
<td></td>
<td>11</td>
<td>M</td>
<td>Closure of two fistulous openings into right atrium</td>
<td>LVH</td>
<td>1.0</td>
<td>8</td>
<td>0.51</td>
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<tr>
<td></td>
<td>12</td>
<td>M</td>
<td>None</td>
<td>LVH</td>
<td>1.0</td>
<td>8</td>
<td>0.51</td>
</tr>
<tr>
<td>P.M.</td>
<td>2 F</td>
<td>Male</td>
<td>None</td>
<td>BVH</td>
<td>1.8</td>
<td>1.5</td>
<td>0.50</td>
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<tr>
<td></td>
<td>7</td>
<td>M</td>
<td>Mild fatigue</td>
<td>BVH</td>
<td>2.2</td>
<td>4</td>
<td>0.49</td>
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<tr>
<td></td>
<td>9</td>
<td>M</td>
<td>Mild fatigue, RAE</td>
<td>BVH, RAE</td>
<td>1.4</td>
<td>6</td>
<td>0.53</td>
</tr>
<tr>
<td></td>
<td>10</td>
<td>M</td>
<td>Closure of fistulous opening into right atrium and foramen ovale</td>
<td>LVH</td>
<td>&lt;1.1</td>
<td>3</td>
<td>0.50</td>
</tr>
<tr>
<td>G.J.</td>
<td>30 M</td>
<td>M</td>
<td>None</td>
<td>Normal</td>
<td>1.2</td>
<td>5</td>
<td>0.50</td>
</tr>
<tr>
<td></td>
<td>40</td>
<td>M</td>
<td>Closure of fistulous opening into right atrium; feeding branch oversewn to main RCA</td>
<td>None</td>
<td>Normal</td>
<td>1.0</td>
<td>0</td>
</tr>
<tr>
<td></td>
<td>41</td>
<td>M</td>
<td>None</td>
<td>Normal</td>
<td>1.0</td>
<td>8</td>
<td>0.51</td>
</tr>
</tbody>
</table>

*Traumatic fistula.

Abbreviations: DOE = dyspnea; RVH, LVH, BVH = right, left, and biventricular hypertrophy; RAE; LAE = right and left atrial enlargement; Qp/Qs = pulmonary blood flow/systemic blood flow; RVEDP = right ventricular end-diastolic pressure; CTR = cardiothoracic ratio; RA = right atrium; MPA = main pulmonary artery; LA = left atrium; LV = left ventricle; ++, +++ = mild and moderate enlargement; RCA = right coronary artery; LMCA = left main coronary artery; LCCA = left circumflex coronary artery; LADCA = left anterior descending coronary artery; LPDCA = left posterior descending coronary artery; Ao = aorta.
did the cardiac configuration change significantly during the years of follow-up. On chest radiographs, pulmonary arterial blood flow appears to be normal in all patients, but in four the main pulmonary artery was slightly prominent. In two of them (R.R., P.M.) the right ventricle was also enlarged. One patient (A.A.) had mild left atrial and left ventricular enlargement and also had electrocardiographic evidence of left ventricular hypertrophy. Chamber enlargement and prominence of the main pulmonary artery segment in these patients remained unchanged over the years and did not regress in the interval after operation.

Four fistulae involved the right coronary artery: in two the main vessel, and in two others a proximal branch. Three patients had fistulae involving the left coronary artery: a branch of the left main coronary in one, the left circumflex in another, and the posterior descending branch of the left circumflex in the third patient. Five fistulous openings were into the right atrium, and two were into the right ventricle.

Repeat angiographic studies were performed after a period of nonoperative management in six patients. In three of them, the size of the coronary artery contributing to the fistula did not change. In two patients who grew significantly during the interval between studies, the coronary artery enlarged, but in each of them the ratio between the diameter of the dilated coronary artery and the diameter of the descending aorta remained the same (figs. 1, 2A, B, C). One patient (F.P.) developed coronary artery disease, presumably atherosclerotic, with proximal occlusion of the dominant right coronary artery and resultant closure of the fistula (fig. 3). She remains asymptomatic.

In three patients (A.A., B.D., G.J.) the sinus of Valsalva from which the fistulous coronary artery arose was dilated at the initial study and did not change during the interval between studies. Except in one patient (A.A.), who had mild dilatation of the proximal left anterior descending coronary artery, all major coronary arteries not contributing to the fistula were normal at the time of initial study and were similar in appearance when restudied.

All four patients had angiograms after operation. Fourteen months postoperatively, patient A.A.'s dilated coronary artery was identical in size to that seen on two preoperative studies, but the bulbus terminal portion of the vessel had enlarged slightly. Eleven months after operation, patient B.D.'s fistula was shown to be closed, but the proximal right coronary artery and its branch supplying the fistula were dilated to the same degree as preoperatively. Ten months postoperatively, the diameter of patient P.M.'s left circumflex coronary artery was 1–1.5 mm smaller than preoperatively, but it remained greatly dilated, and a very small residual shunt into right atrium could be seen on coronary cineangiography (fig. 2D). In patient G.J. the abnormally dilated branch of the right coronary artery was oversewn at operation and was not visualized postoperatively. The right main coronary artery appeared normal. In patients A.A., B.D., and G.J., the dilated sinus of Valsalva had the same appearance after operation as it did preoperatively.

Table 2 contains the mean values and ranges of \( R_e \) and \( \tau \) calculated for the five patients in whom these analyses were made. The data demonstrated that while \( R_e \) was relatively high in the feeding vessels, indicating a greater tendency to unstable flow, \( \tau \) was nearly normal due to vessel dilatation. Critically high \( \tau \) and \( R_e \) existed only in the narrow fistulous communications.

**Discussion**

Our seven patients with coronary arterial-right heart fistulae are generally similar to patients previously reported.\(^1-10\) Shunt flow was small in four patients, and the other three had moderate-sized shunts. Consequently, most patients were asymptomatic, had normal electrocardiograms initially, and had little or no radiographic evidence of increased pulmonary blood flow, generalized cardiomegaly, or specific chamber enlargement (table 1).

Long-term follow-up of patients with coronary arterial-right heart fistulae is reported here for the

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**Figure 1**

*Patient R.R. Left posterior descending coronary artery-right ventricular fistula. (A, B) Age 3 years. Anteroposterior and lateral films following ascending aortic injection demonstrate a dilated left main, circumflex, posterior circumflex, and posterior descending coronary artery. Fistulous communication to the right ventricle was seen on later films. (C, D) Age 11 years, after 8 years of nonoperative management. Repeat study shows no change in the appearance of the dilated left coronary artery. The left circumflex coronary artery-aorta ratio is unchanged from the previous study.*

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first time. After a period of nonoperative management averaging 10 years, interval studies in five of six patients revealed symptomatic, electrocardiographic, hemodynamic, radiographic, and angiographic stability (table 1). During the 15 years between studies, the sixth patient (F.P.) developed occlusive disease of her right coronary artery proximal to the fistula and thereby abolished the shunt. To our knowledge spontaneous occlusion of a coronary artery fistula has not been described previously. A second lesion producing approximately 60% luminal narrowing was observed even more proximally in the right coronary artery. Despite the occlusion the patient has remained asymptomatic, and her left ventricle has a normal appearance on cineangiography.

In four patients, recatheterization after operation revealed no consistent change in right-sided pressures from preoperative studies (table 1). In the patient who had the largest shunt preoperatively (P.M.), fatigue disappeared after operation, and the electrocardiogram improved. All patients had angiography after operation. In one, the dilated proximal branch supplying the fistula had been oversewn back to its origin from the main right coronary artery, and as expected was not visualized on angiography. In three, the dilated coronary artery that had supplied the fistula had essentially the same appearance postoperatively as it had preoperatively. Thus, eliminating the abnormally high flow and reducing flow velocity does not appear to reduce the size of the coronary artery from which the fistula originated. This may be due to the fact that the vessel continues to be subjected to systemic arterial pressure.

The present study has allowed us to examine the several ways in which coronary arterial fistulae may affect the cardiovascular system adversely and, consequently, to reappraise the indications for operation in this anomaly. The left-to-right shunt through the fistula occasionally is large and, under these circumstances, places a volume load on both ventricles. This remains the firmest indication for closure of the fistula.

Rupture of a coronary artery fistula with cardiac tamponade is a rare occurrence. Closure of the fistulous opening into right atrium or ventricle does not change the tension in the wall of the coronary artery, since the diameter of the vessel and the pressure within it remain the same; thus, the possibility of rupture is unlikely to be influenced by operation. Rupture can be prevented by ligating the dilated artery, but because this procedure may result in ischemia or infarction of myocardium supplied by branches of the artery, it has been abandoned.

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**Table 2**

Flow Conditions of Normal Coronary Arteries, Shunt Vessels, and Fistulae in Patients with Coronary Artery Fistulae

<table>
<thead>
<tr>
<th>Parameter</th>
<th>Normal coronary vessels (n = 11)</th>
<th>Feeding vessels (n = 5)</th>
<th>Narrow fistula (n = 4)</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Mean</td>
<td>Range</td>
<td>Mean</td>
</tr>
<tr>
<td>B_s</td>
<td>117</td>
<td>81-170</td>
<td>804</td>
</tr>
<tr>
<td>r (dynes/cm²)</td>
<td>17.0</td>
<td>4.1-35</td>
<td>18.2</td>
</tr>
<tr>
<td>d (mm)</td>
<td>2.8</td>
<td>2.0-5.1</td>
<td>8.1</td>
</tr>
<tr>
<td>Q (cm³/sec)</td>
<td>62</td>
<td>40-80</td>
<td>1568</td>
</tr>
</tbody>
</table>

*See text for explanation of symbols.*
A coronary artery fistula can potentially “steal” blood from other branches of the artery. For that to occur, however, the proximal portion of the artery would have to offer more resistance to flow than the fistulous opening. Such is rarely the case, as the diameter of the coronary arterial lumen is almost invariably much larger than that of the fistulous opening4.21 (fig. 2C). Although angina pectoris has been described in patients with coronary artery fistulae, none of the patients in the present study had this symptom.

Finally, from the results of the present study, it would appear that small shunts will remain small. This observation, as well as the above considerations, suggests that only patients with at least moderate shunts need be considered candidates for operative repair. However, since spontaneous coronary artery occlusion occurred in one of our patients (who was premenopausal, had no conditions known to predispose to coronary disease, and had no angiographically visible plaques in the left coronary artery), it is possible that one of the most important potential complications of coronary artery fistulae is the development of premature coronary atherosclerosis. In this regard, shear-induced intimal damage has been described by Fry,22,23 and may be a precursor to atheroma formation. This phenomenon is believed to account for the tendency for atherosclerosis to localize around arterial branch orifices.24 However, since shear stress in inversely related to the third power of the vessel diameter, under conditions of laminar flow, the remarkable dilatation of the coronary artery feeding the fistula resulted in relatively low intimal shear stresses in these large feeding vessels. From the data in table 2, stresses of the magnitude known to cause experimental endothelial damage (400-800 dynes/cm²) occurred only in the narrow fistulous communication between the feeding vessel and its receiving chamber. It should be emphasized, however, that our analysis assumes constant flow and does not consider variations due to phasic flow through the coronary vessels. Therefore, this analysis tends to underestimate the extremes of local intimal shear stresses as well as Reynolds number.

With this reservation in mind, our analysis suggests that if shear-induced atherosclerosis does develop in these patients it would be expected to occur in the narrow fistulous communication rather than in the large feeding coronary artery. Thus, coronary artery fistulae may be “self-healing,” and it is possible that the entity known as arteriosclerotic coronary aneurysm25 may represent the end result of atherosclerotic occlusion of the fistulous communication of a coronary artery.

Although Reynolds number was elevated in the feeding vessels, unstable (or turbulent) flow has been shown to persist only when Reynolds number exceeds 2000,26 a value that was surpassed only in the narrow fistulae emptying into the right heart. As indicated above, however, our analysis underestimated the extremes of Reynolds number. Nevertheless, although turbulent flow may predispose to feeding-vessel dilatation26 there is no evidence at the present time that any relation exists between turbulent flow per se and atherosclerosis.

In conclusion, although it is impossible to formulate a definitive plan for the management of all patients with coronary artery fistulae, it would seem reasonable to recommend operation in patients with large shunts because of the resulting flow load on the right and left ventricles. It is not clear, however, whether a threat of premature atherosclerosis exists in patients with small shunts, and, if it does, whether it would lead to serious proximal coronary artery disease or to spontaneous closure of the fistulous communication. It is obvious, therefore, that more must be learned about the natural history of these patients before the role of surgery can be precisely defined.

Acknowledgment

We would like to thank Dr. Kirk L. Peterson, Assistant Professor of Medicine, and Director, Cardiac Catheterization Laboratory, Department of Medicine, University of California, San Diego, for performing the second catheterization of patient F.P.

References


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**Figure 3**

Patient F.P. Right coronary artery-right ventricular fistula. Proximal occlusion of right coronary artery with resultant closure of the fistula. (A, B) Age 29 years. Anteroposterior and lateral films following ascending aortic injection demonstrate a dilated right coronary artery with fistulous communication to the right ventricle. (C) Age 44 years. Cineangiogram (left anterior oblique projection) following selective right coronary artery injection demonstrates complete occlusion of the proximal right coronary artery (heavy arrow). An atherosclerotic plaque producing approximately 60% luminal narrowing is seen just distal to the catheter tip (light arrow). Right ventricular collaterals fill distal to the occlusion. Selective left coronary artery injections demonstrated retrograde filling of the dominant distal right coronary artery from posterior circumflex collaterals. The fistulous communication to the right ventricle was no longer seen.
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