SUMMARY An asymptomatic 14-year-old male was found at cardiac catheterization to have a coronary artery fistula involving a vessel originating from the left main coronary artery and terminating in the right heart. Chest X-ray and electrocardiogram were within normal limits and shunt flow was too small to be detected by oximetry.

MANY CASES of coronary artery fistulae have been reported in the literature although this is a relatively uncommon entity. The majority of these reports describe the clinical and angiographic presentation and the immediate results of operative closure. However, little is known of the natural history of these fistulae and thus the indications for and advisability of surgery. An important and previously unreported point in the spectrum of this anomaly is illustrated in the present case in which spontaneous near closure of a coronary artery fistula over a short time period was demonstrated.

Case Report
A 14-year-old male was first noted to have a heart murmur four years prior to admission. His medical history had

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been unremarkable otherwise and he had always been physically active with no evidence of exercise intolerance or any other cardiovascular symptoms. Because of his murmur he was referred for cardiac catheterization.

On examination he was well developed with normal body habitus. Blood pressure was 110/70. First and second heart sounds were normal. A grade IV/V continuous systolic-diastolic machinery murmur was heard along the lower left sternal border and at the apex associated with a palpable thrill. No other murmurs were present. Physiological third and fourth heart sounds were audible. The electrocardiogram and chest X-ray as well as an echocardiogram were within normal limits.

Coronary angiography showed a large anomalous coronary vessel having essentially a common ostium with the left main coronary artery. Flow of contrast material was rapid in the vessel which was seen to terminate in the right ventricle or lower right atrium near the tricuspid valve (fig. 1). One small branch was seen to originate from its proximal portion. This vessel lacks the tortuosity usually seen in coronary arterial branches giving rise to fistulous communications, likely because of the youth of the patient. The left and right coronary arteries were normal. Right and left heart pressures were all normal. Cardiac index was 3.5 L/min/m². Rapid sequential blood samples were obtained for determination of oxygen content and no left-to-right shunt could be demonstrated by this technique.

We chose to follow the patient with endocarditis prophylaxis alone. Nine months later the murmur remained unchanged. One year following catheterization, the murmur was no longer audible. Electrocardiogram and chest X-ray were again unremarkable. Repeat cardiac catheterization at that time showed markedly reduced size of the fistulous vessel (fig. 2). Blood flow, which was rapid in this vessel on the initial study, was much reduced and associated with very slow emptying. The distal end of the vessel was nearly occluded at its cardiac junction. The coronary arteries remained otherwise unchanged.

**Discussion**

The diagnosis of a coronary artery fistula is usually made by the presence of a continuous murmur in an asymptomatic patient since the shunt flow is usually small. Congestive heart failure occurs at a young age only when there is a very large left-to-right shunt. On the basis of the experience at the Mayo Clinic, Edis et al.¹ felt that the natural history of this anomaly was progressive dilatation and elongation of the affected coronary artery and the cardiac chamber receiving the shunt. Symptoms of congestive heart failure appeared in the fourth decade or later. Other rare complications encountered were myocardial ischemia presumably resulting from a steal syndrome with blood flowing from the myocardial capillary bed to the fistula, endocarditis, rupture of the fistula, and pulmonary hypertension. Rittenhouse et al.² also reported eight cases and, in addition, reviewed the 163 patients from the English literature who had been operated on for this anomaly and reached similar conclusions.

Although it seems clear that symptomatic patients and those with large shunt flows should have operative closure of the fistula, the role of surgery in asymptomatic patients remains unclear. Both Edis et al.¹ and Rittenhouse et al.² expressed the opinion that such fistulae should be closed electively when diagnosed, even in the absence of symptoms.

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**Figure 1.** Left) Selective angiogram of the left coronary artery in the right anterior oblique projection showing a large anomalous vessel originating at the left coronary ostium and terminating in either the right ventricle or the right atrium near the tricuspid valve. Because of selective injection, the left circumflex coronary artery is not seen. Right) Selective injection of the fistulous vessel in the left anterior oblique projection. LAD = left anterior descending coronary artery; SB = first septal branch of the LAD.
Such prophylactic treatment for the admittedly rare complications was justified because of the low operative mortality. However, the operative mortality in the literature review reported by Rittenhouse et al. was 2% with a 3.6% incidence of postoperative myocardial infarction.

Unfortunately, there is little information available as to the natural history of coronary artery fistulae with or without surgery, and the frequency with which patients with this anomaly may reach a normal life span without surgery is presently unknown. Jaffe et al. reported six patients with coronary artery fistulae to the right atrium or right ventricle who were followed for 3½ to 17 years without undergoing corrective surgery during this period. Shunt flows ranged between undetectable levels and 2.2:1. Five of these patients demonstrated symptomatic, electrocardiographic, hemodynamic, and angiographic stability. One patient with a fistula from the right coronary artery to the right ventricle developed, during a 15 year follow-up period, occlusion of the right coronary artery proximal to the fistula, resulting in disappearance of the shunt without change in symptomatic or angiographic state. Operative closure of the fistulous opening into the right atrium in four patients resulted in no demonstrable change in hemodynamics or in dilatation of the coronary artery giving rise to the fistula. One patient who had a shunt of moderate size experienced postoperative symptomatic improvement.

Review of the literature reveals no previously recorded cases of spontaneous closure of a coronary artery fistula other than one patient with disappearance of a murmur but without angiographic documentation of fistula closure. The mechanism responsible for closure of such fistulae is unclear. Jaffe et al. in analyzing flow dynamics in coronary fistulae suggested that shear-induced intimal damage resulting from shunt flow may cause atheromatous changes at the narrow fistulous communication with the heart. Our patient with a coronary artery fistula and a small left-to-right shunt demonstrating near closure over a period of only one year raises further questions as to the necessity of early surgical management. Since the natural history of these fistulae remains undefined, perhaps the asymptomatic patient with a coronary artery fistula should be followed periodically rather than being subjected initially to operative closure.

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Circulation. 1978;57:197-199
doi: 10.1161/01.CIR.57.1.197
Circulation is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231
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Print ISSN: 0009-7322. Online ISSN: 1524-4539

The online version of this article, along with updated information and services, is located on the World Wide Web at:
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