A Case of a Right Coronary Artery to Left Ventricle Fistula
Observed over Twenty Years

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Since the natural history of a coronary artery to left ventricle fistula is not well known, a case of such a fistula in a 36-year-old female patient, followed for 20 years is presented. Two-dimensional echocardiography performed as a follow-up examination revealed enlargement of the right coronary artery and the right Valsalva sinus. Two-dimensional color flow mapping showed the jet flow to the apex from the base of the left ventricle only during the diastolic phase. An aortogram confirmed that enlargement of both the right coronary artery and the right Valsalva sinus had occurred during the 20 years. The shunt flow ratio was about 50%. The Symbas procedure, closing of the coronary artery fistula by arteriotomy directly over the site of the fistula, was performed with the aid of extracorporeal circulation. Our findings indicate that enlargement of the shunted coronary artery and its Valsalva sinus can occur gradually where there is a relatively large shunt flow. Therefore, early surgical treatment is recommended in cases like this one.

Key Words: Coronary artery to left ventricle fistula, Long-term observation, Symbas procedure

CASE REPORT

The patient was a 36-year-old female, who was first admitted to our hospital for further examination of a heart murmur at the age of 16 years. At that time, a grade 3/6 to and fro murmur was audible in the fourth left intercostal space near the sternum. An aortogram revealed the influx of the contrast medium to the left ventricle from the right coronary artery and the diagnosis of a right coronary artery to left ventricle fistula was made. Surgery was not performed because she showed no sign of complications such as myocardial ischemia or heart failure. At the time this case was reported as the seventh case of a coronary artery to left ventricle fistula (3). After she was discharged from our hospital, she visited it as an outpatient.

During her second pregnancy at the age of 25 years, the patient had noted palpitation and shortness of breath on effort. The symptoms continued even after her second delivery and gradually got worse. In 1988, at the age of 36, she was admitted a second time for the evaluation of cardiac function and the possibility of cardiac surgery. On the second admission, her blood pressure was 132/42 mmHg and her pulse rate was 82 per minute and regular. On auscultation neither the third nor fourth sound
A Case of Coronary Artery Fistula

Fig. 1. Chest roentgenograms of the patient with right coronary artery to left ventricle fistula, in 1968 and 1988.

was audible. A grade 3/6 to and fro murmur was heard along the third and fourth left intercostal space near the sternum. The chest radiogram taken during this hospitalization showed no change in cardiac silhouette as compared with that observed 20 years earlier and the cardiothoracic ratio was 54% (Fig. 1). An electrocardiogram revealed a slight leftward shift of the electrical axis and higher voltage of precordial leads as compared with 20 years earlier (Fig. 2). An two-dimensional echocardiogram showed enlargement of the right Valsalva sinus and the dimensions of the left ventricle. Furthermore, a duct-like structure was detected by two-dimensional echoardiography on the posterior wall of the left ventricle along the atrio-ventricular groove. Both the right Valsalva sinus and the left ventricular cavity had enlarged during the 20 years (Fig. 3). Two-dimensional color flow mapping revealed a turbulent flow in the duct-like structure in the short-axis view and the jet flow to the cardiac chamber from the orifice of the fistula in the diastolic phase in the apical two-chamber view (Fig. 4). The aortogram at this hospitalization confirmed the enlargement of the shunted right coronary artery which drained into the posterior wall of the left ventricle from the right Valsalva sinus (Fig. 5). Grade 2/4 aortic regurgitation was also present. The total shunt ratio was almost 50%. A treadmill exercise test was negative. Furthermore, exercise thallium-201 myocardial scintigraphy showed no sign of myocardial ischemia. The Symbas procedure, direct closure of the coronary artery fistula by
coronary arteriotomy, was performed with a cardiopulmonary bypass in the Department of Cardiovascular Surgery of our hospital. The maximum diameter of the right coronary artery was 3.5 cm at its proximal portion and the ostium of the fistula was 2 x 2 cm. Palpation during the operation revealed no sclerotic change in the shunted coronary arterial wall. The coronary angiogram three weeks after the operation revealed no shunt flow but did

Fig. 2. Electrocardiograms of the patient in 1968 and 1988.
DISCUSSION

Coronary artery to left ventricle fistulae are exceedingly rare, and with only 58 cases reported in the literature so far (2-6), and account for about 2 to 3% of all coronary artery fistulae (6, 7). The coronary artery to cardiac chamber fistulae are thought to result from an abnormality in embryonic development of the coronary circulation system. When the intramyocardial trabecular sinusoids, which normally become narrow and persist only as Thebesian vessels in adults, are not obliterated, a fistulous communication persists between the coronary arteries and a cardiac chamber (8, 9).

Until now, surgery was indicated in those cases complicated by myocardial ischemia or recurrent infective endocarditis or those with a large shunt flow.
A Case of Coronary Artery Fistula

Fig. 5. Aortograms showing enlargement of the right Valsalva sinus and the right coronary artery in 1988 (B) as compared with 1968 (A).

(9, 10). However indications for surgery for asymptomatic patients or those with a small shunt flow are less well defined and controversial (11). Furthermore, few reports concerning the long-term prognosis after conservative or operative treatment are available (11-14). Edis et al. (12) suggested enlargement of the shunted coronary artery as the natural history of this congenital anomaly. Furthermore, Liberthson et al. (14) recommended early elective ligation of coronary artery fistulae is indicated for all patients because of the high incidence of late symptoms and complications and the increased morbidity and mortality associated with ligation in older patients. Operative complications such as myocardial infarction and thrombosis of an aneurysmal coronary artery have been reported (15, 16). On the other hand, Jaffe et al. (13) reported 6 cases followed for 3 to 17 years. They concluded that little anatomic or functional change occurs in patients with coronary artery fistulae of 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. They also described a case in which the coronary artery fistula closed naturally as the result of atherosclerotic occlusion during a 15-years of observation. They thought it feasible to recommend the operation for patients with large shunt flows. In our case, the right coronary artery drained into the left ventricle. To our knowledge, no other long-term followed-up case of drainage into the left ventricle has been reported. The twenty-year observation of our case revealed enlargement of both the shunted right coronary artery and the right Valsalva sinus. Our case showed that in a coronary artery to the left ventricle fistula with a relatively large shunt flow like this one, the shunted coronary artery and its Valsalva sinus can dilate gradually over the long-term. Considering that the larger the aneurysmal coronary arteries become, the higher the incidence of rupture of the shunted coronary artery and the appearance of congestive heart failure, we recommend early operative treatment as in our case. The incidence of thrombosis is thought to be low, but some authors have reported thrombosis of the aneurysmal coronary arteries after surgery (15, 16). Unfortunately, a thrombus was detected after closure of the fistula in our case. We believe that not only direct closure of the ostium of the fistula, but also complete ligation of both proximal and distal parts of the dilated coronary artery, in addition to a coronary bypass to the distal branch of the aneurysmal artery, is preferred in a case like ours. However the posterior descending branch of the right coronary artery was not large enough to allow a coronary bypass during the operation in this case.

REFERENCES