

Coronary Artery Fistula: Report of A Case and Analytic Survey of Cases Reported in Japan

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Abstract A case of congenital coronary artery fistula (CAF) was reported. In Japan, 100 cases of CAF were reported until November, 1975. In this report, another 44 cases were added for a review. Fistulas arising from the right coronary arteries were more common than those from the left. The 82 per cent of fistulas entered into the right heart while 14 per cent into the left heart. In the 44 cases added in this study, various surgical approaches were employed in 37 cases; ligation of the coronary artery in 9, interlocking mattress suture behind the coronary artery in 4, closure of the fistula from inside of the cardiac chamber in 11, closure of the fistula through a coronary arteriotomy in 6 and addition of subcoronary-artery mattress sutures to Symbas procedure in 3. In the rest of 4 cases, operative methods were obscure. There were no operative deaths in this series.

Key Words: Coronary artery; fistula, Symbas procedure, statistics

Congenital coronary artery fistula (CAF) has been recognized with increasing frequency following the introduction of the selective coronary angiography. In this study, a boy who was successfully treated by Symbas procedure with subcoronary-artery mattress sutures is presented. One hundred cases of CAF reported by Furushima et al¹⁾ in 1976 and 44 cases after this time were subjected for the survey.

Case Report

A 3-year-old boy was admitted to our hospital for the evaluation of a heart murmur, first noticed when he suffered from bronchopneumonia at 6 months of his age. Since then he was totally asymptomatic. Physical

examination revealed a slightly undernourished child with a regular pulse of 120 beats per minute and a blood pressure of 80/40 mmHg. The heart was normal in size and a high-pitched, grade 3/6 to-and-fro murmur was heard best at the 4th intercostal space and left sternal border. The chest roentgenogram revealed normal heart configuration and clear lung fields. The electrocardiogram was normal and a phonocardiogram recorded at the 4th intercostal space and left sternal border demonstrated a to-and-fro murmur without an accentuation of the second pulmonary tone. The right heart catheterization study disclosed marked step-up in oxygen saturation at an apical portion of the right ventricle and normal pressures in the right

cardiac chambers. Left-to-right shunt ratio was 0.16. A retrograde aortogram was shown in Fig. 1, which revealed the large and tortuous left anterior descending coronary artery terminated to a saccular dilatation which drained directly into the right ventricle at the apex. The right coronary artery and the circumflex coronary artery were normal. These studies established the diagnosis of a left coronary artery fistula draining into the right ventricle. The surgery was performed with Symbas procedure. A longitudinal arteriotomy was made on the aneurysmal distal segment of the left anterior descending coronary artery and a fistula of approximately 2 mm in diameter was repaired with over-and-over sutures, followed by a subcoronary-artery mattress suture. Postoperative course was uneventful and he was discharged 21 days after surgery. The

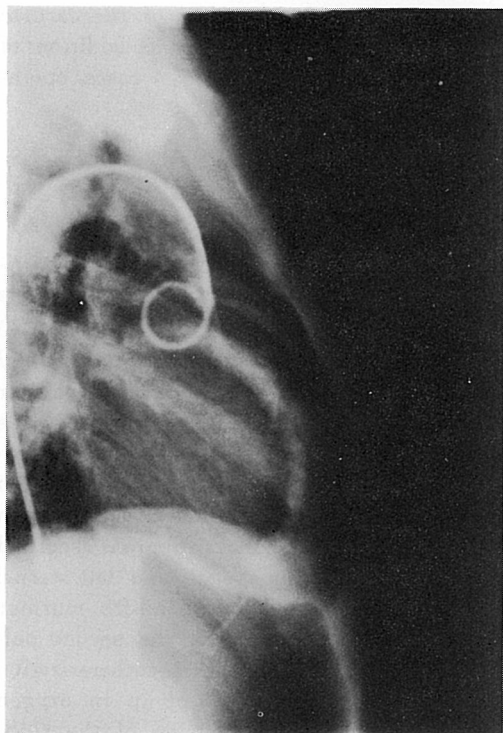


Fig. 1 A lateral view of a retrograde aortogram

patient was reevaluated six months later. The heart murmur was entirely disappeared and angiography revealed that the diameter of the left anterior descending coronary artery is about a half of that before surgery. There was no residual communication.

Analytic Survey of CAF in Japan

Furushima et al¹⁾ surveyed 100 cases of CAF reported in our country until November, 1975. We collected another 44 cases including the present one and reviewed these 144 cases.

Age and sex: Age ranged from 4 months to 72 years. Approximately half of the patients were under 10-year-old. Seventy nine cases were female, 60 were male and five were not described.

Anatomical distribution: The relationship between the artery of origin and the site of drainage is shown in Table 1. Fistulas emptying into the coronary sinus and caval veins were grouped with those into the right atrium. The right coronary artery was more frequently involved (79 cases) than the left (49 cases). In seven patients both coronary arteries were involved and in five the fistulas arose from a single coronary artery. The 82 per cent of fistulas emptied into the right heart; the right ventricle (58 cases) being the most frequent recipient chamber, followed by the right atrium (36 cases) and pulmonary artery (24 cases). Approximately 14 per cent of the fistulas entered into the left heart; the left ventricle (18 cases) and the left atrium (1 case). In 7 patients the recipient chambers were obscure.

Diagnosis: The majority of patients with CAF are asymptomatic. However, reviewed on the present series, approximately 24 per cent of the patients had one to three symptoms of the followings; exertional dyspnea, palpitation, precordial pain, retarded development, repeated respiratory infections and

Table 1 Anatomic Distribution of the Fistula

Recipient chamber	Coronary artery				
	Right	Left	Both	Single	Unclear
Right Atrium	14	22	—	—	—
Right Ventricle	43	12	—	3	—
Pulmonary Artery	5	10	6	2	1
Left Atrium	—	1	—	—	—
Left Ventricle	15	2	1	—	—
Unclear	2	2	—	—	3

heart failure. Heart failure was frequently seen in infants^{1,5,9} and angina-like chest pain usually attacked the patients above 40-year-old^{9,11,14}. Ruptured aneurysm of the coronary artery¹³ and bacterial endocarditis were rarely documented as other consequences.

Clinical manifestation of CAF consisted mainly of a continuous or diastolic murmur over the site of the fistula. Chest roentgenogram, ECG and cardiac catheterization were not specific to CAF. The diagnosis was confirmed by coronary angiography. In the 144 cases, 28 patients were misdiagnosed by physical examination and non-invasive studies for patent ductus arteriosus (8 cases), ventricular septal defect with aortic regurgitation (8 cases), ventricular septal defect (4 cases), aortic regurgitation (4 cases), ruptured aneurysm of Valsalva (2 cases) and others (2 cases). Sixteen out of 28 patients underwent angiographic study, and the diagnosis was corrected. The rest of 10 patients did not receive angiography, and subjected to surgery under misdiagnosis.

Surgical treatment: Among the 44 cases we collected, 37 patients underwent surgery and seven patients were not operated because of minimal shunts and/or patient's refusal. The following surgical approaches have been employed in 37 cases.

1) Ligation of the coronary artery above and below the fistula was made in 9 cases

^{1,5,8,9,18}, with the complication of residual shunt in 2 and myocardial ischemia in one.

2) Interlocking mattress sutures behind the coronary artery (Cooley's procedure) was employed in 4 cases^{9,14}, with the complication of residual shunt in 2 and myocardial ischemia in one.

3) Closure of the fistula from the inside of the recipient cardiac chamber was done in 11 cases^{3,9,10,14,16}, with the side effect of arrhythmia in 4. Involved cardiac chambers were the right atrium in 7, the right ventricle in 3 and the pulmonary artery in one. Arrhythmia seems to happen more commonly in the cases repaired through a right atriotomy.

4) Closure of the fistula through a coronary arteriotomy (Symbas procedure) was made in 5 cases^{4,9,14,15,17}, without complications. Combined method of fistula closure by cardiectomy and arteriotomy (Symbas procedure) was performed in one case.

5) Combination of subcoronary-artery mattress sutures and Symbas procedure was performed in 3 cases⁷, without complications.

In other 4 cases, operative methods were not described. There were no deaths from surgery in this series.

Discussion

Although CAF are relatively rare, this anomaly is being recognized with increasing frequency following wide use of the coronary

arteriography. The influence of the fistula on hemodynamics depends upon not only the distribution of the fistula but also the size of communication. Fistulas entering into the right heart results in a left-to-right shunt and may have mild to moderate pulmonary hypertension. Fistulas connecting to the left heart cause an increase in left ventricular load.

Diagnosis of CAF depends principally upon physical findings and coronary angiography. A characteristic continuous or diastolic murmur heard on the precordial area needs a differentiation of CAF from patent ductus arteriosus, aortic regurgitation, ventricular septal defect and ruptured aneurysm of Valsalva^{1,2,3,14,15}. The meticulous caution should be paid when this anomaly is combined with other cardiac lesions^{1,3,14,15}. A selective coronary angiography discloses the precise anatomical distribution of the fistula and structural changes of the involved artery, even when cardiac catheterization revealed no shunt. An usual consequence of CAF is myocardial ischemia secondary to either decrease in blood supply to the distal coronary branches or coronary artery steal^{1,9,11,14}. These are commonly seen in the patients above 40-year-old. The occurrence of myocardial ischemia is suggested by the occasional anginal symptoms and ECG changes. Congestive heart failure, rupture of aneurysm and bacterial endocarditis are rarely documented as other complications.

Based on the published reports of others, nearly all patients with CAF should be considered candidates for surgery to prevent significant and potentially fatal complications described above. Various surgical approaches have been employed following the site of CAF and surgeon's belief. Among these procedures, we convince that the method of choice is Symbas procedure combined with subcoronary-artery mattress sutures, because there may be numerous minute communications beside the fistula⁷, especially in the

patient with embryologically immature fistula and this combined method prevents recanalization or residual shunts following operation.

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