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# Congenital Coronary Artery Cardiac Chamber Fistula*

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Coronary artery-cardiac chamber fistulas result from an abnormality in embryonic development of the coronary circulation.[4,9] These are the most common congenital anomalies of the coronary circulation. Since the first reported case by Krause[13] in 1865, there are now 200 reported cases of this abnormality in the literature.[16] Recently selective angiography and developments in the methods of intervention on the coronary circulation have re-emphasized the importance of accurate diagnosis and surgical correction of the condition.

In this paper four cases with congenital coronary artery fistula are presented and the symptoms, clinical course, management and complications related to this anomaly are discussed.

The diagnosis of coronary artery fistula was confirmed angiographically, only one patient had surgical correction for the right coronary artery fistula terminating in the right atrium.

## *Clinical Material*

Our patients ranged in age from three to ten years, and the mean age was 7 years. Three out of four, were males (Table I gives the clinical findings and laboratory results).

Two of the patients were asymptomatic (Case 1 and 4, one patient (Case 2) came to the hospital complaining of occasional chest pain. Another patient (Case 3) suffered from fatigue and palpitation. She was

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## TABLE I

### SIGNIFICANT FINDINGS IN 4 CASES

| Case No. | AGE | SEX | Continuous Murmur | Systolic Thrill | Pulmonic 2nd Sound | X-Ray Findings | ECG Qrs Axis | ECG Inter-Pret. | Syst. B. P. | Compli-cation | Coronary Artery | Drainage | Result |
|---|---|---|---|---|---|---|---|---|---|---|---|---|---|
| 1 | 3 Y | M | 3rd-4th LICS Gr III continuous murmur | + | Normal | Right vent. hyp., Pulm congestion | +120 | RVCD | 100/60 | — | Right | Right Vent. | Clinical follow up |
| 2 | 10 Y | M | 3rd-4th LICS Gr IV continuous murmur | + | Normal | Global cardiomegali Pulmonary congestion | + 70 | LVH ST-T chan- | 110/60 | Chest pain | Right | Right vent. | Surgical closure recommended |
| 3 | 10 Y | F | 2nd RICS and apex Gr III Continuous murmur | + | Normal | Left vent. hyp. pulmonary congestion | + 25 | LVH | 115/60 | Infective endocardit CHF fatigue palpitation | Right | Right atr. | Surgical closure performed |
| 4 | 5 Y | M | 2nd RICS Gr II Contiouous murmur | — | Normal | Normal | + 75 | N | 95/50 | Recurrent upper respiratory infection | Left | Right atr. | Clinical follow up |

LICS Left intercostal space RICS Right intercostal space RVCD Right ventricular conduction delay
LVH Lef ventricular hypertrophy CHF Congestive heart failure

admitted to another hospital with the diagnosis of infectious endocarditis and congestive heart failure two months earlier. She was treated successfully. Case 4 had a history of recurrent respiratory infections. All cases had normal systemic arterial pulse pressure. X-rays showed moderate to marked cardiomegaly in three patients. Peripheral pulmonary vascularity was increased in all except one (Case 3). All four patients showed sinus rythm and normal P-R interval. Two patients had left ventricular hypertrophy. One patient also had ST-T wave changes in the left precordial leads, and another patient had right ventricular conduction delay on his ECG. The remaining patient's ECG was normal. Heart murmur was continuous in all patients. It was best heard over the fourth left intercostal space in two patients, and over the second right intercostal space in the other two in whom the murmur was transmitted downwards the right sternal border.

Both right and left heart catheterizations were performed in four patients. The diagnosis of coronary artery-cardiac chamber fistula was confirmed by angiography. Increased oxygen saturation in the right side of the heart was detected in three cases. Right ventricular and pulmonary artery pressures were normal. The pulmonary-to-systemic flow ratio (Qp/qs) was below 1.5 in three cases and higher than 2 in one case.

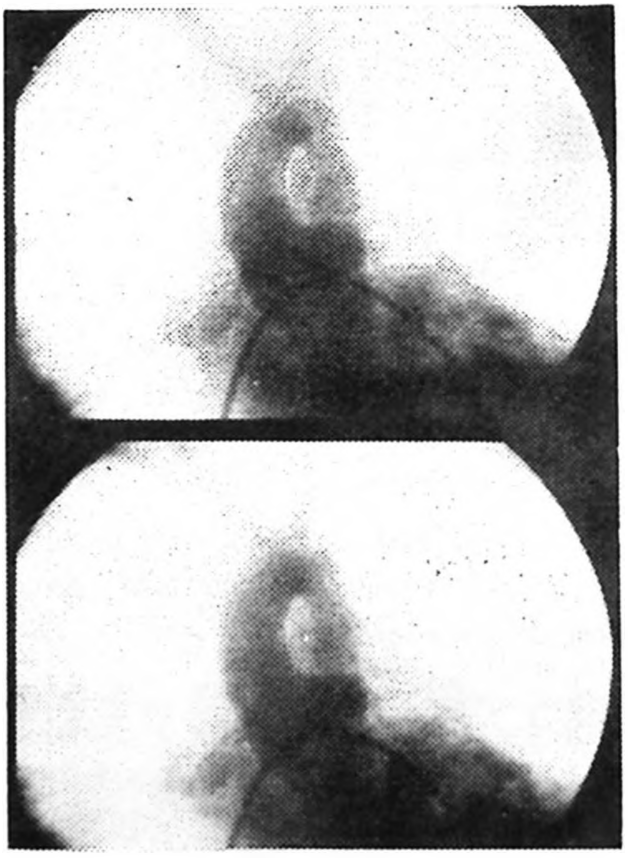

**Figure 1**
a right coronary artery with termination in the right ventricle

The cineangiography revealed the following coronary artery anomalies. The coronary artery fistula originated from the right coronary artery in two patients and terminated in the right ventricle (Figure 1). In one patient the termination was to the right atrium (Figure 2). In the remaining patient a left coronary artery fistula drained into the right atrium. Small ventricular septal defect and subvalvular aortic stenosis were found to be associated lesion in case 2. The other three cases had no other cardio-vascular abnormalities. The patient, whose right coronary artery fistula drained into the right atrium, underwent surgery to have this defect corrected. Closure of the fistula was accomplished by direct liagation at the site of termination. There was no post-operative complications.

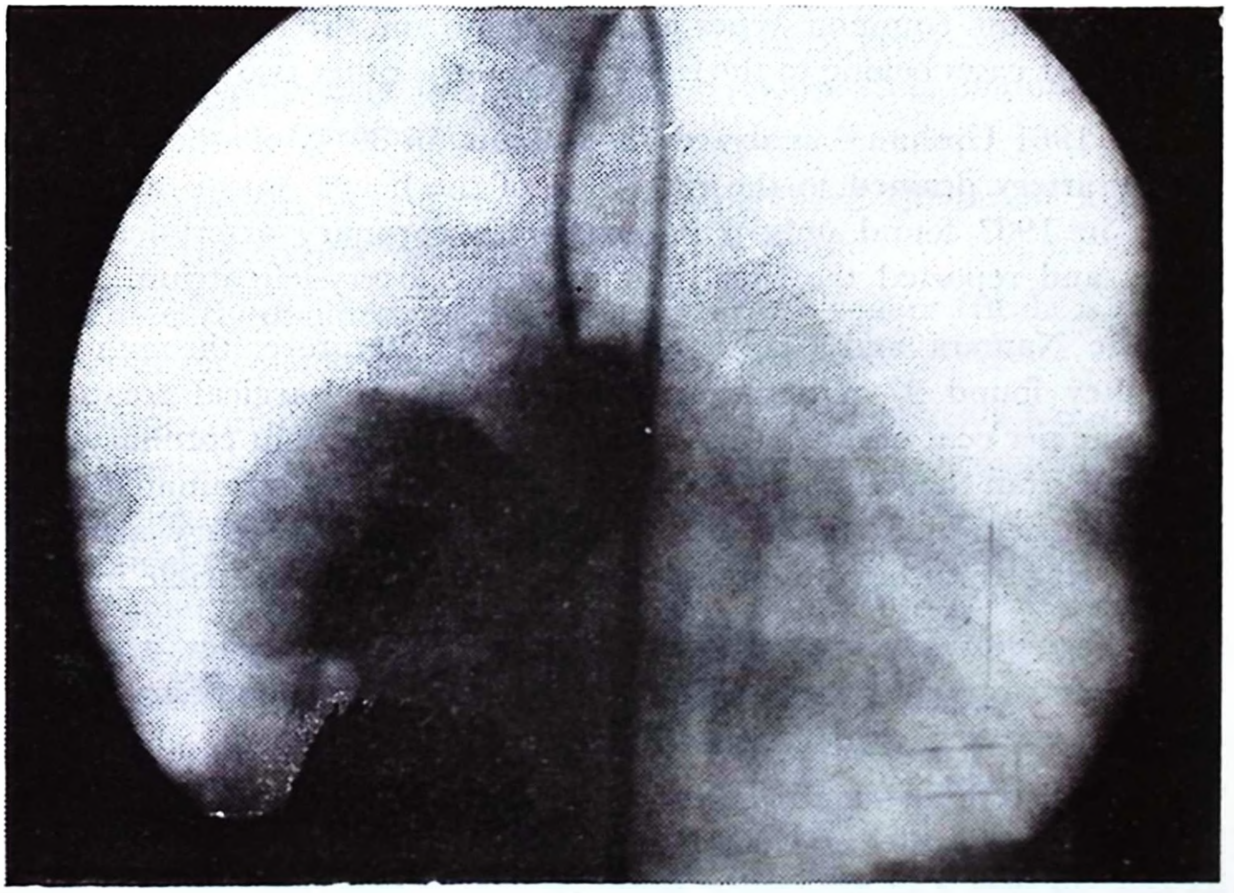

**Figure 2**

a left coronary artery fistula drained into the right atrium

## *Discussion*

Abbott[1] described the condition in detail in 1906 and the first surgical correction of this anomaly was performed by Bjork and Crafoord in 1947.[3] Since then this anomaly has been diagnosed more frequently.[5, 7, 8, 11, 12]

Coronary arterio-venosus fistulas are divided into two major groups on functional grounds.[7] The first group includes anomalies without pulmonary or aortic valvular atresia and the second group comprises coronary anomalies with pulmonary or aortic valvular atresia characterized by retrograde coronary flow.

Depending on the chamber or vessel into which they drain. Coronary arteriovenous fistulas are divided into five types.[18]

*Type I :* Drainage into the right atrium,
*Type II :* Drainage into the right ventricle,
*Type III :* Drainage into the pulmonary artery,
*Type IV :* Drainage into the left atrium,
*Type V :* Drainage into the left ventricle.

The most common types are the second and the first, respectively. Two of our cases belong to the first type and the other two to the second.

In 1961 Upshaw[21] analyzed 73 patients, in 89 % of whom the coronary artery drained to the right side of the heart. Agusti[2] and colleagues in 1967 found only 6 patients with coronary artery-left atrial fistula and reported the first right coronary artery-left atrium fistula.

Mc Namara and Gross[14] reviewed the literature through 1967 and they found 97 patients who had undergone surgical treatment. Fiftynine per cent of the fistulas originated from the right coronary artery, 32 % from the left and 2 % from both. The fistula terminated in the right ventricle in 52 %, right atrium in 24 %, pulmonary artery in 14 %, left atrium or pulmonary vein in 8 % and left ventricle in 2 %.

The majority of patients were asymptomatic. Dyspnea at rest or on exertion was described in 19 % of patients with a coronary artery-cardiac chamber fistula.

A large shunt flow through the fistula may result in congestive heart failure. Daniel and associates[5] reviewed 150 patients and found 21 (14 %) who had congestive heart failure. It was reported that 80 % were over 20 years of age, other authors have also noted that congestive heart failure may develop during infancy.[11, 18]

Subacute bacterial endocarditis has been reported in approximately 10 % of the patients with a coronary artery-cardiac chamber fistula.[14,18,19,20] One of the patients had had subacute bacterial endocarditis. Because of its drastic consequences, both pre and post-operative antibiotic prophylaxis seems advisable. Progressive aneurysmal dilatation of coronary artery fistula has been reported.[17] Disection the fistula

and rupture are rare complications. Only one patient with this complication has been reported.[10] The other symptoms are fatigue, angina or chest pain, palpitation and hemolysis.

The condition is usually first suspected by the discovery of a continous cardiac murmur which is heard along the left and right lower sternal borders. In a fistula to the atrium a systolic accentuation is often noted. Differentiation from patent ductus arteriosus can often be made on the basis of the characteristics as well as the location of the murmur.[15] Fifty-four per cent of the patients were reported to have electrocardiographic abnormalities. Fifty per cent had evidence of enlarged heart of increased pulmonary vascular markings on chest x-ray.

Cardiac catheterization was undertaken in most patients. There was no significant difference in magnitude of the shunt flow in the symptomatic patients. The angiocardiogram was of great value not only in establishing the diagnosis, but also in demonstrating anatomic details of the anomalous vessels.

The aortagrams clearly showed generalized dilatation and tortuosity of the coronary artery bearing the fistula.

The early diagnosis of the coronary artery venous fistula is important both for management and surgical correction.

Despite many reports, management of these patients has still not been clearly established. In those patients who develop congestive heart failure, angina, or recurrent subacute bacterial endocarditis the need for surgical closure of the fistula is clear. The indication for operation in asymptomatic patients or those with small fistulas is not as well defined. It has been recommended that those patients who are asymptomatic should not to be subjected to closure of the fistula.[6, 11] Jaffa and associates described 6 patients, followed for an average of ten years with stable hemodynamics. In one of them the fistula eventually closed spontaneously.

The risk of death or morbidity following closure of a coronary artery-cardiac chamber fistula has been shown to be very low. Several authors have suggested that all fistulas should be closed in order to prevent the complications as well as to relieve the chronic circulatory overload.[7, 9]

*Summary*

Four cases of coronary arteriovenous fistula are reported with a brief review of the literature. Differentiation of this condition from patent ductus arteriosus on the basis of the murmur characteristics and location

was discussed Retrograde aortagraphy was found to be the best diagnostic method. Early correction should be carried out to prevent complications. Both pre and post-operative antibiotic prophylaxis are advisable to prevent subacute bacterial endocarditis.

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