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CONGENITAL LEFT CORONARY ARTERY - RIGHT VENTRICLE FISTULA*

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Coronary artery-cardiac chamber fistulas are the most common congenital malformations of coronary circulation in infancy, childhood and adolescence[1]. Congenital arteriovenous connection results from an abnormality in embryonic development of the coronary circulation, and its incidence appears to be one in 50,000 cases of congenital heart disease, with no sex predilection[1,2].

A rare type of congenital left coronary artery-right ventricle fistula encountered and treated in our clinic is the subject of this paper.

Case Report

A 16-year-old female Caucasian patient was admitted to the Cardiology Department of the Hacettepe Medical Center in March 1981 for evaluation of a cardiac murmur which had been present since her childhood and a chest pain that had started recently. She had no limitations in her physical activities.

On physical examination, arterial blood pressure was 135/70 mmHg, pulse 72/min. A systolo-diastolic cardiac murmur was heard over the left mid-sternal region between the second and fourth intercostal spaces. Other systemic findings were in the normal range. A telecardiogram showed a prominent pulmonary conus with an increase in pulmonary vasculature (Figure 1). An electrocardiogram was within normal range with a sinus rhythm and with normal P-R intervals. Cardiac catheterization was performed, with the following results:

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TABLE I

Results of Cardiac Catheterization

Place of catheter	O_2 Concentration	Pressure (mmHg)
Pulmonary capillary	–	10
Pulmonary artery	75	15/8
Right ventricle	65	15/3
Left ventricle	100	100/10
Aorta	100	100/60
Superior vena cava	54	–
Inferior vena cava	70	–

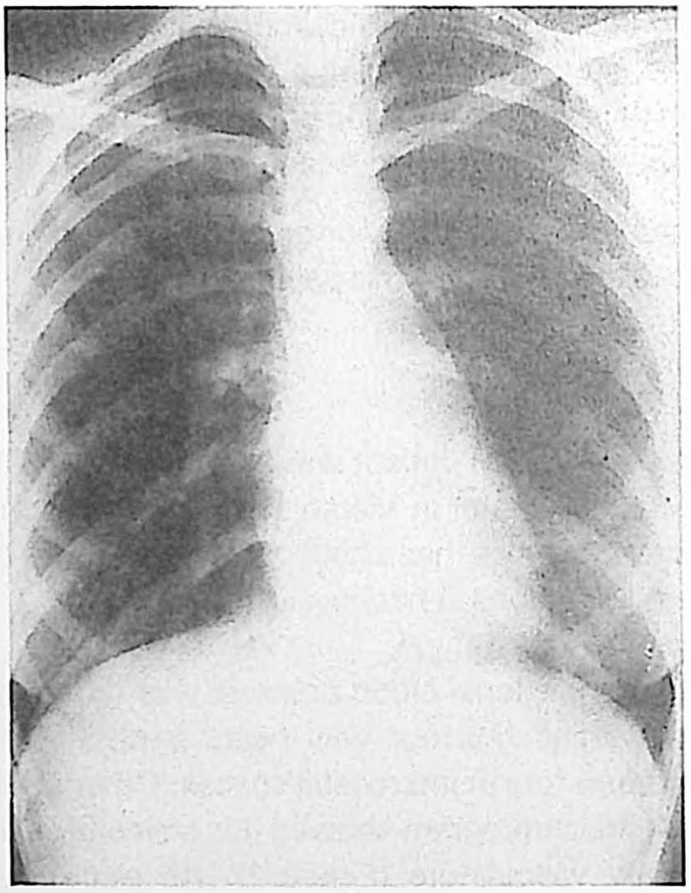

Figure 1

Chest roentgenogram.

Pulmonary systemic flow ratio (Q_p/Q_s) was 1.2. Cineangiography and coronary angiography proved the presence of a left coronary artery-right ventricle fistula with dilatation of the left coronary artery (Figure 2).

The patient underwent open heart surgery November 23, 1981. The left coronary artery was enlarged (about 1.5-2 cm in diameter). An aberrant fistulous tract was

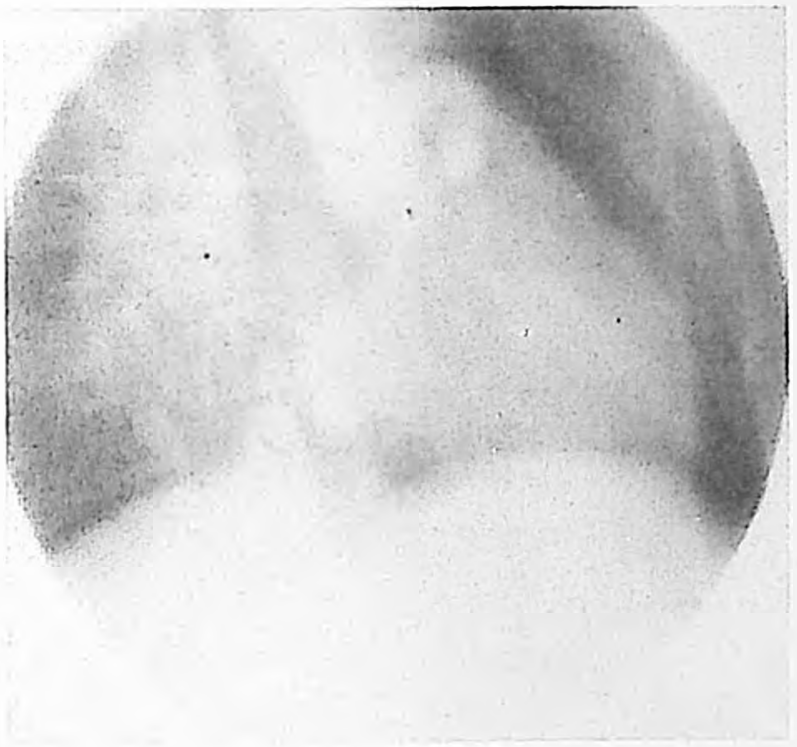

Figure 2

Arteriogram. Contrast material injected at the aortic root travels through the dilated left coronary artery and fistulous tract and fills the right ventricle.

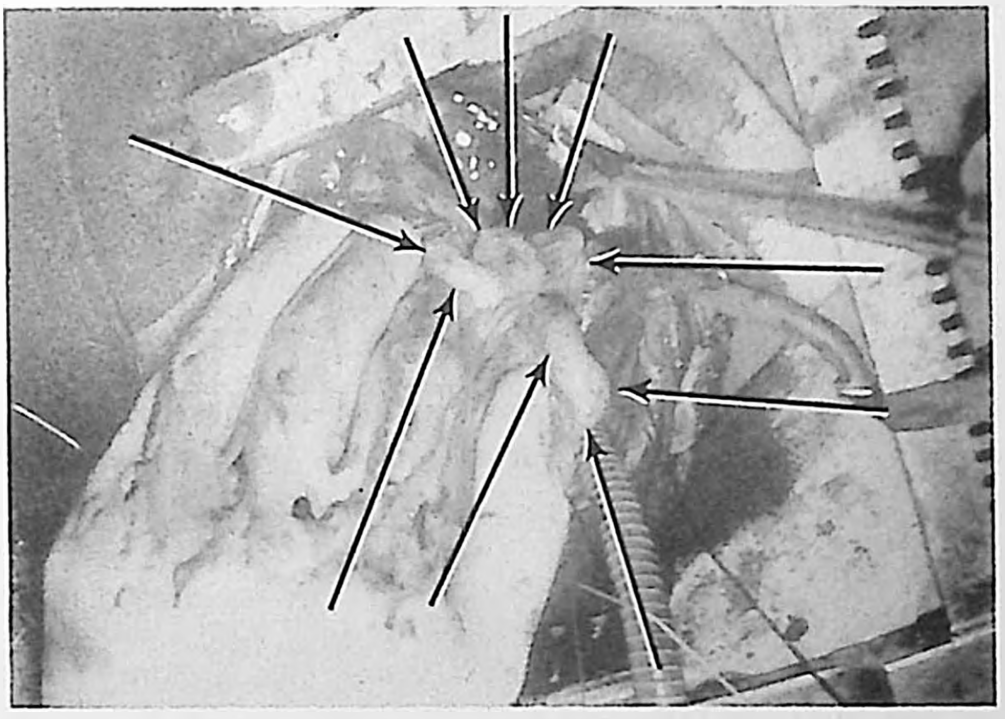

Figure 3

Intraoperative view. The fistulous tract is traveling along the atrioventricular groove (arrows).

present traveling along the atrioventricular groove, finally entering the right ventricle just above the outflow tract with a palpable thrill (Figure 3). Right ventriculotomy was performed under normothermic conditions. The opening of the fistula was noted to be about 6 mm in diameter, just above the outflow tract, and arterial blood was pouring into the right ventricle (Figure 4). The aorta was cross-clamped and the fistulous opening was closed by two 2-0 interrupted sutures reinforced with teflon pledgets, and the sutures were tied outside the right ventricular wall. After closure of the fistula the thrill disappeared. The aortic clamp was released and there was no arterial blood coming into the right ventricle. The right ventricle was closed and the operation completed. The postoperative period was uneventful.

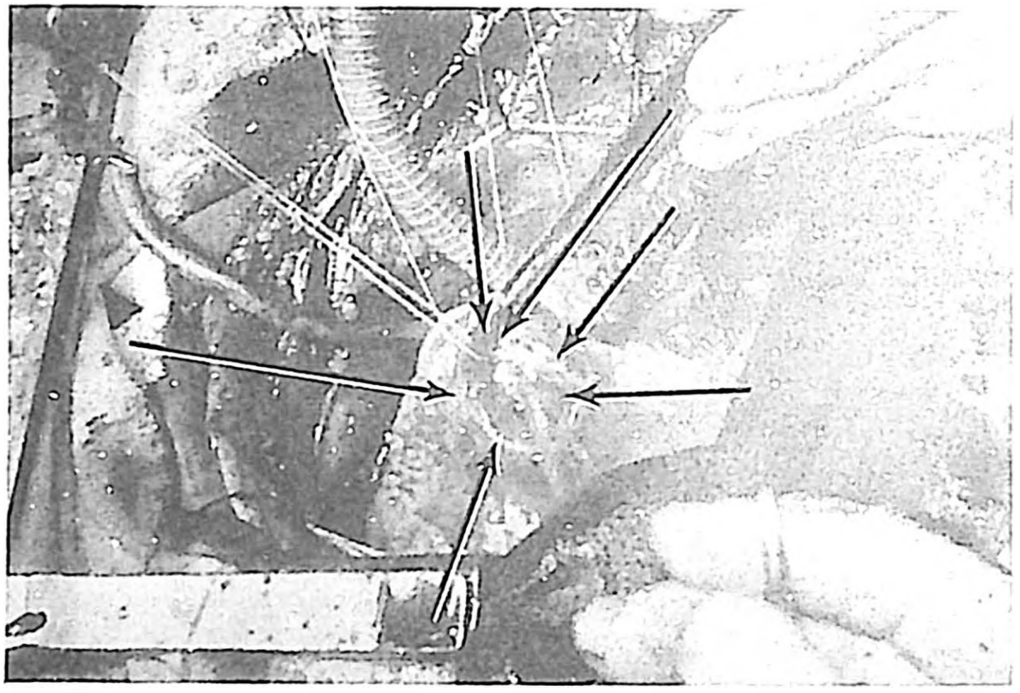

Figure 4

Intraoperative view. The arterial blood of the fistulous tract is pouring into the right ventricle.

Discussion

A congenital coronary artery-cardiac chamber fistula was first reported by Krause in 1865. Abbott described the condition in detail in 1906, and the first surgical correction of this anomaly was performed by Bjork and Crafoord in 1947 (cited by Rittenhouse et al)[3].

Coronary artery fistulas develop from persistence of the intertrabecular sinusoids due to an arrest in the development of the embryonic myocardium[1]. In many cases, as in ours, the fistula originates from a single vessel, frequently tortuous, with a single origin and a single termination[4]. In some cases, however, multiple tortuous vessels may drain into cardiac chambers at multiple sites[5].

Fistulas from the right coronary artery are more common (50-60%) than from the left coronary artery (30-40%) and rarely occur in both coronary systems (2-10%)[6]. The fistula terminates in the right ventricle, right atrium, pulmonary artery, coronary sinus, left atrium, pulmonary veins, left ventricle and/or left persistent superior vena cava[6,7]. Many of these fistulas are small, and their size seems to be proportional to the age of the patient and the resistance of the recipient site[8].

The majority of these patients are asymptomatic[3]. Symptoms occur with increasing frequency in older patients[9]. Symptoms which may occur with a significant number of shunts include congestive heart failure, angina pectoris, bacterial endocarditis, dyspnea, fatigue, frequent upper respiratory tract infections, paroxysmal nocturnal dyspnea and hemoptysis[3,7]. These symptoms probably result from changes in the coronary blood flow by the passage from the normal coronary tree to the dilated coronary arteries[2]. Heart failure is most commonly seen in early infancy and after the fourth decade of life[6]. Atrial fibrillation, frequently seen in older patients, may precipitate heart failure at its onset[3,9]. Aneurysmal dilatation of the fistulous tract is the result of a gradual weakening of the vessel wall, which is secondary to the increased blood flow[3]. Dissection and rupture of the fistula are rare[1,2].

Development of premature atherosclerosis is a potential complication of these fistulas[3]. In atherosclerotic heart disease even a small shunt may exacerbate angina pectoris. Coronary steal syndrome, secondary to the fistula, causes angina pectoris or ischemia[1,3].

A fistula should be suspected during the clinical examination when a continuous cardiac murmur or only the systolic and diastolic components are heard along the left and right sternal borders. In our case the latter type of murmur was auscultated.

This type of murmur can also be detected by auscultation in other malformations, such as aorticopulmonary window, patent ductus arteriosus, ventricular septal defect and aortic regurgitation, rupture of the sinus of Valsalva and absence of the pulmonic valve[7,8]. Chest x-rays are not diagnostic, and an ECG may reveal ischemic changes due to the coronary steal syndrome. The diagnosis of this anomaly is established by cardiac catheterization, cineangiography and selective coronary angiography.

Despite many reports, management of these patients has still not been clearly established. In patients who develop congestive heart failure, angina, bacterial endocarditis, large aneurysm, thrombosis and rupture of the fistula, pulmonary hypertension and progressive radiologic changes, or in symptomatic cases, the need for surgical closure of the fistula is clear[7,9,10,12].

The surgical technique varies according to preoperative and intraoperative findings. It consists of ligation of the fistula as close as possible to its entry to the cardiac

chamber[10-12]. Generally an operation can be performed by simple ligation of the fistula, but in some cases a cardiopulmonary bypass may be necessary[8,10]. Horiuchi et al[11] mentioned the potential danger of injury to the nodal arteries during simple ligation.

Operation for a coronary fistula is usually a safe procedure with a very low morbidity rate[3,10,11]. Myocardial ischemia, infarction and arrhythmias are the most commonly encountered complications[3,12,13]. We believe that this operation should be performed with a cardiopulmonary bypass or at least with a pump on standby for safer closure of the fistula.

Summary

A rare type of congenital fistula between the left coronary artery and the right ventricle, in which the fistula was draining just above the outflow tract, is presented. The patient was operated on utilizing cardiopulmonary bypass, and the fistulous opening into the right ventricle was closed with interrupted ti-cron sutures supported with teflon pledgets. The benefits of using a cardiopulmonary bypass in these operations or at least having it on standby in the operating room are emphasized for safer closure of the fistula when needed.

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