Congenital Right Coronary Artery-Right Atrial Fistula

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SUMMARY

A rare type of congenital fistula between the right coronary artery and the right atrium, where fistula was draining just above the inferior vena caval orifice, is presented. Patient was operated on utilizing cardiopulmonary bypass and the fistulous opening into the right atrium was closed by interrupted ti-cron sutures supported with teflon pledgets. The benefits of using cardiopulmonary bypass in these operations or at least having it stand-by in the operating room are emphasized for safer closure of the fistula when needed.

Additional Indexing Words:

Congenital cardiac fistula Myocardial ischemia Coronary steal syndrome Cardiac failure Bacterial endocarditis

CORONARY artery disease in infancy, childhood and adolescence is an uncommon condition. Anomalous origin and distribution of the coronary arteries and coronary artery-cardiac chamber fistulas are the most common congenital coronary artery malformations in these age groups.

Congenital arteriovenous fistula is the most frequently encountered hemodynamically significant coronary artery anomaly and its incidence appears to be 1 in 50,000 cases of congenital heart disease with no sex predilection.[1)]

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

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Case Report

N.O., 10-year-old Caucasian girl, the last of 7 children of a family, was referred to our hospital complaining of fatigue, palpitation and dyspnea on exertion. Her siblings were in good health. She had been treated in another hospital with the diagnosis of cardiac failure and bacterial endocarditis.

Physical examination revealed BP: 115/60 mmHg and PR: 80/min in sinus rhythm. Her general condition was satisfactory and she was not cyanotic. Auscultation of the heart revealed a systolo-diastolic murmur along the right and left sternal borders, loudest at the 3rd and 4th IC spaces. Liver was palpable 6 cms below the costal margin. Examination of other systems was non-contributory.

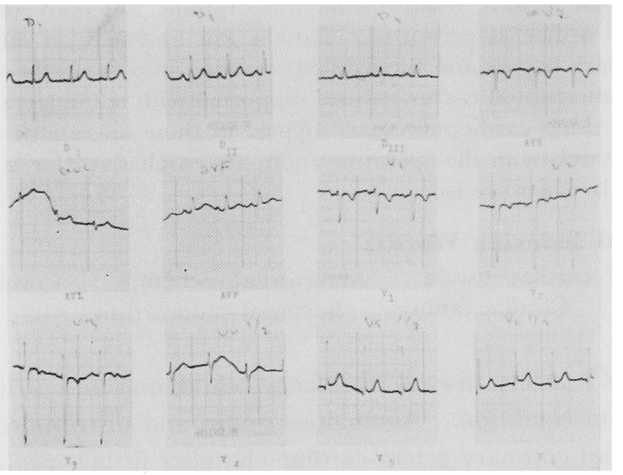

Fig. 1. Electrocardiogram.

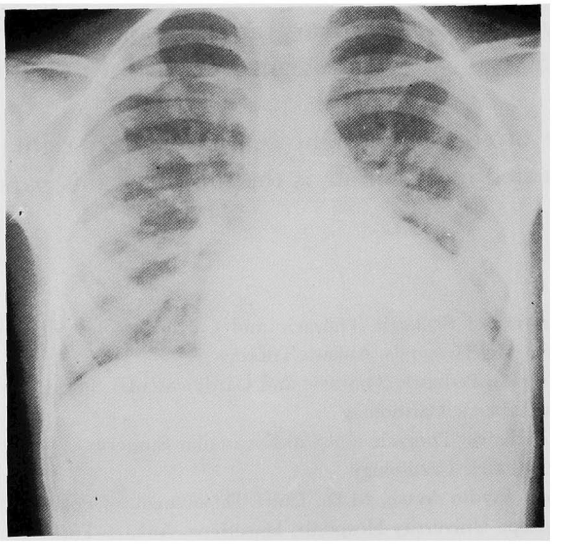

Fig. 2. Chest roentgenogram.

ECG (Fig. 1) displayed slight left ventricular hypertrophy (voltage criteria). Increased pulmonary vascularity and enlarged pulmonary conus were noted in chest X-rays (Fig. 2). Cineangiography and coronary angiography proved the presence of a right coronary artery-right atrial fistula with dilatation of the proximal right coronary artery (Fig. 3).

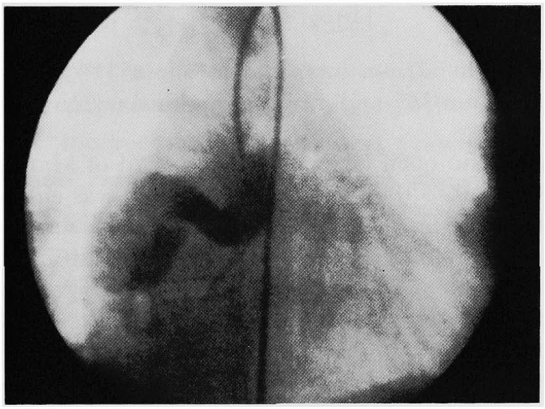

Fig. 3. Preoperative arteriogram. Contrast material injected at the aortic root travels through the dilated right coronary artery and fistulous tract and fills the right atrium.

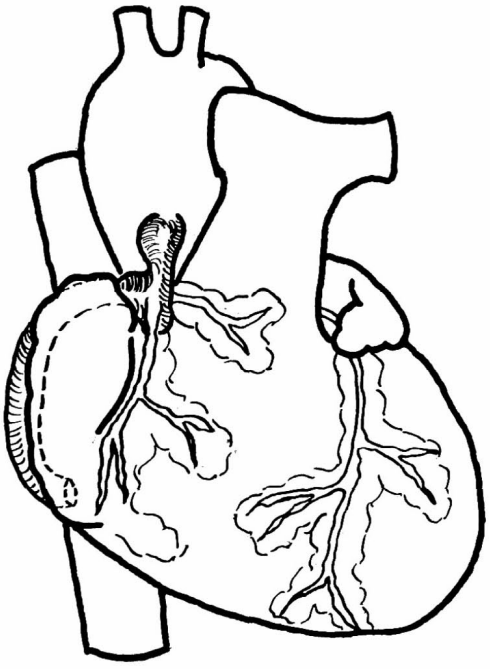

Fig. 4. Schematic representation of the intraoperative view. The fistulous tract is passing behind the superior vena cava and traveling along the interatrial groove, finally ending in the right atrium with a single entry just above the inferior vena caval orifice.

Cardiac catheterization showed:

	Pressures (mmHg)
Pulmonary artery	30/20
Right ventricle	30/0–5
Left ventricle	110/0–10
Aorta	110/80

Oxygenation of the right atrium increased to about 11% of its normal. Left to right shunt was 5.3 l/min/M^2 and ratio of pulmonary/systemic flow was 10/4.7 (=2.1).

She was subjected to open heart surgery on 23rd of March, 1979. The right coronary artery proximal to the right atrial appendix was enlarged (about 1 cm in diameter). An aberrant fistulous tract was present passing behind the superior vena cava and traveling along the interatrial groove, and finally entering into the right atrium just above the inferior vena caval orifice (Fig. 4). Right atrium was opened under normothermic conditions. Opening of the fistula was noted to be about 4 mm in diameter just above the inferior vena caval orifice, and arterial blood was pouring into the right atrium. Aorta was cross-clamped and fistulous opening was closed by two 2–0 interrupted sutures reinforced with teflon pledgets and sutures were tied outside of the right atrial wall and teflon pledgets left inside the right atrium. Aortic clamp was released and there was no arterial blood coming into the right atrium. Right atrium was closed and the operation terminated.

Postoperative course of the patient was uneventful and repeat cineangiography undertaken before the discharge of the patient showed no draining into the right atrium (Fig. 5).

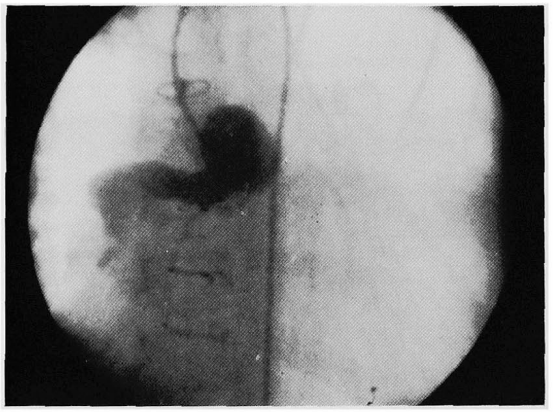

Fig. 5. Postoperative arteriogram. Contrast material given to the aortic root fills the proximal fistulous tract, but does not fill the right atrium. The opaque fistulous silhouette is amputated at the lower level of the right atrium where the fistula's entry is closed.

Discussion

A congenital coronary artery-cardiac chamber fistula was first reported by Krause in 1865. Abbott described this malformation more thoroughly in 1906 and Björk and Crafoord performed the first surgical correction in 1947 (quoted by Rittenhouse et al).[2]

Coronary arteriovenous fistulas develop as the result of the embryonic arrest of obliteration of the intertrabecular sinusoids.[1] Fistulas from the right coronary artery are more common (50–60%) than from the left coronary artery (30–40%), and rarely fistulas from both coronary systems may take place (2–10%).[1)-6)] Major coronary artery may communicate with the right ventricle, right atrium, pulmonary artery, coronary sinus, left atrium, pulmonary veins, coronary veins, left ventricle, and/or left persistent superior vena cava.[1),2),6),7)] Many of these fistulas are small and their size seems to be directly proportional with the age of the patient and inversely proportional with the resistance in the recipient site.[8)] The fistula may communicate with the recipient chamber or vessel via a single opening or multiple openings. Aneurysmal dilatation of the fistulous tract is the result of gradual weakening of the vessel wall secondary to increased blood flow.[2)]

Majority of these patients are asymptomatic.[2)] Symptoms which occur in cases with significant amount of shunts may consist of congestive heart failure, angina pectoris, bacterial endocarditis, dyspnea, fatigue, frequent upper respiratory tract infections, paroxysmal nocturnal dyspnea, and hemoptysis.[2),7)] Wedell and Teloh stated that heart failure tended to occur either in early infancy or beyond 40 years of age (cited by Wenger).[1)] A precordial continuous murmur is heard in these cases. This type of murmur can also be auscultated in other malformations such as aortico-pulmonary window, patent ductus arteriosus, ventricular septal defect with aortic regurgitation, rupture of sinus of Valsalva, absence of the pulmonary valve and other rare types of cardiac malformations like aortico-left ventricular and aortico-right ventricular tunnels.[9)] Certain conditions such as pulmonary hypertension and congestive heart failure can cause a continious murmur to be atypical.[4)]

Development of premature atherosclerosis is a potential complication of coronary artery-cardiac chamber fistulas.[2)]

The diagnosis of this anomaly is established by cardiac catheterization, cinecardioangiography and selective coronary angiography. ECG may reveal ischemic changes due to coronary steal syndrome. The treatment of coronary arteriovenous fistulas is mostly surgical, although indications for operation are not very well defined in the literature for asymptomatic cases and those with small fistulas.[2)] Several surgical groups suggest that all these fistulas

should be closed once they are diagnosed.[1),2),10)] The acceptable indications by the majority of the authors include[1),2),7),10)-12)] 1) congestive heart failure, 2) myocardial ischemia and/or chest pain, 3) bacterial endocarditis, 4) large aneurysms of the involved artery, 5) thrombosis and rupture of the fistula, 6) pulmonary hypertension, 7) significant murmur with progressive radiologic changes, and 8) symptomatic cases.

The surgical technique vary according to the preoperative and intraoperative findings. It mainly consists of ligating the anomalous coronary artery as close as possible to its entry into the cardiac chamber.[10),11)] Although simple ligation of the fistula is possible in most cases, some may require cardiopulmonary bypass for surgical closure.[2),3),6)] Horiuchi et al[13)] called for the potential danger of injuring the nodal arteries during simple ligation of the fistulas draining into the right atrium or superior vena cava. Fistulas with multiple entry sites in the recipient chamber can be repaired by tangential arteriorrhaphy.[3),14)]

Operation for coronary arteriovenous fistula is usually a safe procedure with a very low morbidity and mortality rate.[2),10)] Myocardial ischemia, myocardial infarction and arrhythmias are the most commonly encountered complications after surgery resulting in morbidities and mortalities.[2)] We believe that operation for this malformation should be performed with cardiopulmonary bypass or at least with pump stand-by for a safer closure of the communication and to avoid the above postoperative hazards.

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