Coronary Artery - Cardiac Chamber Fistula

Prinya Sakiyalak, MD
Charun Gherunpongs, MD
Somchai Sriyoschart, MD
Kampol Prachuabmoh, MD

From the Division of Cardiothoracic Surgery, Department of Surgery, Faculty of Medicine and Siriraj Hospital, Mahidol University, Bangkok, Thailand.

Fistulous communication between the coronary artery and cardiac chamber was first reported by Krause[1] in 1865. Abbott[2] described the condition more thoroughly in 1906 and Björk and Crafoord[3] performed the first surgical correction in 1947. Since that time coronary artery cardiac chamber fistulas have been reported with increasing frequency. The advent of aortography and selective coronary angiography has accurately defined the anatomy, thus allowing precise diagnosis[4,5] However, this anomaly occurs infrequently and only a few centers have had extensive experience with operative correction.

CLINICAL MATERIAL

Between 1962 and 1979 seven patients underwent closure of fistulas between the coronary arteries and the cardiac chambers (Table 1). The ages ranged from 7 years to 46 years (average 17 years). There were six females and one male. Six patients had congenital defect whereas only one patient had an acquired type. The major complaint was dyspnea on exertion as appeared in six patients. Two patients (patient no. 3 and no. 4) were admitted with clinical diagnosis of subacute bacterial endocarditis but in only one patient the diagnosis was confirmed by positive hemoculture. The electrocardiogram was abnormal in one patient. Chest roentgenogram revealed mild cardiomegaly with increased pulmonary vasculature in 4 patients. Cardiac catheterization was performed in six patients and oxygen step up was noticed at the atrial level in 3 cases, at ventricular level in one case and at pulmonary artery in one case. In one patient, no oxygen step up was detected. Preoperative diagnoses were correct in only 3 patients. The other 4 patients had diagnoses of mitral stenosis, patent ductus arteriosus and rupture of the aneurysm of sinus of Valsava. The fistulas originated from the right coronary artery in 5 patients and from the left coronary artery in 2 patients. The fistulas ended at the right atrium in 4 cases, at right ventricle in 2 cases and at the pulmonary artery in one case. Closure of the fistulas at their opening with the aid of cardiopulmonary bypass was accomplished in 5 patients where as direct ligation from outside of the cardiac chamber was performed in 2 patients. There was no operative mortality and overall results were excellent. No evidence of recurrence has been detected after periods of 1-17 year follow up. (Table 2)

DISCUSSION

Coronary artery cardiac chamber fistula is a rare anomaly. Upshaw[6] reviewed the English literatures up to 1962 and found 58 patients with this anomaly. Rittenhouse[7] performed a collective review from 65 English literatures up to 1974 and found 163 patients underwent surgical treatment of coronary artery-cardiac chamber fistula. Among 2,000 cases of congental heart disease which were operated upon at Siriraj Hospital during the same period of time, there were only 7 cases of such anomaly and the first case in this series was previously reported by Kitiyakara et al[8] in 1965. Six patients had congenital anomaly where as in only one patient the fistula occurred after a stabbed wound of the heart. The congenital anomaly results from an abnormality in embryonic development of the coronary circulation when the intramyocardial trabecular sinusoids fail to be obliterated. Usually

Table 1 Preoperative clinical status of seven patients with coronary artery-cardiac chamber fistula

No	Age & Sex	Symptom	ECG	Chest Roentgenogram	Catheterization	Diagnosis Preoperative
1.	17 F	Dyspnea	Normal	Normal	No catheterization	Mitral stenosis
2.	9 M	Asymptomatic	Normal	Normal	O_2 step up at PA	PDA
3.	15 F	Dyspnea + fever	Normal	prominent pulmonary conus + RPA	O_2 step up at RA	Rupture aneurysm sinus of Valsava
4.	7 F	Dyspnea + fever	Normal	mild cardiomegaly	O_2 step up at RV	Coronary A-V fistula
5.	7 F	Dyspnea	LVH	mild cardiomegaly	O_2 step up at RA	Rupture aneurysm sinus of Valsava
6.	46 F	Dyspnea	Normal	mild cardiomegaly	No step up	Coronary AV fistula
7.	19 F	Dyspnea	Normal	Normal	O_2 step up at RA	Coronary AV fistula

Table 2 Types of fistula and operative procedures

No.	Type	Fistula	Operation	Cardiopulmonary bypass	Result
1.	Congenital	RCA-RA	Closure	Yes	Good
2.	Congenital	RCA-RV	Closure	Yes	Good
3.	Congenital	RCA-RA	Closure	Yes	Good
4.	Congenital	LCA-RV	Ligation	No	Good
5.	Congenital	RCA-RA	Closure	Yes	Good
6.	Congenital	LCA-RA	Ligation	No	Good
7.	Acquired	RCA-RA	Closure	Yes	Good

there are no associated anomilies, but atresia of the semilunar valve or patent ductus arteriosus may be seen occasionally. In Upshow's series[6], the right coronary artery was involved in 30 patients (53%), the left coronary artery in 18 patients (33%) and both coronary arteries in 10 patients (14%). In Rittenhouse's[7] review, the right was involved in 89 patients (54%), the left in 66 patients (41%) and both coronaries in 8 patients (5%). Entrance into the chamber of the right side of the heart is more common in both eries as in our experience.

There are several hemodynamic consequences and adjustments to a coronary artery-cardiac chamber fistula characterized by the size of the communication and the chamber of termination. Fistulas terminating in the pulmonary artery overload the pulmonary circulation and left ventricle as a result of left to right shunt. Those emptying onto the right atrium or right ventricle cause a volume overload on the right ventricle as well. Although the left to right shunt through a coronary artery-cardiac chamber fistula may be large, a marked increase in pulmonary vascular resistance with reversal of flow from right to left has not been reported.[7] Pulmonary hypertension has been described but is quite unusual.[9] Although most of the reports have indicated that majority of patients were asymptomatic, six out of seven patients in our series were admitted to the hospital with chief complaint of having dyspnea on exertion. Subacute bacterial endocarditis has been reported in approximately 10% of patients with coronary artery-cardiac chamber fistula. Other symptoms which have been described such as angina, paroxysmal nocturnal dyspnea, congestive heart failure, hemoptysis, edema and palpitation were not found in our patients. The diagnosis is usually suspected from detection of a continuous murmur in an unusual location for a patent ductus arteriosus. However, at times it may be difficult to differentiate coronary artery-cardiac chamber fistula from patent ductus arteriosus, ventricular septal defect with aortic valve incompetence, rupture of the aneurysm of sinus of Valsava, or aortopulmonary window. Gasul[10] points out that if the point of entry is into the atrium, the systolic component is dominant with mid systolic accentuation. If the fistula communicates with the left ventricle, the diastolic murmur is loud. With right ventricular entry, two phases of murmur are approximately equal in intensity. Differentiation from patent ductus arteriosus can often be made on the basis of the murmur characteristics as well as location. Phonocardiography may be a useful means of documenting the type of murmur and area of maximum intensity. In our series, the correct preoperative diagnosis was made only in 3 cases where as rupture of the aneurysm of sinus of Valsava, patent ductus arteriosus and mitral stenosis were among the incorrect diagnosis.

The roentgenographic features of coronary artery-cardiac chamber fistula to the right side of the heart are, in general, characteristic of left to right shunt. Those terminating in left heart chambers may be associated with enlargement of the left side of the heart and some times diminished vascular marking may be noted[9]. In patients with a small shunt the chest roentgenogram may be entirely normal as demonstrated in 3 of our reported cases. Mild cardiomegaly was noted in 3 other cases and prominent pulmonary conus and right pulmonary artery was noticed in one

patient. There was no evidence of increased pulmonary vascular marking in our cases which may indicate small shunt in all.

Although 54% of the patients reviewed by Rittenhouse demonstrated electrocardiographic changes, they were not specific or diagnostic. Many patients with this lesion have normal electrocardiogram as demonstrated in 6 of our reported cases. There was only one patient who had left ventricular hypertrophy in the electrocardiogram. 61% of 130 patients reveiwed by Mc Namara and Gross[11] had evidence of right or left ventricular overload. Sakakibara[12] reported high incidence of atrial fibrillation in patients with a fistula draining into the right atrium. Electrocardiographic changes of myocardial infarction are quite unusual despite the potential for myocardial ischemia from a coronary steal. Only 5 of the 163 patients reviewed by Rittenhouse[7] had evidence of myocardial infarction on their preoperative electrocardiograms.

Cardiac catheterization with selective coronary angiography are undoubtedly the best method for precise diagnosis. Two of our reported cases had the preoperative diagnosis of rupture aneurysm sinus of Valsava because they had only an aortic root injection and selective coronary angiography was not performed. One patient had preoperative diagnosis of patent ductus arteriosus due to an oxygen step up to pulmonary artery without confirmation by an aortography. The other patient who had a diagnosis of mitral stenosis did not have cardiac catheterization. At present, the diagnosis could be made accurately by cardiac catheterization and selective coronary angiography.

INDICATION FOR OPERATION

In patients who develops congestive heart failure, angina or recurrent subacute bacterial endocarditis, the need for surgical closure of the fistula is clear. In asymptomatic patients or those with small fistulas, the indication for operation is less defined. Several authors[7,10] have suggested that all fistulas should be closed once they are diagnosed since the operative mortality and morbidity associated with the operation are very low. There have been some arguments that many patients have lived a normal lifespan with their coronary fustulas detected only on postmortem examination[13]. Besides that, there was a report of a case with spontaneous closer of the fistula[14]. Based on these findings, it has been recommended by some authors[15] that aysmptomatic patients should not be subjected to surgical closure of the fistula.

OPERATIVE TREATMENT

The operative approach in most patients has been direct ligation[16]. In many of the earlier reports the coronary fistula was ligated proximal to its entrance into the cardiac chamber, however high incidence of myocardial ischemia resulted. More recently most authors have advised ligation at the point of entrance to the cardiac chamber, thus avoiding the hazard of myocardial ischemia[13]. Some fistulas must be approached utilizing cardio-pulmonary bypass such as the fistulas which open into the atrium, or right ventricular chamber near the ventricular septum and tricuspid annulus. Kitiyakara et al[8] were among the first group to advocate the use of open heart surgery with aid of inflow occlusion and hypothermia. Mc Namara and Gross[11] recommended using extracorporeal circulation in a particularly difficult location and angiomatous fistulas with multiple communications (usually to the right ventricle). We have used cardio-pulmonary bypass in all cases which the fistulas communicated to the right atriums. Direct ligation at their entrances was used on two other occasions with success.

The overall result following closure of the coronary-artery fistula has been excellent. There was no operative mortality in our report. Follow up study revealed no recurrence of the fistula. The low operative mortality seems to justify operation even in asymptomatic patients to prevent complications which have been described.

CONCLUSION

Seven cases of coronary artery-cardiac chamber fistula operated upon between 1962-1979 were reviewed. This anomaly is rare but the diagnosis should be kept in mind to differentiate from other anomalies which give similar clinical manifestations. Cardiac catheterization with selective coronary angiography are the best methods for confirmation of the diagnosis. The indication for closure of the fistula is clear in symptomatic patient. For those who are asymptomatic, operation should still be considered to prevent possible complications.

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