Coronary Arteriovenous Fistula- Clinical Features, Morphologic Aspects, Diagnostic Criteria, Management Options, and Outcome

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Abstract

Objective: Coronary artery fistula is an unusual coronary anomaly. To the spectrum of coronary cameral communications, we want to add this separate entity of coronary cameral fistula by presenting the clinical features, morphologic aspects, diagnostic criteria, management options, and outcome in various clinical settings for better understanding of this developmentally intriguing, clinically complex, and therapeutically challenging disorder.

Methods: From June 1992 through June 2016, 16 patients were treated for coronary cameral fistula at our institution. Ages ranged from 1 to 53 years. There were 8 male subjects and 8 female subjects. Morphologically, isolated fistulas arise from both the right and left coronary arteries, probably with similar frequency, but terminate much more commonly in the right heart or pulmonary artery than in the left heart. But in our series 76.4% (n=13) of the fistula originated from the left coronary artery and 23.6% (n=4) from the right coronary artery. The termination or drainage to the right ventricle in 47% (n=8), to the right atrium in 29.4% (n=5), to the coronary sinus and left atrium in 5.9% each (n=1 each) and to the pulmonary artery in 11.6% (n=2); In all patients the diagnosis was established by means of 2-dimensional echocardiography and transesophageal echocardiography and confirmed by means of angiography and contrast enhanced tomography scan. Fifteen patients were treated surgically, and 1 patient was treated with coil embolization. Postoperative echocardiograms obtained for all patients before discharge confirmed complete obliteration of the tunnel.

Results: There was no early or late mortality. All patients were discharged in stable condition. During follow-up at 3 months, 1 year, 5 years, 10 years and more than 10 years, all patients were in New York Heart Association class I, and echocardiography showed no residual shunts.

Conclusions: Coronary cameral fistula is a rare congenital anomaly. It can be seen isolated or in association with other cardiac defects. It may arise from one coronary system or involve both coronary systems. Fistulas draining into the coronary sinus are prone to develop congestive cardiac failure. Fistulas draining in Right atrium or coronary sinus are prone to develop atrial fibrillation & its sequel. Cardiac cath and coronary angiography gives definitive diagnosis and planning management and to rule out other cardiac lesions. Computed tomography angiog may replace angiography in the future for noninvasive diagnosis. Surgery is indicated for lesions not amenable to percutaneous closure. Surgical closure can be done with or without cardio pulmonary bypass by ligation, tangential arteriography or closure via cardiac chamber. Most of the fistula can be dealt with in the interventional catheterization laboratory. However, occasionally large fistulas have a difficult origin and may present particular challenges to the interventional catheterization team so that they require surgical management. The location and the size of the fistula dictates technical details. Follow-up reveals excellent functional recovery.

INTRODUCTION

The prevalence of Coronary arteriovenous fistula (CAF) is about 0.1-0.8% based on coronary angiography or echocardiography studies. CAFs originate mostly from the right coronary artery and the left anterior descending artery and have proximal involvement. Most of them drain into the right atrium, right ventricle and pulmonary artery. Few of them drain into the left ventricle or left atrium. To the spectrum of coronary cameral communications, we want to add this separate entity of coronary cameral fistula by presenting the clinical features, morphologic aspects, diagnostic criteria, management options, and outcome in various clinical settings for better understanding of this developmentally intriguing, clinically complex, and therapeutically challenging disorder.

MATERIAL AND METHODS

This is a retrospective and prospective study of 16 patients with CAF who were treated between November 1992 through July 2016, at our institution. Ages ranged from 1 to 53 years (mean, 19.3 years), with a male/female ratio of 8:8. All patients were symptomatic. Clinical presentation included one or more of the following complaints. All patients had shortness of breath, of whom 15 were in New York Heart Association (NYHA) class I and 1 was in NYHA class IV. One patient presented with palpitation. Recurrent respiratory tract infection was present in 4 patients, and angina of NYHA class II was present in 1 patient. Physical
examination revealed a continuous murmur at the right or left parasternal border in the third, fourth, and fifth intercostal spaces in all 16 patients. Initial chest radiography showed cardiomegaly in all patients, with cardio thoracic ratios ranging from 55% to 80% and increased pulmonary vascularity. Electrocardiography showed sinus rhythm, normal axis, and right ventricular hypertrophy, with strain pattern in 14 patients, two patients were in atrial fibrillation. Two dimensional (2D) echocardiography and Doppler scanning showed a dilated coronary with continuous Doppler signals in the draining chamber (Table 1, Figure 1). Cardiac catheterization and coronary angiography were performed in all patients. Cardiac catheterization showed a step up in draining chamber ranging from 77% to 92.9% and left-to-right shunt ranging from 1.2:1 to 4.5:1 L/min/m², with a normal pulmonary vascular resistance index ranging from 0.2 to 2.5 Wood units except in one patient with fistula to left atrium. Coronary angiography showed, single fistula in 15 patients and to 2.5 Wood units except in one patient with fistula to left atrium.

Coronary angiography showed, single fistula in 15 patients and multiple fistula in 1 patient. Fistula was originating from the left anterior descending coronary artery (LAD) in 5 patients, Left circumflex coronary artery (LCX) in 8 patients, and right coronary artery (RCA) in 4 patients. The site of termination of fistula was in right ventricle (RV) in 8 patients, right atrium (RA) in 5 patients, pulmonary artery (PA) in 2 patients and coronary sinus (CS) and left atrium (LA) in 1 patient each (Figures 2,3). One patient underwent Computed tomography scan showed, left circumflex coronary fistula opening into the coronary sinus (Figure 4). Patient in NYHA IV required medical stabilization before surgery. Fifteen patients underwent surgical correction and one patient underwent coil embolisation. The surgical technique used was, off pump ligation in 5 (33.3%) patients (Figure 5), on pump closure in 10 (66.6%) patients. Off Pump – Terminal fistula without associated anomalies and On Pump – RA, RV inflow, MPA & LA or associated anomalies. A detail of surgical procedure is listed in (Table 2). Mean cardiopulmonary bypass time was 86 minutes and mean aortic cross clamp time was 46 minutes. Associated procedures were performed like, Atrial septal defect closure in 2 patients, Tetrology of Fallot correction and open mitral commissurotomy in one patient each. One patient underwent coil embolisation, Using JL diagnostic catheter SOFT TTERUMO 0.035” x 260 centimeter wire passed from LAD to fistula to RV to PA which was snared out from Right femoral vein approach and thus a-v rai created. Despite multiple attempts 5Fr. PDA sheath could not be tracked from RFV across the fistula due to very tortuous course and using mother & child approach finally 5Fr long PDA sheath tracked from arterial side into LAD into fistula 8Fr. vascular plug deployed from right femoral artery (RFA) to LAD to FISTULA and position confirmed with JL catheter injection in left system to confirm vascular plug position beyond distal LAD (Figure 6).

RESULTS

There was no immediate mortality or no myocardial infarction or arrhythmias. Intra operative and immediate post-operative echocardiography showed, no residual or recurrent shunt in all (Figure 7), and good ventricular function in all except one who had biventricular dysfunction. Mean ventilation time was 7.5 hours and mean intensive care unit stay was 2.5 days. Follow up was done at 3 months, 1 year, 5, 10 and more than 10 years. Average follow up was 82.5% with average follow up duration was 114 months and all patients were in NYHA class I, one patient was developed left ventricular dysfunction at 3 years follow up, one patient with atrial fibrillation developed moderate tricuspid regurgitation and right ventricular dysfunction at 5 years follow up. The longest follow up was 17 years and on follow up no residual or recurrence of fistula and coronary arteries remained dilated and tortuous with marginal reduction in the size. Only one patient underwent coil embolisation recently and 1st follow up showed no residual or recurrent fistula but dilated coronary artery. There was no late mortality.

DISCUSSION

Coronary artery fistula (CAF) is a unusual vascular anomaly that communicates between one of the coronary arteries and a cardiac chamber or a large thoracic vessel. The first reported case of a CAF was in 1865 by Krause [1]. Its prevalence is about 0.1-0.8% based on coronary angiography and echocardiography studies [2-5]. The true incidence is difficult to evaluate because about half of the cases may be asymptomatic and clinically undetectable until an echocardiogram or catheterization is performed. CAFs comprise 14% of congenital coronary artery anomalies [4], and represent 0.4% of all congenital cardiac

Table 1: Echocardiographic data with origin of fistula, termination of fistula and their numbers.

<table>
<thead>
<tr>
<th>Coronary Artery Origin</th>
<th>Termination of Fistula</th>
<th>No of Patients</th>
</tr>
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<tbody>
<tr>
<td>LCx</td>
<td>RA</td>
<td>4</td>
</tr>
<tr>
<td>LCx</td>
<td>CS</td>
<td>1</td>
</tr>
<tr>
<td>LCx</td>
<td>LA</td>
<td>1</td>
</tr>
<tr>
<td>LCx</td>
<td>PA</td>
<td>1</td>
</tr>
<tr>
<td>LAD</td>
<td>RV</td>
<td>4</td>
</tr>
<tr>
<td>LAD</td>
<td>PA</td>
<td>1</td>
</tr>
<tr>
<td>RCA</td>
<td>RV</td>
<td>3</td>
</tr>
<tr>
<td>RCA</td>
<td>RA</td>
<td>1</td>
</tr>
</tbody>
</table>

Abbreviations: LAD- Left Anterior Descending Coronary Artery; LCX-Left CircumflexCoronary Artery; RCA-Right Coronary Artery; RA-Right Atrium; CS- Coronary Sinus; RV- Right Ventricle; PA-Pulmonary Artery; LA- Left Atrium.
**Figure 2** Angiography.
A. Left coronary angiography shows, LAD to PA fistula. LAD-Left anterior descending coronary artery, PA-Pulmonary artery.
B. Aortic root angiogram shows, LAD to RV fistula. LAD-Left anterior descending coronary artery, RV- Right ventricle.
C. Aortic root angiogram shows, RCA to RV fistula. RCA- Right coronary artery, RV- Right ventricle.

**Figure 3** Aortic root Angiography.
A. LCX to RA fistula.
B. LCX to RV fistula.
C. LCX to CS fistula.
**Abbreviations:** LCX- Left Circumflex Coronary Artery; RV- Right Ventricle; CS- Coronary Sinus.

**Figure 4** Computed coronary angiography showing LCX to CS fistula.
A-F: Origin, course and termination of LCX-CS (* mark).
**Abbreviations:** LCX- Left Circumflex Coronary Artery; CS- Coronary Sinus.
Figure 5 Intraoperative images.
A-C- LAD to RV fistula
D-F- LCX to RV fistula
Abbreviations: ALAD- Left Anterior Descending Coronary Artery; LCX- Left Circumflex Coronary Artery; RV- Right Ventricle.

Table 2: Operative techniques used for the patients.

<table>
<thead>
<tr>
<th>OPERATIVE TECHNIQUE</th>
<th>No. OF PATIENTS</th>
</tr>
</thead>
<tbody>
<tr>
<td>Ligation</td>
<td>5 (31.25%)</td>
</tr>
<tr>
<td>Through cardiac chamber</td>
<td>6 (37.5%)</td>
</tr>
<tr>
<td>Through MPA</td>
<td>2 (12.5%)</td>
</tr>
<tr>
<td>Tangential arteriorrhaphy</td>
<td>1 (6.25%)</td>
</tr>
<tr>
<td>Combined ligation and through cardiac chamber</td>
<td>1 (6.25%)</td>
</tr>
<tr>
<td>Combined arteriorrhaphy and through cardiac chamber</td>
<td>1 (6.25%)</td>
</tr>
</tbody>
</table>

Abbreviations: MPA- Main Pulmonary Artery.

Figure 6 Left coronary angiogram showing, LAD to RV fistula with vascular plug (Arrow).
Abbreviations: LAD- Left Anterior Descending Coronary Artery; RV- Right Ventricle.

malformations [6]. Approximately 10-30% of patients with a CAF also have another congenital cardiovascular anomaly [7,8]. The most commonly seen defects include variations of tetralogy of Fallot, patent ductus arteriosus, atrialseptal defect, ventricular septal defect and pulmonary stenosis. The majority of CAFs arise from the right coronary artery and the left anterior descending artery; the circumflex coronary artery is rarely involved. They most frequently drain into the right ventricle, right atrium or pulmonary artery. Few of them drain into the coronary sinus, left ventricle, left atrium, pulmonary vein or superior vena cava. Generally, most CAFs manifest as a single fistula; cases of multiple fistulas are rare. CAFs can be found at any age and are not gender specific.

Pathophysiology

According to the site of drainage, CAFs have varied physiologic presentations. Latson et al., described the physiology of CAFs in detail [9]. CAFs that drain into the right side of the circulation create a left-to-right shunt of oxygenated blood back to the pulmonary circulation. Those that drain into the
Acquired CAFs have been reported to be due to an incremental use of intravascular procedures and frequency in recent years. It is possible that this increase may result in little or no dilation of the proximal coronary artery from which they arise, and are themselves no larger at any point than the normal expected proximal coronary artery diameter. Fistulas that, at any point, are larger than two times but less than three times the expected proximal normal coronary artery diameter, or that are associated with a similar range of dilation of the proximal associated coronary artery, are considered to be medium-size fistulas. Fistulas that are more than three times the proximal coronary artery diameter are considered large. This classification may be useful in making clinical decisions.

**Clinical manifestation**

Most CAFs are found incidentally during cardiac catheterization. The clinical manifestations vary according to the size of the fistula, drainage site and patient’s age. About half of them are asymptomatic, but angina, myocardial infarction, heart failure, arrhythmia, endocarditic and aneurysm rupture have been reported. Said et al., reported that in 76 patients with CAFs, 55% of the patients were a symptomatic, 34% had chest pain, 13% had congestive heart failure and 1% had arrhythmia [18]. Symptoms and complications of CAFs are less common in children but more significant in adults. In the literature review and analysis by Liberthson et al., in 1979, symptoms occurred in only 19% of young patients (20 years), where as 63% of patients over the age of 20 had either symptoms or complication due to the fistulas [21]. The most common physical finding is a heart murmur. The typical murmur of a moderate or large CAF is continuous murmur that tends to be crescendo-decrescendo in both systole and diastole, louder in diastole, however. Differential diagnosis includes persistent ductus arteriosus, pulmonary arteriovenous fistula, and ruptured sinus of Valsalva aneurysm, ventricular septal defect with aortic valve incompetence, aorto pulmonary window, and systemic arteriovenous fistulas. Isolated systolic or diastolic murmurs have occasionally been reported [22]. The site of maximal intensity of the murmur is related to the site of drainage. Signs of pulmonary plethora and cardiomegaly at X-ray and ECG signs of right ventricular volume overload could be noted if there is large volume of flow through a CAF that produces a left-to-right shunt. Myocardial ischemia with abnormal 201 Tl perfusion image can be detected in large portion of patients with CAF [23]. The absence of Tl perfusion defect in patients with CAF may be due to micro-fistula without evident steal phenomenon of coronary blood flow. Coronary angiography is the major diagnostic tool. It can demonstrate the size, anatomy, number, origination and termination site of the fistulas. Cardiac echocardiography is also useful for diagnosis [24–26]. Magnetic resonance imaging and multi-detector computed tomography are also used to evaluate the anatomy, flow and function of CAFs [27–29].
Racial differences

Most CAFs were reported from studies in Caucasian people. Chiu et al., presented the first report of Oriental CAF patients with long-term follow-up and with a large number of patients [30]. From September 1992 to August 2007, 152 CAFs were detected in 28,210 coronary angiograms from 125 Chinese patients (incidence: 0.4%). Of the 125 patients, 58% of CAFs originated from the left anterior descending artery and 29% of CAFs originated from the right coronary artery. Most of CAFs (63%) drained to the pulmonary artery. Chiu et al., classified the CAFs into two types: type I in 99 patients with 124 solitary coronary to cardiac chamber or great vessel fistula; type II: 26 patients with 28 coronary artery-left ventricular multiple micro fistulas. The incidence (0.09%) of type II CAFs in this report is significantly higher than those previously reported in Caucasian people. This incidence of type II CAFs was 0.015% in one of the largest series of 33,600 consecutive angiograms [31]. Type I CAFs predominantly originated from the proximal segments (76%) and type II fistulas all originated from the mid (50%) or distal (50%) segments of the coronary artery. Single-, double-, and triple- CAFs were detected in 79%, 20%, and 1% of patients, respectively. The incidence of bilateral/multiple fistulas was also higher than those reported in Caucasian people. Coexistent coronary lesions were noted in 41% of patients. Fistula related symptoms included stable angina in 55, myocardial infarction in 2 and heart failure in 2 and sudden death with ventricular fibrillation in 1, and syncop e in 1. Twenty-four (20%) of patients had coexistent congenital anomalies. Unlike those reported CAF in Caucasian people, the most common coexistent congenital anomaly was myocardial bridge. Most patients received medical treatment because of mild symptoms. Only 9 patients underwent coronary intervention or and surgery for CAFs.

Treatment

The management strategy of patients with CAF depends on the size of the fistula, presence of symptoms, the anatomy of the fistula, the patient’s age and whether the patient has other associated cardiovascular disorders. Small CAFs are usually asymptomatic, and may close spontaneously [32-34]. Patients with a small CAF have a good long-term prognosis and should be treated conservatively. Medical treatment with either beta blockers or calcium channel blockers is suggested [35,36]. Prophylaxis for bacterial endocarditis is recommended in all CAF patients and in patients after complete fistula occlusion for at least 1 year [37]. There appears to be good consensus that all symptomatic patients should undergo closure of medium or large CAFs. Saide et al., suggested surgery or percutaneous transcatheter embolization to treat patients that have moderate or large CAFs with $\frac{Q_p}{Q_s} \geq 2.18$. The first successful surgical closure was reported by Börck and Carfoo in 1947 [38], where as the first therapeutic embolization was performed in 1974 by Zuberbuhler et al., [39]. Because transcatheter closure of CAF is associated with a much shorter recovery time and avoids a scar, it is considered the procedure of choice when fistula closure is indicated. Catheter closure can be performed with a variety of techniques, including detachable balloons, stainless steel coils, controlled release coils, controlled-release patent ductus arterious coils, patent ductus arterious plugs, regular and covered stents, and various chemicals [14,40-45]. Surgical ligation should be reserved for patients who have a complex and distally located fistula, or have adjacent vessels at risk. In addition, surgical ligation may be preferred when correction of other congenital defects or coronary artery bypass grafting is required 47. Mortality related to surgical closure or transcatheter closure of isolated CAFs is slow (1%) 48. Incomplete closure has been seen in ~10% of patients treated by catheter techniques or surgical ligation [45].

Until now, there is limited longitudinal information about the long-term prognosis of patients with CAFs following surgical or transcatheter treatment. Cheung and colleagues [6] found 96.9% of the patients in their series remained asymptomatic at the mean follow-up of 9.1 years, and approximately 10% had demonstrable recurrence of fistula without hemodynamic disturbance. Half of the patients underwent follow-up angiography, which showed a dilated and tortuous native coronary artery or more frequently showed thromboses with a short proximal stump [6,46,47]. None of our patient showed thrombosis or occlusion of coronary artery, but coronary artery remained dilated as long as 16 years after surgery. Grifka et al., reported [46], the long-term patient outcome after fistula occlusion remains unknown, but intermediate-term results reveal persistent coronary artery dilatation in many of these patients. Therefore, patients who have undergone coil occlusion of coronary fistulae require close follow-up; and, in certain cases, the use of antiplatelet therapy or low-dose anticoagulant may be warranted [46]. We have not used antiplatelet or anticoagulant in our patients.

Prognosis

Recent results of transcatheter and surgical approaches indicate a good prognosis from both techniques. Life expectancy is considered normal. However, risk of degenerative atherosclerotic disease may be higher if ectasia and dilatation of the coronary artery persist or progress.

In young surgical patients, one can anticipate the involution of the dilated segment of the feeding vessel; this is not the case in adults [6,45-47].

CONCLUSION

Coronary cameral fistula is a rare congenital anomaly. CAF may be isolated or in association with other cardiac defects. It may arise from one coronary system or involve both coronary systems. Fistulas draining into the coronary sinus are prone to develop congestive cardiac failure. Fistulas draining in RA/CS are prone to develop AF & its sequel. Most of the fistulas drain into the right side of the heart but it may originate from the left coronary system than the right. Cardiac cath and coronary angiography gives definitive diagnosis and planning management and to r/o other cardiac lesions. MDCT angio may replace angiography in the future for noninvasive diagnosis. Percutaneous closure is the preferred choice of closure. Surgery is indicated for lesions not amenable to percutaneous closure. Surgical closure can be done with or without CPB by ligation, tangential arteriorrhaphy or closure via cardiac chamber. Off Pump ligation technique can be used for Terminal fistula without associated anomalies. On Pump closure required in fistula draining in – RA, RV Inflow, MPA & LA or associated anomalies. Watch for ischemic changes while ligating the fistulas without CPB. Intraoperative cardioptlegia
delivery and intraoperative trans esophageal echocardiography is helpful in assessing adequate fistula closure. Early and Long term follow up showed good outcome. With atrial fibrillation, long term follow up showed right ventricular dysfunction and tricuspid regurgitation. Severe coronary artery dilatation (>10 mm) may warrant anticoagulation with warfarin, especially in patients with sluggish coronary flow, although there is little information available concerning the risk of coronary thrombosis in this group.

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