

Congenital solitary coronary artery fistulas characterized by their drainage sites

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Abstract

Last centuries have witnessed tremendous sophistication and progress in the detection, diagnosis and treatment of coronary artery fistulas (CAFs). In many countries, CAFs were reported to be visualized and treated using several imaging techniques and different management strategies. Reports from nearly all continents of the globe have contributed to the description of CAFs, not only in Asia and Europe but also throughout North and Latin America. However, these reports have to be cautiously analyzed as many of them were published as a case report and careful interpretation is warranted due to possible publication bias. A literature search was performed using PubMed search interface to select papers dealing with congenital CAFs in adult population between 2000-2009. A total of 233 subjects were collected, and analysed according to their drainage site and treatment modality. They were divided into two subgroups: percutaneous transluminal embolization group (PTE group, $n = 122$) and surgical ligation group (SL group, $n = 111$). In the SL group, atherosclerotic coronary artery disease (19%) and associated congenital lesions (23%) were more prevalent compared with the PTE group (9% and 8%), respectively. Infective endocarditis was more frequently seen in the SL group besides syncope, congestive heart failure and hemopericardium. In both groups multimodality diagnostic workup composed of several non-invasive

and invasive imaging techniques for fistula visualization were performed and drainage sites into the different cardiac chambers and intrathoracic great vessels were similarly distributed in the two groups.

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Key words: Congenital anomalies; Solitary coronary artery fistulas; Adult population; Diagnostic modalities; Therapeutic options

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INTRODUCTION

When we consider congenital coronary artery fistulas (CAFs), we have to bear in mind that there are two separate entities: solitary CAFs and coronary artery-ventricular multiple microfistulas (MMFs). The current review focuses exclusively on solitary CAFs.

During the last decennia contributions of different investigators, researchers and scientists across the globe guided us to understand more but not yet all about CAFs.

Although coronary angiography (CAG) is the gold standard for detection and visualization of CAFs, other non-invasive techniques are frequently applied to delineate the anatomical morphological features and assess the function of the fistula. These modalities include transesophageal echocardiography (TEE)^[1], 2-dimensional echo^[2], 3-dimensional TEE imaging^[3], cardiovascular magnetic resonance imaging (CMR)^[4] and multidetec-

tor computed tomography (MDCT)^[5]. Furthermore, direct imaging, by different modalities, at the time of surgical correction may be required; intraoperative ECG monitoring can be a very useful tool to guide and detect perioperative ischemic changes and TEE is also helpful to demonstrate wall motion abnormalities^[1] and confirm the complete ligation of the fistula.

In the last decades, CAFs have been more often discovered and diagnosed as a result of initially raised clinical and echocardiographic suspicion and subsequently due to the frequent application of CAG. The angiographic incidence of CAFs is estimated at 0.2%-0.8% of patients undergoing CAG^[6,7]. While there is a common opinion that surgical or transcatheter intervention to obliterate the fistula and preserve the normal coronary blood flow should be performed in patients with large shunts, documented ischemic changes, threatening future complications, and concomitant congenital or acquired coronary or valvular heart disorders, the optimal management of asymptomatic or mildly symptomatic patients with a medium-sized left-to-right shunt is not yet established. The hemodynamic indication for intervention as assessed by the pulmonary-to-systemic flow ratio should be more than 1.5^[8]. The current treatment strategies, depending on the magnitude of left-to-right shunt and the morphological features and functional characteristics of CAFs are: watchful waiting and close follow-up, conservative medical management, percutaneous transcatheter "therapeutic" embolization (PTE) and surgical ligation (SL). The reported success rate of PTE in a mixed (paediatric and adult subjects) series of 15 patients of Alekyan *et al*^[9] in 2002, was 93% with one early death and no recurrence after a follow-up period up to 13 years. Another series in 2001 by Wang *et al*^[10] demonstrated that surgical closure of the fistula under cardiopulmonary bypass was successful without residual fistula with no mortality or significant morbidity. Early surgical intervention is safe (100% survival and 100% closure rate) in CAFs with a perioperative mortality of 0%-4% with an increasing incidence with age of postoperative surgical complications and sequelae from less than 1% in patients under 20 years of age to 23% in those above 20 years of age^[11,12].

A high successful percutaneous closure rate (75%-87%) has been reported by Reidy *et al*^[13] in the 1990s and Trehan *et al*^[14] in 2004, respectively. Armsby *et al*^[15] reported an angiographic recurrence rate of 9% after PTE and the recurrence after surgical closure varied from 16% to 22% as reported by Kamiya *et al*^[16].

A literature search was performed using PubMed search engine to select papers published in English language dealing with congenital CAFs in adult population between 2000-2009. References were then cross-checked for other relevant publications. The keywords used were congenital coronary anomalies, CAFs and adult population. The reports were screened for those that stated the drainage site and adult population group. Papers concerning pure paediatric population were neglected. A total of 63 reports were selected.

From the literature between 2000 and 2009, 233 patients who were treated for congenital CAFs were collected. They were divided into two groups according to the treatment strategy. The first group consisted of 122 subjects treated with percutaneous transluminal "therapeutic" embolization (PTE) techniques^[9,14,15,17-27] and the second group composed of 111 patients was treated with surgical ligation (SL) of the fistula^[3-5,28-34].

TERMINATION SITES INTO DIFFERENT CHAMBERS OF THE HEART AND TO OTHER THORACIC VESSELS

CAFs are abnormal congenital communications between one or more coronary arteries and any cardiac chamber or intrathoracic vessel bypassing the capillary bed and are classified according to Greenberg *et al*^[35] as anomalies of termination. CAFs are among the most hemodynamically significant coronary artery anomalies^[36].

Two distinct congenital types are recognized: solitary CAFs and MMFs^[37]. Usually fistulas originate from the right coronary artery in 15%-53% and from the left coronary artery in 42%-67% of cases^[2,6,36,38]. The overwhelming majority is unilateral (80%-89%) followed by the bilateral fistulas (16%) and finally they may involve all three coronary arteries as multilateral fistulas (4%-5%)^[2,6,36]. Fernandes *et al*^[2] reported multiple fistulas in 11% of the subjects without differentiation between bilateral and multilateral contribution. It has been noted that bilateral^[39] CAFs terminate more often into the pulmonary artery (PA) (56%) while in contrast the unilateral CAFs ended in the PA in 17% of the cases^[36].

Several authors have disclosed the termination sites of CAFs as indicated in (Table 1)^[16,36,40,41]. Termination into the right side of the heart occurs in more than 90% of cases^[2,42]. Generally, acquired CAFs, coronary artery-ventricular multiple microfistulas and paediatric population are excluded from the current review.

Right ventricle

Termination into the right ventricle (RV) has been reported from several countries across the globe, Asia and Europe^[9,10,43-45] represented by several authors. Among the non-invasive diagnostic workup imaging techniques were myocardial perfusion test, echocardiography, CMR and MDCT. The invasive gold standard CAG was performed in all reports. The postoperative course was reported to be uneventful in patients with surgically ligated fistulas^[43]. Conservative medical management was the therapeutic strategy in the majority of the reports^[9,10,43-45]. They included symptomatic adult subjects who had continuous cardiac murmur and all were in sinus rhythm.

Right atrium

Reports about termination to the right atrium (RA) came from North and Latin America^[3,4], Asia and Europe^[5,17,18,44-48].

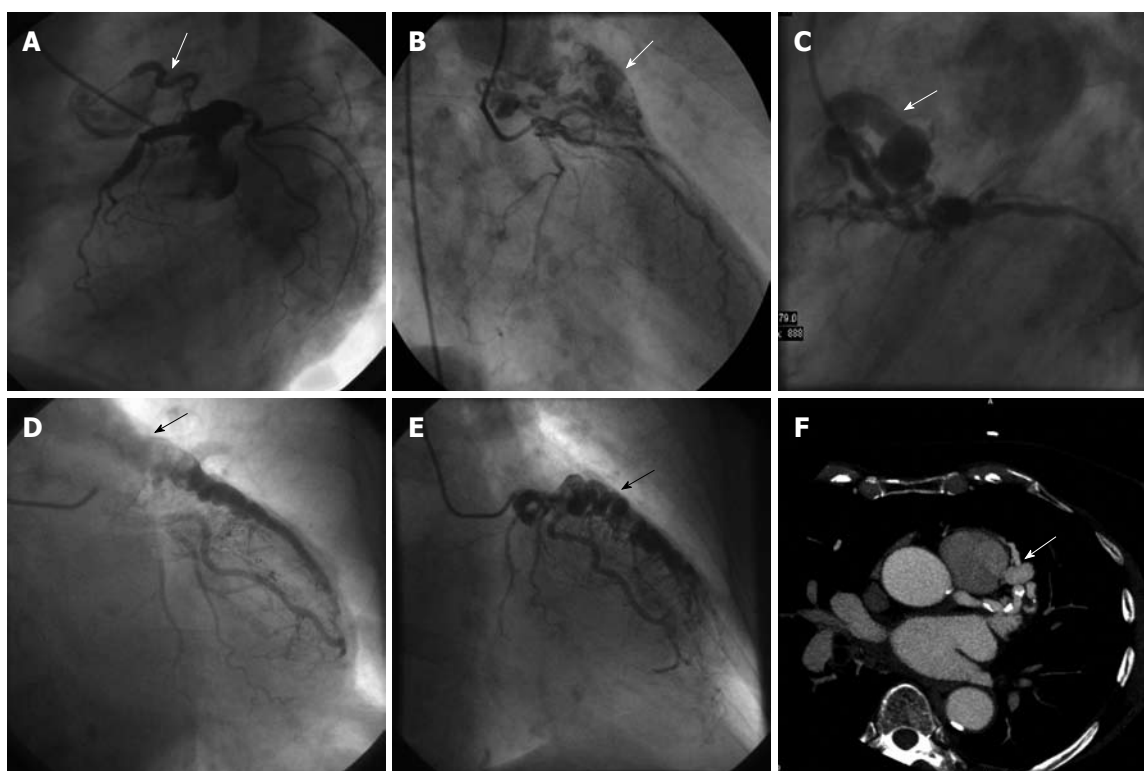


Figure 1 Different morphologic appearances of coronary artery-pulmonary artery fistulas (arrows). A: Left lateral view of coronary angiogram demonstrating a fistula with a single origin, tortuous pathway and single termination (arrow); B: Right anterior oblique (RAO) view showing a fistula with multiple origin and outflow associated with a plexiform pathway (arrow). Shallow filling of the coronary arteries is visible; C: Left lateral projection illustrating a fistula with multiple origin and termination associated with aneurysmal formation (arrow), and total occlusion of the left anterior descending coronary artery (LAD) is appreciated; D and E: Sequential frames in RAO projection depicting a huge tortuous LAD fistulating into the pulmonary artery (arrows); F: Multislice cardiac gated computed tomography scan demonstrating the fistula running from the LAD into the pulmonary artery (arrow), and the fistula is connected to the pulmonary artery at the anterior side.

Table 1 Termination sites of congenital solitary CAFs (%)

Termination sites	McNamara 1969 ^[40] (n = 172)	Levin 1978 ^[36] (n = 363)	Hobbs 1982 ^[41] (n = 122)	Kamiya 2002 ^[16] (n = 266)
Right ventricle	40	41	3	34
Right atrium	35	26	7	18
Pulmonary artery	16	17	66	38
Coronary sinus	--	--	--	--
Left atrium	6	5	7	2
Left ventricle	3	3	17	5
Superior vena cava	--	1	--	--
Right ventricle outflow tract	--	--	--	--
Other sites	--	--	--	3

CAFs: Coronary artery fistulas; --: Not applicable.

The clinical features were variable including asymptomatic presentations, dyspnoea, congestive heart failure, chest pain, syncope and myocardial infarction^[3-5,17,18,44-48]. In the reviewed reports, multimodality imaging techniques were instituted including 2D echocardiography^[4,46-48], 3D transesophageal echo^[3], MDCT, 4-detector row^[47] or 128-detector row^[5], CMR^[4], aortogram^[48], cardiac catheterization and CAG^[5,17,18,44-47].

The pulmonary-to-systemic flow ratio was found to be more than 1.5^[17,18,47]. Normal PA pressure (PAP) was described in some reports^[18]. Mild to moderate elevation of PAP was also elaborated in some studies^[17].

Successful surgical ligation with neither postoperative morbidity nor mortality was performed. The procedures were conducted using cardiopulmonary bypass^[5,48] or utilizing off-pump technique^[3,4,47]. In the reports of PTE treated patients, the use of coils^[18] or Amplatzer duct occluder^[17] was reported. One report showed complications of myocardial infarction after procedural period^[17]. The patient required an urgent coronary artery bypass grafting.

PA

Many papers have reported the drainage of the fistula to the PA (Figure 1). These originated from different continents, Europe^[19,20,28-30,45,49-51], Africa^[52], Asia^[21,22,53-60] and North America^[23].

Many reports were published describing unilateral^[19,28,30,45,53,58], bilateral^[21,29,54,57] and multilateral^[60] coronary artery-PA fistulas. All reports included adult symptomatic patients presented with typical or atypical chest pain, acute coronary syndrome, dyspnoea, syncope, fatigue, arrhythmias or congestive heart failure. Non-invasive and invasive diagnostic multimodalities were established and included the following techniques: echocardiography, CMR, myocardial perfusion test, MDCT, cardiac catheterization and CAG^[19-23,28-30,45,49-60]. The treatment modalities in these reports were conservative medical management, SL with extra corporeal circulation or without cardiopulmonary bypass and endovascular closure of the fistula. The materials used for transcatheter

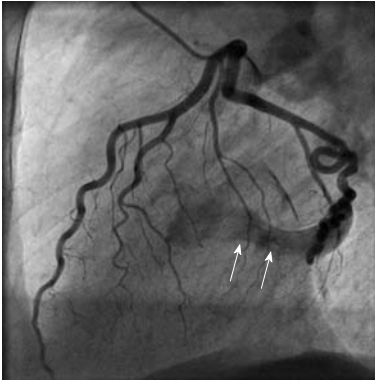


Figure 2 A fistula originating from the circumflex coronary artery and terminating into the coronary sinus (arrows).

occlusion of the fistula were coils, detachable balloon and stent-graft (Jostent). Among those reports of surgical or non-surgical exclusion of the fistula, there was no significant morbidity or mortality. One report described the successful application of a harmonic scalpel during SL of quadruple fistula all draining into the PA^[57].

Coronary sinus

Drainage into the coronary sinus (CS) (Figure 2) has been reported in the world literature from the following continents; Australia^[61], Europe^[62-68], Asia^[69-76] and North America^[31,32,77].

It was found in these reports that the patients may have lack of symptoms throughout life and others may develop or possess gradual progression of symptoms over the years varying from 5 to 40 years^[67,69,71]. While some become symptomatic in infancy and childhood requiring early management. The reports^[61-68] included patients presented with dyspnoea, typical and atypical chest pain, palpitation, atrial fibrillation, continuous cardiac murmur, pericardial effusion and cardiac tamponade^[69-75], infective endocarditis, congestive heart failure, abnormal echocardiographic findings, respiratory tract infection and vertigo^[31,32,76,77].

Multimodality imaging for the precise diagnosis of CAFs (origin, pathway and termination) is essential. This provides complete anatomical and functional data. Several reports described the diagnostic workups, including non-invasive and invasive techniques such as myocardial perfusion test, transthoracic and TEE, MDCT, CMR, cardiac catheterization and CAG^[31,32,61-77]. None of the reports mentioned the use of intracoronary Doppler ultrasound.

The reported pulmonary-to-systemic flow ratio was found to be significant^[61,67,69-71,75-77] in some papers and negligible in others^[62,63], calculated by the oxymetric method in all^[61-63,67,69-71,75-77]. The PAP was normal or proved to be mildly elevated in the majority of the reports^[62,69,72,75,77]. Only one report described significant elevation of the pulmonary pressure^[67].

The reported treatment strategies were SL and conservative medical management. Surgical ligation was

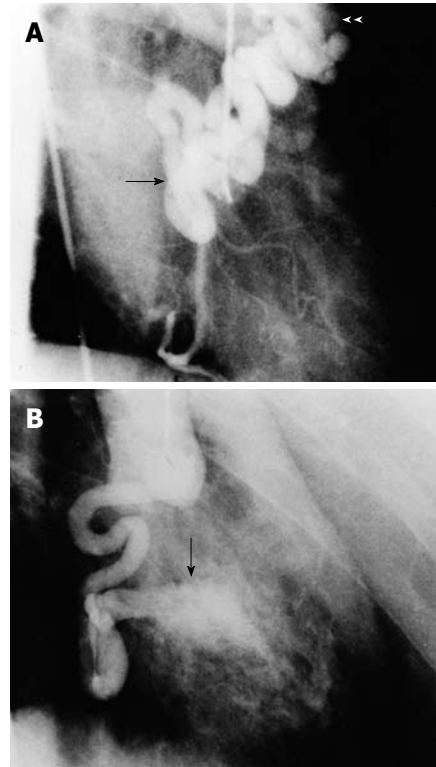


Figure 3 Different morphologic appearances of a dilated right coronary artery terminating into the pulmonary artery and the left ventricle (arrows).

A: RAO projection of a fistula originating from the proximal segment (arrow) of the right coronary artery and terminating into the pulmonary artery (arrowheads); B: A fistula from the distal segment (arrow) ending to the left ventricle.

performed with cardiopulmonary bypass^[67,68,71,73]. Conservative medical management consisted of β -blockers and calcium channel antagonists and/or watchful follow-up policy. Non of the reports dealt with PTE. Increased success rate of surgical closure of CAFs draining into the CS could be reached by the instillation of intraoperative fistula image guidance with TEE^[67,68,71,73] and/or on site CAG^[65] as a complementary imaging facility to each other. The latter is not widely applied in the operating rooms. These image guidance techniques during surgery could be used in cases with complex anatomy and multiple sites of origin or termination. Generally, cardiopulmonary bypass is especially required in cases with associated valvular or coronary heart diseases, drainage into CS and complex fistula anatomy. This has the advantage of direct intracardiac closure of the fistulous opening and furthermore for identification of the distal end by briefly interrupting the aortic cross-clamp or infusing cardioplegic solution.

Left ventricle

These coronary-cameral fistulas ending at the left ventricle (LV) (Figure 3) are extremely rare. Arterioarterial shunts cause isolated increased LV pressure and workload. These fistulas are mainly associated with a diastolic heart murmur. Flow through the fistulous shunt occurs exclusively in diastole mimicking aortic regurgitation murmur.

During systole, the fistulous opening draining into the LV is obliterated. In the reports from Europe^[33] and Asia^[44,78], echocardiography suspected the diagnosis of CAFs^[33] which was confirmed non-invasively by CMR and invasively by CAG. Non-invasive CMR and 64-row MDCT^[78] delineated the morphologic features of the fistula.

Superior vena cava, pulmonary vein, right ventricular outflow tract and left atrium

Few reports have been collected concerning the termination into superior vena cava, pulmonary vein (PV), right ventricular outflow tract (RVOT) and left atrium mainly from Asia^[10,44,79] and to a lesser extent from North America^[75]. Termination into the PV is very rare^[24]. Drainage to the RVOT is highly infrequent^[79]. These were all adult symptomatic patients who were treated by surgical closure of the fistula and performed off-pump^[79] or by percutaneous coil occlusion of the fistula^[24]. There are very few reports concerning the termination of the fistula into LV^[10,44].

TREATMENT MODALITY

In the PTE group, the presenting symptoms were dyspnoea, chest pain and angina pectoris, palpitation and fatigue. On the contrary, besides these symptoms, syncope, congestive heart failure and hemopericardium were found more frequent in the SL group.

Drainage sites were comparable in both groups. The pre-intervention diagnostic workup in the PTE group provided the decision making after echocardiography, myocardial perfusion test, MDCT, cardiac catheterization and CAG. The decision making was reached in the SL group after diagnostic workup using echocardiography, myocardial perfusion test, MDCT, cardiovascular magnetic resonance, chest CT scan, cardiac catheterization and CAG. Clinical presentation with infective endocarditis was found in 10% of the patients in the SL group but in none of the PTE group.

Surgical ligation with cardiopulmonary bypass was used in 67% of the patients in the SL group and off-pump procedures were reported in 9%, while no cardiopulmonary bypass was reported in 24% of the patients.

Occlusion materials used in the PTE group were: all kinds of coils, 84%, stent 3%, ADO and Amplatzer septal occluder 3%, balloon (silicon and latex) 2%, umbrella devices 5% and Grifka and floppy tips of guide wires in 3% of the patients.

Complete closure of the fistula was confirmed with clinical examination, non-invasive and invasive assessment using echocardiography, phonocardiography, myocardial perfusion test and CAG in the PTE group. But in the SL group, echocardiography, myocardial perfusion test, MDCT and cardiac catheterization and CAG were usually used.

CONCLUSION

In the current review, the wide spread spectrum of con-

genital CAFs is not fully represented in this limited series of reports. However, this review does emphasize the two separate manifestations of the fistulas: the congenital solitary CAFs which is the cornerstone of this report and MMFs. MMFs are excluded from the current study because they form a different entity.

CAFs are usually detected on CAG. They may be simply suspected by 2-D echocardiography. Multi-imaging modality (TEE, MDCT, CMR, CAG and cardiac catheterization) is required to adequately demonstrate the precise anatomical features (origin-pathway-termination aneurysmal formation, multilaterality, multiplicity, tortuosity) and accurate functional and shunt flow characteristics. These may be very helpful during the follow-up period for the operated or non-operated patients. Intra-operative image guiding increases the success rate of the surgical results.

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