Acquired Coronary Cameral Fistulas: Are These Collaterals Losing Their Destination?

SALAH A. M. SAID, M.D., AND TJEERD VAN DER WERF, M.D., PH.D.*

Departments of Cardiology, Hospital Streekeziekenhuis Midden-Twente, Hengelo, and *Heart Centre, Academic Hospital Nijmegen, Nijmegen, The Netherlands

Summary

Background: The majority of coronary cameral fistulas (CCFs) are congenital in origin. On the other hand, acquired coronary cameral fistulas, having various etiopathogenic origins, are increasingly recognized.

Hypothesis: The aim of this study was to assess the possible involvement of coronary atherosclerosis in the pathogenesis of acquired coronary cameral fistulas.

Methods: Between 1993 and 1996 coronary cameral fistulas were detected in seven adults patients with coronary atherosclerosis ($n = 4$) and following myocardial infarction ($n = 3$) with a mean age of 59.3 years (range 40–77). They were analyzed at our hospital.

Results: Myocardial infarction (MI) was documented in six patients and was localized at the same territory of the fistula-related artery in three of them. All patients remained asymptomatic after the detection of the fistula. Five patients had associated cardiac disorders. Two patients were treated conservatively with medical therapy. Coronary artery bypass grafting (CABG) was performed in three patients. One patient died while awaiting CABG. Thirty-four adult cases with acquired CCFs were collected from the current literature. The right coronary artery was the origin of the fistula in 37% and they terminated into the right heart-side in 72% of cases. They remained asymptomatic in 62% of cases.

Conclusions: It could be concluded that acquired CCFs may complicate the course of severe atherosclerosis or myocardial infarction in certain adult patients. The symptomatology and treatment strategy is comparable in the congenital and acquired types. The distribution of involvement of the right or left coronary arteries is equally divided in both the acquired and congenital types. Further studies are needed to investigate the precipitating factors for the occurrence of and incidence of acquired CCFs in patients with severe atherosclerosis or post-MI subjects.

Key words: acquired coronary cameral fistula, coronary atherosclerosis, collaterals, myocardial infarction

Introduction

Coronary cameral fistulas (CCFs) are etiologically divided into congenital and acquired types. The vast majority of these lesions are congenital in origin.^{1–4}

Acquired CCFs may occur after heterogeneous causes of endogenous or exogenous trauma: exogenous trauma such as sharp^{5, 6} and blunt chest injury,⁷ or endogenous surgical or nonsurgical trauma such as barotrauma after percutaneous transluminal coronary angioplasty (PTCA),^{8–11} myocardial infarction (MI),^{12–16} permanent ventricular pacing,¹⁷ endomyocardial biopsy,¹⁸ and cardiac surgery.^{19–21} Coronary cameral fistulas may be spontaneously associated with severe coronary atherosclerosis.^{22, 23} In view of the increased use of endovascular procedures, acquired fistulas are reported increasingly.²⁴

This report presents the clinical manifestations and angiographic findings of seven adult patients with incidentally detected acquired CCFs associated with severe coronary artery disease or MI. The literature concerning the acquired CCFs is reviewed.

Study Patients

Seven adult patients with acquired CCFs (Figs. 1 and 2) were analyzed at District General Hospital Streekeziekenhuis

Address for reprints:

Salah A.M. Said, M.D.
Department of Cardiology
Hospital Streekeziekenhuis Midden-Twente
Geerdinksweg 141
555 DL Hengelo, The Netherlands

Received: June 5, 1998

Accepted with revision: October 26, 1998

FIG. 1 Acquired single coronary cameral fistula (CCF). Right coronary artery (RCA) in right anterior oblique projection (RAO), showing proximal total occlusion with bridge collaterals (*) filling the periphery (arrow) of the RCA and simultaneously blushing the right atrium (arrowheads). An example of acquired single CCFs originating from the RCA and terminating into the right atrium. The small caliber RCA, mainly seen in acquired CCF, is well appreciated. C = right Judkins catheter.

FIG. 2 Acquired multiple coronary cameral fistulas. Angiogram of the left coronary artery in right anterior oblique projection demonstrating multiple fistulas originating from the circumflex artery (Cx) to the lumen of the left ventricle (small arrows). Total occlusion of the left anterior descending (arrowhead) and significant stenosis of the Cx (o) arteries are well appreciated. C = left Judkins catheter.

Midden-Twente, Hengelo, The Netherlands, between 1993 and 1996.

Acquired CCFs are defined as a direct communication of a coronary artery with a cardiac chamber, provided that cardiac or coronary intravascular "iatrogenic or spontaneous" procedure/event or extravascular thoracic trauma has taken place, especially when extravasation of contrast is visible from the coronary artery into or adjacent to the left ventricle in an infarcted wall segment. Furthermore, the fistula arises proximal to severe coronary obstructive lesions.

None of our patients underwent therapeutic endovascular procedure prior to detection of the fistula, and none had penetrating or nonpenetrating thoracic trauma.

Results

This study consisted of seven adult patients (six men, one woman), ranging in age from 40 to 77 years (mean age 59.3 years). Patient characteristics are summarized in Table I. When shunts were calculated, none were over 1.6. The patients had seven CCFs. The origin of the CCFs was the right coronary artery (RCA) in five and the left coronary artery in two patients. Of these, the left anterior descending artery (LAD) participated in one while the circumflex artery (Cx) was the fistula-related artery in one patient. The sites of termination of the fistulous tract were the right atrium in four, the left ventricle in two and the pulmonary trunk in one patient. These CCFs were incidentally found during coronary angio-

graphic studies performed for analysis of ischemic heart disease, congestive heart failure, and cardiac arrhythmias.

Association of CCF with mitral valve prolapse (MVP) was seen in two patients (Nos. 1 and 7). Mitral regurgitation was present in two patients (Nos. 2 and 7).

Myocardial infarction occurred in five patients (Nos. 1, 2, 3, 4, and 6) and it was localized in the same territory of the fistula-related artery in three of these (Nos. 2, 3, and 6) (Figs. 1–3). Mean left ventricular end-diastolic pressure (LVEDP) was marginally elevated at 13 mmHg (range 5–20).

A systolic cardiac murmur was heard in two patients (Nos. 1 and 7). One patient (No. 6) had congestive heart failure and atrial fibrillation, with an elevated LVEDP of 16 mmHg. The fistula terminated directly into the lumen of the left ventricle. Echocardiographic measurement of left ventricular end-diastolic dimension (LVEDD) was evaluable in four patients with a mean of 56.3 mm (range 50–66). It was found dilated (LVEDD $\geq$ 64 mm) in two cases (Patients No. 1 and 6).

Severe coronary artery stenosis was defined as angiographic stenosis of $\geq$ 80%. Severe narrowing of the fistula-related artery (FRA) was found in four patients (Nos. 1, 2, 3, and 5) and total occlusion of the FRA in three (Patients No. 4, 6, and 7). Well-developed collateral circulation to the myocardial region of the severely stenotic or totally occluded FRA was demonstrated in four patients (Nos. 3, 4, 5, and 6). Bridge collaterals to the periphery of the FRA were found in Patients No. 3 and 4. Collateral circulation was angiographically undetected in three cases (Patients No. 1, 2, and 7). Three patients (Nos. 4, 5, and 6) had dual coronary artery disease (CAD), and triple CAD was found in four cases (Patients No. 1, 2, 3, and 7).

TABLE I Characteristics of seven adult patients with acquired coronary cameral fistula

Patient No.	Date of detection/ age/sex	Mode of detection Etiology	Collaterals and percent of stenosis CCF	MI	Associated disorders	CABG
1	14-02-1994 59/M	CAG Atherosclerosis	LAD-80%→coll→Cx Cx-100%, RCA-80% CCF: RCA → RA	1994 ASMI	MVP/RR	LIMA→ LAD VSM→ RCA VSM→ Cx
2	21-07-1993 72/F	CAG, Atherosclerosis	LAD-100%, pCx-90%, coll→ RCA, RCA-100%. Graft D-LAD: patent Graft OM-PL: 100% CCF: Cx → LV	1987 ALMI	MR	VSM→ D- LAD VSM→ OM- PL
3	03-05-1995 46/M	CAG Myocardial infarction	LAD-80%, D-70%, Cx-70%, coll→RCA, RCA-90%, bridge coll. to periphery CCF: RCA → RA	IMI, silent	None	LIMA→ LAD-D RIMA→ OM2 VSM→ PL-PD
4	15-11-1995 40/M	CAG Atherosclerosis	Cx-90%, RCA-100%, bridge coll. to periphery CCF: RCA → RA	1995 PMI, 1996 ILMI	None	Medical 2× PTCA of Cx
5	06-12-1995 53/M	CAG Atherosclerosis	Cx-99%, bridge coll. to periphery and RCA, RCA-95% CCF: RCA → PT	None	None	Medical
6	12-06-1995 68/M	CAG Myocardial infarction	LAD-100%, AL-90%, pCx-80%, RCA-coll. to LAD CCF: LAD → LV	1990 ASMI, pericarditis 1995 reMI	CHF/AF	PTCA of AL and Cx
7	03-06-1996 77/M	CAG Atherosclerosis	LAD-90%, Cx-90%, RCA-100% CCF: RCA → RA	IPMI	MVP/MR	Died, cardiac death while awaiting CABG

Abbreviations: M = male, F = female, CAG = coronary angiography, CABG = coronary artery bypass grafting, CCF = coronary cameral fistula, LAD = left anterior descending coronary artery, Cx = circumflex coronary artery, RCA = right coronary artery, LIMA = left internal mammary artery, RIMA = right internal mammary artery, VSM = vena saphena magna, D = diagonal branch, OM = obtuse marginal branch, PL = posterolateral branch, AL = anterolateral branch, coll = collateral, PD = posterior descending artery, RA = right atrium, LV = left ventricle, PT = pulmonary trunk, MI = myocardial infarction, reMI = recurrent, ASMI = anterosseptal, ALMI = anterolateral, IMI = inferior, PMI = posterior, ILMI = inferolateral, MVP = mitral valve prolapse, MR = mitral regurgitation, RR = hypertension, AF = atrial fibrillation, CHF = congestive heart failure, → = to.

Coronary artery bypass grafting (CABG) was performed in three patients (Nos. 1, 2, and 3). Two patients (Nos. 4 and 6) underwent PTCA for concurrent coronary artery stenosis of non-fistula-related arteries.

Review Subjects

Thirty-four adult cases were collected from the current literature in the period between 1971 and 1996; they had 43 acquired CCFs. The etiology varied from endovascular barotrauma and endomyocardial biopsy to chest injury.⁵⁻²⁵ The origin was the RCA in 16 of 43 (37%) and the left coronary artery (LCA) in 27 (63%) of the CCFs. They terminated into the right side of the heart in 31 of 43 (72%) and into the left side of the heart in 12 (28%) of the fistulas. Of 34 (62%) patients, 21 had no symptoms after detection of the CCF. Chest

pain occurred in 23% and dyspnea in 15% of the subjects. Four patients (12%) were managed by surgical ligation, 82% medically, 3% by percutaneous transcatheter embolization (PTE); 3% died while awaiting CABG.

On follow-up, 14 subjects remained asymptomatic. Spontaneous closure and near-closure was demonstrated in six subjects. Surgical ligation was performed in one patient. Three patients died. No details were available in the remaining 10 subjects.

Discussion

Coronary cameral fistulas are commonly congenital in origin¹⁻⁴ but they may be acquired subsequent to a heterogeneous group of causes.^{5-23, 25} The various reported causes of acquired CCFs and speculative factors thought to contribute

FIG. 3 Angiogram of a 30-year-old man (not included in the present series) of the right coronary artery (RCA) (arrows) in the right anterior oblique (RAO) projection demonstrating severely dilated and tortuous congenital coronary cameral fistula originating from the distal RCA (arrowheads) and terminating directly into the lumen of the left ventricle (LV). C = pigtail catheter.

to their development are listed in Table II. In our own series, all patients had an acquired CCF secondary to severe atherosclerotic changes and associated with MI. Coronary cameral fistulas were associated with MI in two and with severe atherosclerosis in five patients.

In the former two patients, it is possible that the fistula had formed as a result of the transmural MI facilitated by the occurrence of soft necrotic myocardial tissue in the infarction region. This complication has been described previously in literature.¹²⁻¹⁶

In the remaining five patients with associated severe atherosclerosis, the most likely mechanisms for the formation of the CCF are (1) aberrant development of collateral vessels, i.e., collaterals that "lost their way" or direction and terminated elsewhere (inadvertently into a cardiac chamber) than at the target myocardial region; (2) dilatation of arteriovenous channels that may permit enough shunting of blood to be visualized during coronary angiography.^{14, 22, 23} In one patient (No. 6), the development of post-MI pericarditis may also have played a role or facilitated the development of such a CCF. These CCFs are believed to be acquired secondary to atherosclerosis and MI because of the absence of dilatation (mostly seen in congenital cases) of the involved coronary artery and the presence of a consequent direct relation between the area of termination and the stenotic or occluded coronary artery responsible for the fistula formation.

Although most of the patients with CCF are asymptomatic, a few may initially present with a catastrophic manifestation such as syncope and cardiac arrest.²⁶

Termination of CCF in the left ventricle has been reported,²⁷ but is infrequent (3%).² In two of our patients, the CCF terminated into the lumen of the left ventricle. The LCA was the fistula bearing artery in both of these.

Coronary cameral fistulas may be single or multiple.²⁸ Bilateral or multiple CCFs, while rarely reported in congenital CCFs,²⁸ were even rarer both in our own series (1 of 7 cases) and in the current literature review of acquired CCFs (2 of 34 cases). Coronary cameral fistulas are either isolated²⁹ or associated with local aneurysmal formation^{27, 30} of the fistula-related artery. None of our patients showed aneurysmal formation of the fistulous tract or the fistula-related artery.

TABLE II Pathogenesis and speculative factors contributing to the formation of acquired coronary cameral fistula

First author/year (Ref. No.)	Etiology	Factors contributing to fistula formation
Meng 1985 (8)	PTCA	Intracardiac perforation
Iannone 1990 (10)	PTCA	Oversized balloon, inappropriate wire tracking
Grill 1991 (9)	PTCA	Soft infarcted tissue surrounding ruptured artery
El-Omar 1995 (11)	PTCA	Oversized balloon, inappropriate wire tracking, and intracardiac perforation
Lee 1989 (13)	MI	Limited bleeding and rupture of the coronary arteries into necrotic myocardial site creating the coronary cameral fistula
Shimizu 1993 (15)		
Somsen 1994 (16)		
Searcy 1971 (23)	MI	Aberrant development of collateral vessels leading to coronary cameral fistula formation, ^{22, 23} or by reopening of the Thebesian veins ¹⁴
Ryan 1977 (14)		
Ray 1992 (22)		
Lee 1988 (19)	Cardiac surgery	Direct injury of right atrial artery leading to the development of coronary cameral fistula
Cheng 1973 (5)	Trauma	Traumatic laceration of coronary artery by stab wound
Saeian 1991 (17)	Cardiac pacing	Erosion of the permanent ventricular pacing lead into the interventricular septum creating coronary cameral fistula
Sandhu 1989 (18)	Biopsy	Result of repeated right ventricular endomyocardial biopsy

Spontaneous closure has been reported for congenital CCFs,^{31, 32} and it has also been demonstrated in acquired CCFs.^{18, 25} It has been estimated that spontaneous closure occurs in 1–2% of the patients.³¹

Patients with acquired or congenital CCFs with a large-size shunt are treated either with percutaneous transcatheter embolization techniques^{20, 33} or, when the anatomy is not amenable, by surgical ligation method. In patients who cannot undergo surgery because of the risk of congestive heart failure and endocarditis, close long-term follow-up is recommended and antibacterial prophylactic measures should be considered. In the present series, the magnitude of the left-to-right shunt is small (mean Qp/Qs 1.35). In the review subjects (34 cases), the shunt size was not reported. Of the 7 patients with acquired CCFs in our series and the 34 acquired CCFs from literature, none had symptoms attributable to the fistula. Based on the findings in our own series and literature review, the symptomatology in the acquired CCFs is not unique, but is usually related to the underlying cause: in our series, for instance, it was related to severe obstructive coronary atherosclerotic lesions and MI.

In the literature review (34 cases), most patients with acquired CCFs were treated medically, but surgical ligation (4 times) or PTE (once) were performed in few subjects. At follow-up, spontaneous closure or near-closure of the CCF was documented in six of the review cases. Three patients with acquired CCFs (related to PTCA in two patients and secondary to MI in the third) died.

Conclusions

As collaterals “lose their way,” acquired CCFs can probably develop in patients with severe atherosclerosis and MI. No difference was found between congenital² and acquired CCFs in terms of vessel involvement. In our series (seven cases), the systemic to pulmonary shunt was small. In the current literature review and in our series, the acquired CCFs showed no specific or unique symptomatology, and the majority of patients with acquired CCFs were treated medically.

The question remains whether patients with acquired CCFs require endocarditis prophylaxis as recommended in congenital CCFs.

Acknowledgement

The authors wish to thank Mrs. H.E. Blijdenstein for her expert secretarial assistance during the preparation of the manuscript.

References

1. Yamanaka O, Hobbs RE: Coronary artery anomalies in 126; 595 patients undergoing coronary arteriography. *Cathet Cardiovasc Diagn* 1990;21:28–40
2. Levin DC, Fellows KE, Abrams HL: Hemodynamically significant primary anomalies of the coronary arteries. Angiographic aspects. *Circulation* 1978;58:25–34
3. Gillebert C, van Hoof R, van de Werf F, Piessens J, de Geest: Coronary artery fistulas in an adult population. *Eur Heart J* 1986;7: 437–443
4. Said SAM, Landman GHM: Coronary-pulmonary fistula: Long-term follow-up in operated and non-operated patients. *Int J Cardiol* 1990;27:203–210
5. Cheng TO, Adkins PC: Traumatic aneurysm of left anterior descending coronary artery with fistulous opening into left ventricle and left ventricular aneurysm after stab wound of chest. *Am J Cardiol* 1973;31:384–390
6. Rangel A, Badui E, Verduzco C, Valdespino A, Enciso R: Traumatic coronary arteriovenous fistula communicating the left main coronary artery to pulmonary artery, associated with pulmonary valvular insufficiency and endocarditis: Case report. *Angiology* 1990;41:156–160
7. Toda S, Nakamura A, Iwamoto T, Nakaji S: Successful surgical treatment of aortic regurgitation with coronary artery fistula due to blunt chest trauma—a case report. *J Jpn Assoc Thorac Surg* 1991; 39:1087–1092
8. Meng RL, Harlan JL: Left anterior descending coronary artery-right ventricle fistula complicating percutaneous transluminal angioplasty. *J Thorac Cardiovasc Surg* 1985; 90:387–390
9. Grill HP, Chew PH, Weiss JL, Merillat JC, Hill G, Cadden J, Brinker JA: Contrast echocardiographic diagnosis of PTCA-induced coronary artery-left ventricle fistula. *Am Heart J* 1991;121: 194–198
10. Iannone LA, Iannone DP: Iatrogenic left coronary artery fistula-to-left ventricle following PTCA: A previously unreported complication with nonsurgical management. *Am Heart J* 1990;120: 1215–1217
11. El-Omar MM, Hargreaves MR, Venkataraman A, Been M: Coronary ventricular fistula as a complication of PTCA: A case report and literature review. *Int J Cardiol* 1995;51:113–116
12. Yu R, Sharma B, Franciosa JA: Acquired coronary artery fistula to the left ventricle after acute myocardial infarction. *Am J Cardiol* 1986;58:557–558
13. Lee KMH, Pichard AD, Lindsay J: Acquired coronary artery-left ventricular pseudoaneurysm fistula after myocardial infarction. *Am J Cardiol* 1989;64:824–825
14. Ryan C, Gertz EW: Fistula from coronary arteries to left ventricle after myocardial infarction. *Br Heart J* 1977; 39:1147–1149
15. Shimizu M, Yoshio H, Ino H, Takeda R, Okada Y: Histopathologic findings of a left coronary artery fistula to the left ventricle after myocardial infarction. *Cardiology in the Elderly* 1993; 1:335–336
16. Somsen GA, Liem KS: Coronary artery-left ventricular fistulas probably due to transmural myocardial infarction: A case report. *Cardiologie* 1994;1:65–67
17. Saeian K, Vellinga T, Troup P, Wetherbee J: Coronary artery fistula formation secondary to permanent pacemaker placement. *Chest* 1991;99:780–781
18. Sandhu JS, Uretsky BF, Zerbe TR, Goldsmith AS, Reddy PS, Kormos RL, Griffith BP, Hardesty RL: Coronary artery fistula in the heart transplant patient: A potential complication of endomyocardial biopsy. *Circulation* 1989;79:350–356
19. Lee RT, Mudge GH, Colucci WS: Coronary artery fistula after mitral valve surgery. *Am Heart J* 1988;115:1128–1130
20. Reidy JF, Anjos RT, Qureshi SA, Baker EJ, Tynan MJ: Transcatheter embolization in the treatment of coronary artery fistula. *J Am Coll Cardiol* 1991;18:187–192
21. Pattee PL, Chambers RJ: Acquired postoperative coronary arteriovenous fistula. *Cathet Cardiovasc Diagn* 1992;26:140–142
22. Ray SG, Cowan MD, Kennedy JA: Spontaneous acquired fistula from the right coronary artery to the right ventricular cavity. *Br Heart J* 1992;67:323–324
23. Searcy RA, Stein PD, Gunesan G, Bruce TA: Arterio-atrial shunting in coronary atherosclerosis. *Chest* 1971;59:398–401

24. Karim MA: Coronary artery aneurysmal fistula: A late complication of stent deployment. *Int J Cardiol* 1996;57:207–209
25. Bata IR, MacDonald RG, O'Neill BJ: Coronary artery fistula as a complication of percutaneous transluminal coronary angioplasty. *Can J Cardiol* 1993;9:331–335
26. Bartorelli AL, Pepi M, Sganzerla P, Montorsi P, Loaldi A, Ravagnani P, Fabbicchi F: Syncope with cardiac arrest as the first manifestation of two congenital left coronary artery-to-main pulmonary artery fistulae. *Am Heart J* 1994;127:207–209
27. Ogino K, Hisatome I, Kotake H, Furuse T, Mashiba H, Kuroda H, Mori T: A case of four coronary artery fistulae originating from three vessels associated with aneurysm. *Eur Heart J* 1987;8:1260–1263
28. Abhyankar AD, Mok NS, Helprin GA, L Pressley: Multiple coronary-pulmonary fistulae involving all three coronary arteries: A case report. *Int J Cardiol* 1996;57:181–183
29. Ashraf SS, Shaukat N, Fisher M, Clarke B, Keenan DJ: Bicornary-pulmonary fistulae with coexistent mitral valve prolapse: A case report and literature review of coronary-pulmonary fistula. *Eur Heart J* 1994;15:571–574
30. Takahashi M, Sekiguchi H, Fujikawa H, Mito H, Eto M, Hojo Y, Chiba Y, Kitamura S, Shimada K: Multicystic aneurysmal dilatation of bilateral coronary artery fistula. *Cathet Cardiovasc Diagn* 1994;31:290–292
31. Farooki ZQ, Nowlen T, Hakimi M, Pinsky WW: Congenital coronary artery fistulae: A review of 18 cases with special emphasis on spontaneous closure. *Pediatr Cardiol* 1993;14:208–213
32. Shyam Sunder KR, Balakrishnan KG, Tharakan JA, Titus T, Pillai VRK, Francis B, Kumar A, Bhat A, Shankaran S: Coronary artery fistula in children and adults: A review of 25 cases with long-term observations. *Int J Cardiol* 1997;58:47–53
33. Perry SB, Rome J, Keane JF, Baim DS, Lock JE: Transcatheter closure of coronary artery fistulas. *J Am Coll Cardiol* 1992;20:205–209