

# Cardiovascular magnetic resonance and computed tomography in the evaluation of aneurysmal coronary-cameral fistula

Efstathios E Detorakis<sup>1\*</sup>, Emmanouil Foukarakis<sup>2</sup>, George Karavolias<sup>3</sup>, Alkiviades Dermitzakis<sup>2</sup>

1. Department of Radiology, CT and MRI unit, Euromedic International, Heraklion, Crete, Greece

2. Department of Cardiology, General Hospital of Heraklion "Venizeleio - Pananeio", Heraklion, Crete, Greece

3. Second department of interventional cardiology, Onassis cardiac surgery centre, Athens, Greece

\* **Correspondence:** Efstathios E. Detorakis, MD, 15 Iktinou str. 71305, Heraklion, Crete, Greece  
(✉ [edetorakis@hotmail.com](mailto:edetorakis@hotmail.com))

Radiology Case. 2015 Jul; 9(7):10-21 :: DOI: 10.3941/jrcr.v9i7.2305

## ABSTRACT

Coronary artery fistulas represent abnormal communications between a coronary artery and a major vessel like venae cavae, pulmonary arteries or veins, the coronary sinus, or a cardiac chamber. The latter is called coronary cameral fistula is a rare condition and is most of the times congenital but can be also post traumatic or post surgical, especially after cardiovascular interventional procedures. Most patients are asymptomatic and coronary-cameral fistulae are discovered incidentally during angiographic evaluation for coronary vascular disorders, while other patients have a clinical presentation ranging from angina pectoris to heart failure. In this article, we report a rare case of an aneurysmal right coronary cameral fistula draining into the left ventricle. Echocardiography usually represents the first diagnostic imaging approach, but often due to a poor acoustic window may not show the entire course of the fistula which is crucial for the final diagnosis. ECG-gated cardiovascular CT may play an important role in the evaluation of the origin, course, termination and morphology of the fistula, its relation to the adjacent anatomical structures as well as the morphology and contractility of the heart. Cardiac MRI instead plays an additional crucial role regarding not only the above mentioned factors but also in estimating the blood flow within the fistula, providing more detailed information about the cardiac function but also about myocardial wall viability.

## CASE REPORT

### CASE REPORT

A 42-year-old asymptomatic man was referred to our clinic for routine medical examinations. The patient was a nonsmoker, nondiabetic, normotensive with no family history of coronary artery disease.

Upon auscultation a continuous heart murmur was heard while the rest physical examination was unremarkable. Electrocardiogram (ECG) showed normal sinus rhythm without significant ST-T wave changes. Cardiac enzymes and routine laboratory index findings were also within normal limits.

### Imaging findings

A transthoracic echocardiogram with color Doppler interrogation revealed a large pulsatile vascular structure originating from the right coronary cusp (RCC), probably corresponding to the right coronary artery (RCA), with a tortuous course first along the right and consecutively the left atrio-ventricular groove, possibly terminating into the left ventricle (LV). Pulse wave-Doppler with sample volume at the entry site of coronary fistula in right coronary sinus, recorded a predominantly diastolic flow. The vascular structure was further evaluated by transesophageal echocardiogram that also verified the origin of the vascular structure from the right coronary sinus, its tortuous course and registered a draining diastolic jet into the left ventricle at the base of posterior wall with concomitant aortic valve regurgitation (Fig.1).

In order to further identify the vascular structure and to delineate its origin, course and draining site, as well as its relation with the coronary arteries further imaging investigation was followed first with cardiac computed tomography (Cardiac CT) and then with cardiac MRI (CMR).

An ECG-gated 64-MDCT (LightSpeed VCT 64-slice CT scanner, GE Healthcare, USA) scan was performed to better define the coronary anatomy. Scout view showed an obliterated right cardiophrenic angle of a slightly increased radiopacity comparing to the normal pericardial fat pad (Fig.2).

Nonionic contrast material iopromide (Ultravist® 370, Bayer HealthCare Pharmaceuticals, 80mL at 4.5 mL/s) was injected with bolus tracking over the ascending aorta. Retrospective ECG gated curved multiplanar reconstruction (MPR) and maximum intensity projection (MIP) images revealed a giant RCA aneurysm with a maximum diameter of 5.6cm originating from the right coronary sinus. Homogeneous contrast enhancement was detected along the course of the fistula as well as partially calcified atherosclerotic lesions of its aneurysmal wall (Fig.3A and B). After a serpiginous course first at the level of the right and then the left posterior atrioventricular groove the dilated RCA appeared to drain at the level of the left ventricle base, corresponding to an aneurysmal coronary-cameral fistula (CCF) (Fig.3 C). MIP images revealed thin collateral branches originating from the fistula draining the neighboring part of the epicardium of the right and left ventricle and the interventricular septum (Fig.3 D).

Volume rendering technique images (VRT) offered a three-dimensional overview of the origin, dimensions, course and termination of the CCF and its anatomic relation with the remaining epicardial vessels, the large vascular structures of the mediastinum and the cardiac chambers (Fig.4 A,B,C,D and Fig.5 A).

The left coronary artery (LCA) and its branches, including the posterior descending artery, were normal in origin, course, and caliber.

In order to have a more dynamic evaluation of the CCF, the patient also underwent ECG gated CMR imaging was performed in end-expiratory breath-hold period using a 1.5 T system (Signa CV/i, GE Medical Systems, Milwaukee, Wisconsin, USA) in line with recommendations of the Society of CMR (SCMR) and the European Society of Cardiology (ESC) CMR Working Group[1].

The imaging protocol included double inversion recovery (IR) black blood sequence in axial plane (Fig. 5B) and balanced-steady state free precession cine sequences (b-SSFP) in two, three and four-chamber view as well as in short-axis view (Fig.6). Subsequently, i.v administration of gadolinium (Gadovist® 0.1mmol/kg, Bayer) enhanced T1-weighted inversion recovery gradient echo (IR-GRE) sequences were acquired for first pass perfusion. Data acquisition for late gadolinium enhancement (LGE) started after 10 min in standardized apical, mid-cavity and basal short axis levels covering all AHA 17-segment model [2]. Inversion time (TI) was adapted to nullify the signal of myocardium (Fig.7). Cardiac volumes and left ventricle ejection fraction (LVEF) were calculated using the Signa® CV/i™ analysis software package (Signa® CV/i™; General Electric Medical Systems, USA).

All CMR sequences confirmed the presence of a giant aneurysmal coronary-cameral fistula originating from the right coronary sinus coursing along the posterior atrioventricular groove and finally draining into the base of the left ventricle (Fig.4 and 5). Cine cardiac imaging through b-SSFP sequences, especially in 4-chamber view revealed blood flow with turbulent flow areas throughout the fistula but most important its draining jet into LV, verifying the findings of the transesophageal echocardiogram (Fig. 6 A and B). Unfortunately, flow assessment of the fistula with phase - contrast velocity mapping was not diagnostic due to respiratory motion artifacts.

Right ventricular dimensions and function as well as thickness of right and left ventricular walls were within normality while mild dilatation of the LV and slightly compromised LV contractility with ejection fraction around 50% was registered. No perfusion defect was identified during first pass perfusion sequence and LGE sequences did not show any evidence of prior myocardial infarction or myocardial fibrosis of the LV wall (Fig. 7).

### Management plan

A selective coronary angiogram also confirmed the above findings of a giant aneurysm of the right coronary artery with a tortuous course, draining into the left ventricle (Fig. 8). The patient subsequently underwent surgery for closure of the fistula. The great saphenous vein graft was then anastomosed from the aortic root to the posterior descending artery and the acute marginal branch of the RCA. Right coronary artery giant aneurysm was excised and a 2.0 cm Dacron patch was used to close the orifice of RCA. The patient made a full recovery.

## DISCUSSION

*Etiology and Demographics*

Coronary artery anomalies include pathology of origination and course, anomalous collateral vessels as well as anomalies of coronary termination [3]. Coronary artery fistulae (CAF) are considered a rare major anomaly and represent abnormalities of termination, establishing a pathological communication between a coronary artery and any segment of the systemic or pulmonary circulation (coronary-vascular fistulae, CVF) or a coronary artery and a chamber of the heart, bypassing the myocardial capillary bed (coronary-cameral fistulae, CCF) [3,4].

Most of CAF are congenital but their exact incidence is unknown. They rarely may be acquired of traumatic iatrogenic (usually post-surgical), traumatic accidental (most from penetrating chest injury) and spontaneously occurring CCFs (Table 1).

CCFs have no gender predilection while most of them are congenital and discovered during childhood [5]. They can be divided into multiple micro-fistulas or small or large solitary macro fistulas, as in our case, arising from one or more coronary arteries and terminating into one of the four cardiac chambers. The acquired types of CCFs can also be single or multiple, arising from one or more coronary arteries entering into one of the four cardiac chambers [3]

The exact incidence of CCFs is unknown but according to literature CCFs are seen in 0.1% of patients undergoing coronary angiograms [6] (Table 2). The prevalence of these fistulas in adult patients is low and that of aneurysmal coronary fistulas is even lower because the majority of them are diagnosed and treated during childhood [7].

The RCA represents the most frequent site of origin of CCFs (55-65%), followed by the LCA (35%), while in a small percentage both coronary arteries may be involved (5%). Low pressure structures like the right cardiac chambers are the most common sites of CCFs drainage. Major draining sites are the right ventricle (40%), right atrium (26%), pulmonary arteries (17%) and less frequently the superior vena cava or coronary sinus and least often the cardiac atria [8,9,10].

RCA fistula terminating into a left heart chamber and especially the left ventricle is exceedingly rare with the incidence being reported as 1.2% of all CAF and to our knowledge only a few case reports have been published [4,11,12,13,14,15]. The association of a CCF with coronary artery aneurysm, as in our case, is considered even more rare [16].

*Clinical & Imaging findings*

The majority of coronary artery fistulas are asymptomatic in the early years and they may be occasionally detected later in life during angiographic procedures, after cardiac surgery, such as coronary artery bypass grafting, valve replacement, and after interventional manipulations like repeated myocardial biopsies [5,6,7,8]. The clinical features depend upon the size and location of these fistulas and range from

asymptomatic continuous murmur, to large left-to-right shunt and congestive heart failure, myocardial ischemia distal to the fistula due to a "myocardial steal" or even rupture or thrombosis of the fistula. Hence, larger fistulas may result in congestive cardiac failure or angina pectoris at the extremes of life, in infants or older adults [17].

A two-dimensional echocardiography with color Doppler interrogation usually represents the first diagnostic approach in the diagnosis of a coronary fistula, revealing an abnormal vascular communication regarding a coronary artery. Unfortunately, echocardiography is an operator dependent technique and very often imaging quality may have limitations imposed due to the poor acoustic window quality and diagnosis of CAF and especially CCF may be extremely difficult or even impossible since it may not show the entire course of the fistula.

Even though cardiac catheterization with coronary angiography remains the gold standard for the diagnosis of CAFs, is an invasive technique, but it only shows an intraluminal approach of the lesion and may hamper a complete evaluation by overlapping between a tortuous fistula and adjacent cardiovascular structures [18].

Nowadays more sophisticated and non-invasive techniques such as MDCT and CMR may play an important role in the diagnosis of these vascular anomalies. Both techniques nicely demonstrate the origin, course, size and termination site of a CCF and its relation with the adjacent anatomical structures.

ECG-gated cardiac CT offers a significant improvement of temporal resolution and with MPR and MIP reconstructions may show the exact origin of the fistula, providing with in detail measurement of its dimensions and course but also its anatomic relation with the adjacent vascular structures and the cardiac chambers as well (Fig.2 and 3).

In case of aneurysmal CAF cardiac CT is important in the evaluation of its walls, demonstrating the wall thickness and morphology, especially in case of atherosclerotic lesions or eventual mural thrombus. VRT image reconstructions create an impressive overview of the fistula and its course which may be extremely useful for surgical planning (Fig. 3).

CMR imaging apart from evaluating the anatomy of the fistula, may further measure the blood flow within its lumen. Phase contrast velocity mapping of flow through planes transecting the fistula should also provide an accurate measurements of cardiac output, shunt flow, turbulent flow jet areas or even regurgitation [19].

CCFs may cause volume overloading of the draining heart chamber and heart failure. Evaluation of cardiac anatomy may be done with double IR black blood and cine SSFP sequences by measuring cardiac dimensions including chamber volumes (left ventricle end diastolic volume, LVEDV, left ventricle end systolic volume, LVESV, ml), myocardial mass (left ventricular mass index, g/m<sup>2</sup>) and wall thickness, but also cardiac function (LV and RVEF, stroke volume). In our case,

even though the patient was asymptomatic, CMR with b-SSFP cine sequences showed a mild dilatation of the LV as well as a slightly compromised left ventricular function with LVEF around 50%. These findings are related to the increased chronic volume overload of the LV caused by the fistula. Other possible CMR findings related to chronic volume overload, not depicted in our case, may be an increased thickness of LV myocardial walls.

In some cases, fistula, due to coronary artery steal phenomenon, leads to ischemia of the segment of the myocardium perfused by the coronary artery [20]. Important additional information about the myocardial wall status are given by the LGE sequences. Late gadolinium enhancement CMR sequences offer the capacity to quantify the extent of myocardial fibrosis that represents a pathophysiological end point related to most cardiomyopathies related to ischemia or chronic volume overload. The presence of LGE areas at the level of the myocardial wall, regarding only the epicardium, myocardium or endocardium or even with a transmural distribution have been related to a poorer patient's prognosis, so it is of crucial importance to identify or to exclude their existence [21].

Even though our patient didn't present LGE areas within the myocardial wall and made a full recovery, the mild LV dilatation combined with a slight decrease of the LVEF suggest a closer clinical follow-up.

#### Treatment & Prognosis

There is general agreement that symptomatic patients and those with a large diameter CAF, being symptomatic or not, should be treated with closure of the fistula by either surgical or transcatheter approaches. Untreated larger fistulas may predispose to congestive heart failure and premature coronary artery disease in the affected vessel [22]. The current options include closure of the fistula by surgical ligation alone either with or without cardiopulmonary bypass or surgical ligation accompanied by coronary artery bypass surgery (by using a venous graft as in our case) in cases of large caliber fistulas or transcatheter closure of small caliber fistulas with devices, including detachable balloons, stainless steel coils, controlled-release coils and Amplatzer PDA plug [21,22,23]. Transcatheter closure of these fistulas has an excellent outcome with lesser morbidity and mortality, lower cost, shorter recovery time, and avoidance of thoracotomy and cardiopulmonary bypass [22,23]. Patients with a low caliber CCF and without established congestive heart failure or coronary artery disease may have an excellent prognosis.

#### Differential Diagnoses

The differential diagnosis of a CCF include a coronary-vascular fistula, a coronary artery aneurysm in case of a large caliber fistula or even a patent ductus arteriosus (PDA) in case of a small caliber vascular structure. In case of a coronary-vascular aneurysm it is important to show the origin and the termination of the fistula into a major vessel that can be achieved with an echocardiographic approach with echo color Doppler and can be further verified by cardiac CT and CMR. The case of a coronary artery aneurysm can be easily ruled out with cardiac CT by excluding a communication with a cardiac

chamber or another vascular structure. In suspected cases of PDA echocardiography with echo color Doppler may identify the abnormal communication between the aortic arch and the pulmonary artery circulation while ECG-gated contrast enhanced cardiac CT may further exclude any involvement of the coronary arteries and establish the presence, patency, caliber and length of the PDA.

#### TEACHING POINT

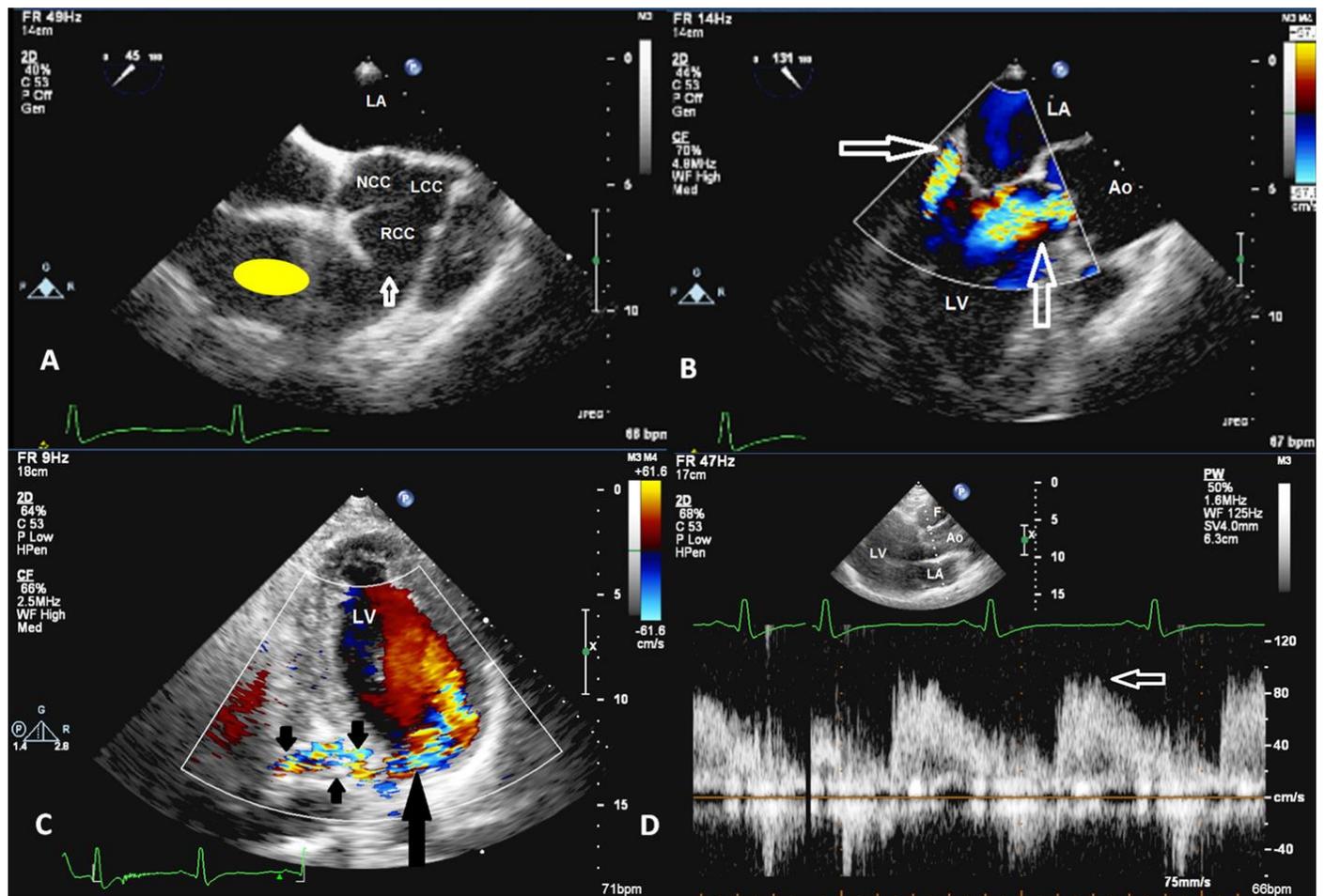
Cardiac computed tomography may play an important role in the diagnosis of a coronary-cameral fistula with multiplanar reconstructions and maximum intensity projection image reformats detecting its origin, course, morphology and termination but also its relation with the adjacent anatomic structures, while volume rendering technique images offer an impressive overview of the fistula that would be helpful for surgical planning. With cardiovascular magnetic resonance we can measure the blood flow volume within the coronary-cameral fistula's lumen and have additional detailed information about the cardiac function and morphology of the cardiac chambers as well as the myocardial wall viability that may be of crucial importance for the prognosis of these patients.

#### REFERENCES

- Schulz-Menger J, Bluemke DA, Bremerich J, et al. Standardized image interpretation and post processing in cardiovascular magnetic resonance: Society for Cardiovascular Magnetic Resonance (SCMR) board of trustees task force on standardized post processing. *J Cardiovasc Magn Reson.* 2013 May 1;15:35. doi: 10.1186/1532-429X-15-35. PMID: 23634753
- Cerqueira MD, Weissman NJ, Dilsizian V, et al. Standardized myocardial segmentation and nomenclature for tomographic imaging of the heart. A statement for healthcare professionals from the Cardiac Imaging Committee of the Council on Clinical Cardiology of the American Heart Association. *J Cardiovasc Magn Reson.* 2002;4:203-10. doi:10.1081/JCMR-120003946. PMID: 12135124
- Angelini P. Coronary artery anomalies--current clinical issues: definitions, classification, incidence, clinical relevance, and treatment guidelines. *Tex Heart Inst J.* 2002;29(4):271-8. PMID:12484611
- Said SA, Schiphorst RH, Derksen R, et al. Coronary-cameral fistulas in adults (first of two parts). *World J Cardiol.* 2013 Sep 26;5(9):329-36. doi: 10.4330/wjc.v5.i9.329. Review. PMID: 12135124
- Padfield GJ. A case of coronary cameral fistula. *Eur J Echocardiogr.* 2009 Jul;10(5):718-20. doi: 10.1093/ejechocard/jep049. Epub 2009 May 4. PMID: 19414488

6. Thomas M, Davis RC. Asymptomatic extensive coronary cameral fistulae involving the left ventricle. *Can J Cardiol.* 2008 Jul;24(7):e46. PMID: 18612509
7. Jagia P, Goswami KC, Sharma S, et al. 16-MDCT in the evaluation of coronary cameral fistula. *AJR Am J Roentgenol.* 2006 Aug;187(2):W227-8. PMID: 16861516
8. Yuksel S, Yasar E, Nar G, et al. Prevalence and characteristics of coronary-cameral communications in adult patients: Coronary Angiographic Analysis of 16,573 Patients. *Med Princ Pract.* 2014 Jun 7. [Epub ahead of print]. PMID: 24924735
9. Ho HH, Cheung CW, Jim MH, et al. Images in cardiology: coronary-cameral fistula. *Heart.* 2005 Dec;91(12):1540. PMID: 16287738
10. Sommer RJ, Hijazi ZM, Rhodes JF Jr. Pathophysiology of congenital heart disease in the adult: Part I: Shunt lesions. *Circulation* 2008;117(8):1090-9. PMID: 18299514
11. Arani DT, Greene DG, Klocke FJ. Coronary artery fistulas emptying into left heart chambers. *Am Heart J.* 1978 Oct;96(4):438-43. PMID: 696564
12. Araki H, Kageyama M, Inami S, et al. Right coronary artery to left ventricle fistula associated with three-vessel coronary artery disease: a case report. *J Cardiol* 50: 193-197, 2007 (in Japanese). PMID: 17941195
13. Funabashi N, Asano M, Komuro I. Right coronary artery aneurysm with fistula to left ventricle: multislice CT appearance. *J Thorac Imaging* 21: 63-65, 2006. PMID: 17941195
14. Feuchtner G, Junker D, Bonatti J, Friedrich G. Right coronary artery fistula into left ventricle: dynamic compression shown by multislice computed tomography. *Eur J Cardiothorac Surg* 32: 933, 2007. PMID: 17904377
15. Oncel D, Oncel G. Right coronary artery to left ventricle fistula effective diagnosis with 64-MDCT. *Int J Cardiovasc Imaging* 23:287-291, 2007. PMID: 16924400
16. Nakamura Y, Yutani C, Imakita M, et al. A huge coronary aneurysm resulting from a coronary artery-to-left ventricle fistula. *Intern Med* 37: 366-369, 1998. PMID:9630195
17. Sharland GK, Tynan M, Qureshi SA: Prenatal detection and progression of right coronary artery to right ventricle fistula. *Heart* 1996, 76:79-81. PMID: 8774334
18. Jamil G, Khan A, Malik A, Qureshi A. Aneurysmal coronary cameral fistula. *BMJ Case Rep.* 2013 Jun 3;2013. pii: bcr2013008649. doi: 10.1136/bcr-2013-008649. PMID:23737570
19. Kilner PJ, Gatehouse PD, Firmin DN. Flow measurement by magnetic resonance: a unique asset worth optimising. *J Cardiovasc Magn Reson.* 2007;9(4):723-8. PMID: 17613655
20. Kurt IH. Myocardial infarction caused by a fistula between the left anterior descending coronary artery and the left ventricle. *Tex Heart Inst J.* 2009;36(2):177-9. PMID: 19436820
21. Stirrat J, White JA. The prognostic role of late gadolinium enhancement magnetic resonance imaging in patients with cardiomyopathy. *Can J Cardiol.* 2013 Mar;29(3):329-36. doi: 10.1016/j.cjca.2012.11.033. PMID:23439019
22. Latson LA. Coronary artery fistulas: how to manage them. *Catheter Cardiovasc Interv.* 2007; 70: 110-116. PMID: 17420995
23. Vijayalakshmi IB, Narasimhan C, Agarwal A. Transcatheter closure of left coronary cameral fistula with Amplatzer duct occluder II. *J Invasive Cardiol.* 2013 May;25(5):265-7. PMID: 23645054

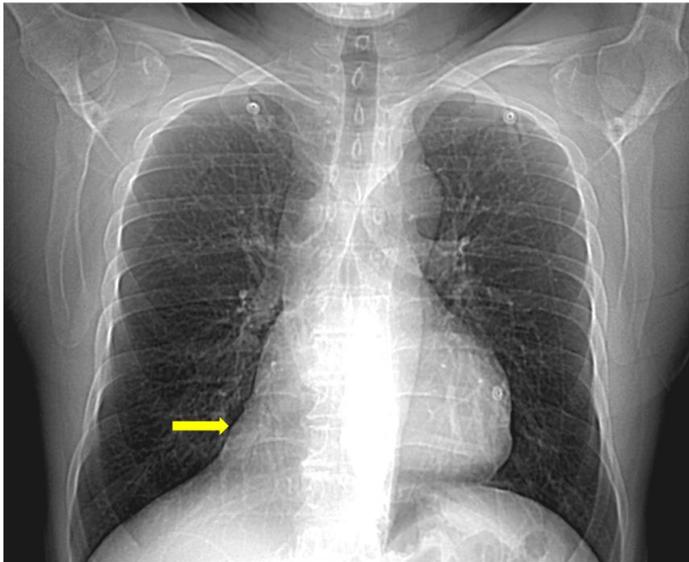
FIGURES



**Figure 1:** 42 year old male with aneurysmal coronary-cameral fistula.

Findings: Transesophageal echocardiogram in a midesophageal aortic valve short-axis view (A) revealing a large pulsatile structure (yellow ovoid sign) originating from the right coronary sinus (empty arrow) probably corresponding to the RCA. Transesophageal color Doppler image in aortic valve long axis view (B) showing simultaneously the aortic valve regurgitation jet (vertical arrow) and the draining jet of coronary fistula into left ventricle at the base of posterior wall (horizontal arrow). Transthoracic color Doppler image in a modified apical view (C) indicating the possible tortuous course of the vascular structure along the atrio-ventricular groove (small black arrows) and consecutively the draining site into the left ventricle (big black arrow). Pulse wave-Doppler in a transthoracic long axis view (D) with sample volume at the entry site of coronary fistula in right coronary sinus, recording the predominantly diastolic flow (empty arrow).

NCC= non coronary cusp, LCC= left coronary cusp, RCC= right coronary cusp, LA= left atrium, LV= left ventricle, Ao= aorta  
 Technique: The ultrasound study was performed using a Philips iE 33 Matrix (Royal Philips Electronics, Eindhoven, Netherlands) with a transthoracic S5-1sector array transducer (1-5MHz) and a transesophageal multiplane S7-2t omni sector array transducer (2-7MHz).



**Figure 2 (left):** 42 year old male with aneurysmal coronary-cameral fistula.

Findings: Scout view from ECG-gated cardiovascular CT, shows obliteration of the right cardiophrenic angle which seems slightly more radiopaque than the common radiopacity of the pericardial fat pad (yellow arrow) corresponding to the anatomic location of the vascular structure figured at the echocardiographic study.

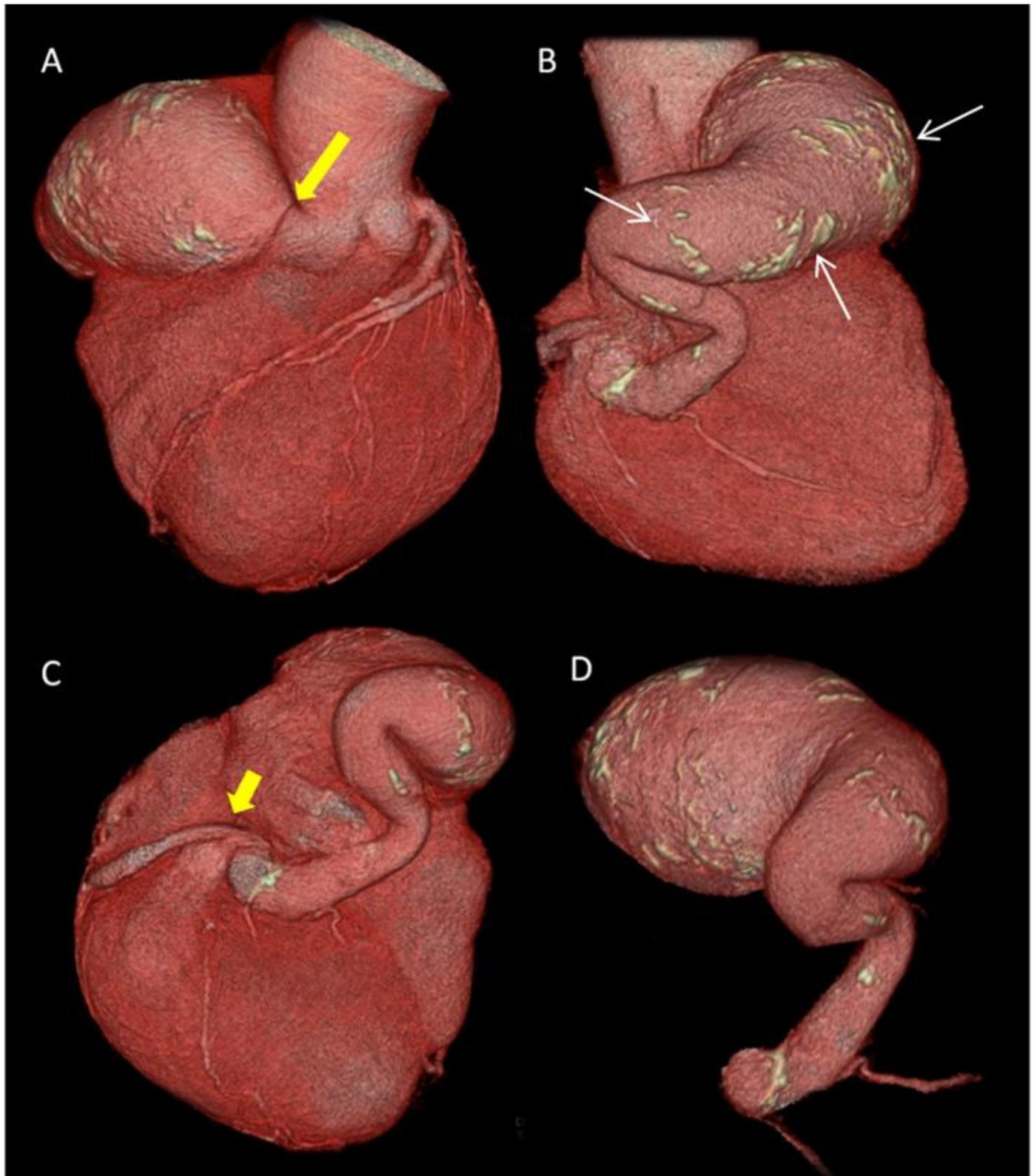
Technique: ECG-gated 64-MDCT, (LightSpeed VCT 64-slice CT scanner, GE Healthcare, USA, 100 mAs, 120 kV, slice thickness, 0.625 mm, rotation time 0.40 second).



**Figure 3:** 42 year old male with aneurysmal coronary-cameral fistula.

Findings: ECG-gated cardiovascular CT, mediastinal window, width/level of 350/30 HU, in axial plane showing in (A) the origin of the CCF from the right coronary sinus (black round sign) and in (B) the giant aneurysmal dilatation of its proximal portion (black asterisk). Multiplanar reconstruction image in (C) shows the course and tortuosity of the fistula until its termination at the level of the LV base (yellow arrow). Maximum intensity projection image in (D) showing calcified atherosclerotic lesions along the fistula's wall and thin collateral branches originating from the distal part of the fistula draining the adjacent epicardium (thin white arrows).

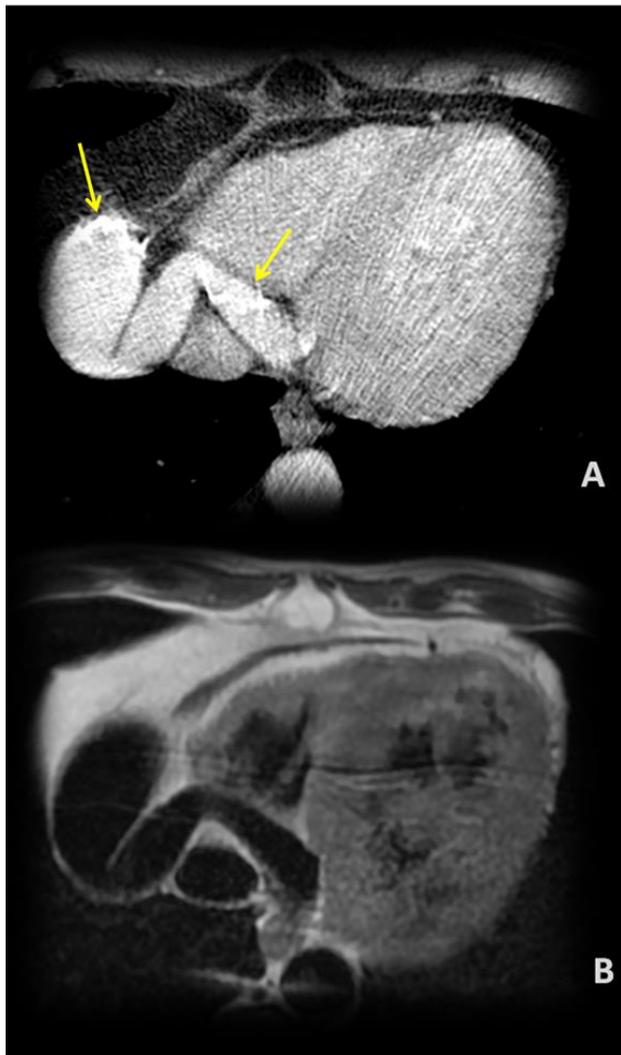
Technique: ECG-gated 64-MDCT, (LightSpeed VCT 64-slice CT scanner, GE Healthcare, USA, 100 mAs, 120 kV, slice thickness, 0.625 mm, rotation time 0.40 second, scanning time 10 seconds, 1.15 pitch, heart rate of 55 bpm) after i.v administration of contrast material (Ultravist 370, 80mL at 4.5 mL/s).



**Figure 4:** 42 year old male with aneurysmal coronary-cameral fistula.

Findings: ECG-gated cardiovascular CT, Volume rendering technique image reconstruction showing an overview of the coronary-cameral fistula from its origin at the level of the right coronary sinus (yellow arrow) (A), the giant aneurysmal dilatation of its proximal portion with atherosclerotic wall lesions giving a coarse appearance of its wall (multiple thin white arrows) (B), and the tortuous course followed along the posterior atrio-ventricular groove before draining into the left ventricle (short yellow arrow) (C). Isolated image of the CCF showing thin collateral branches at its distal portion, directed to the epicardium (D).

Technique: ECG-gated 64-MDCT, (LightSpeed VCT 64-slice CT scanner, GE Healthcare, USA, 100 mAs, 120 kV, slice thickness, 0.625 mm, rotation time 0.40 second, scanning time 10 seconds, 1.15 pitch, heart rate of 55 bpm) after i.v administration of contrast material (Ultravist 370, 80mL at 4.5 mL/s).



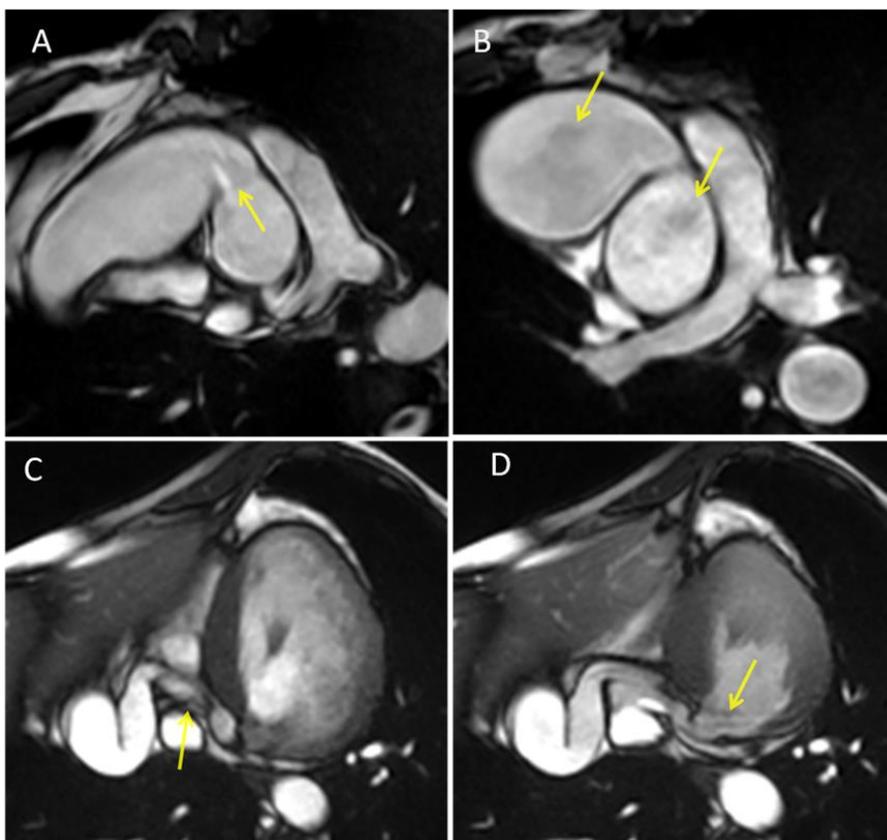
**Figure 5 (top left):** 42 year old male with aneurysmal coronary-cameral fistula.

Findings: (A) ECG-gated cardiovascular CT, mediastinal window, width/level of 350/30 HU, in axial plane showing the course of the fistula and the calcified atherosclerotic lesions on its wall (yellow arrows).

Technique: ECG-gated 64-MDCT, (LightSpeed VCT 64-slice CT scanner, GE Healthcare, USA, 100 mAs, 120 kV, slice thickness, 0.625 mm, rotation time 0.40 second, scanning time 10 seconds, 1.15 pitch, heart rate of 55 bpm) after i.v administration of contrast media (Ultravist 370, 80mL at 4.5 mL/s).

Findings: (B) In comparison with (A), ECG-gated cardiovascular MRI, T1- weighted turbo fast spin echo "black blood" sequence, almost at the same plane with (A), shows the fistula course and its relation with the adjacent anatomic structures.

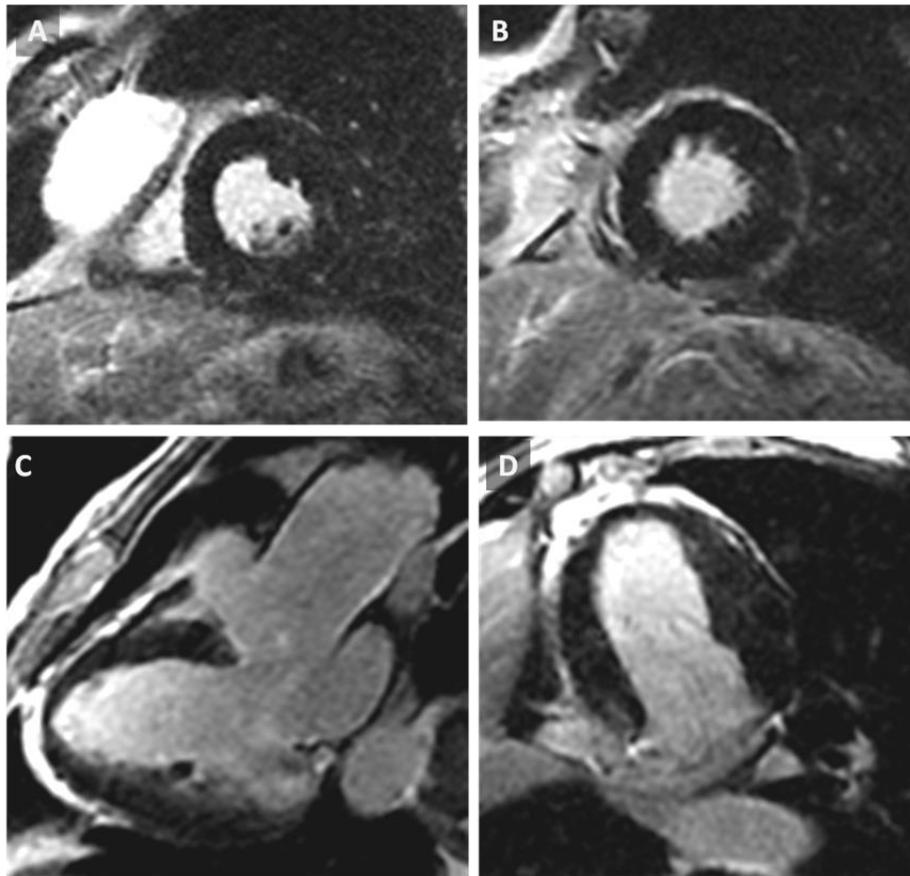
Technique: ECG-gated CMR, 1.5 T system Signa CV/i, GE Medical Systems, USA, with a 5-element cardiac phased-array coil, FOV: 340-400 mm; TR/TE: 6.0/3.8ms, matrix: 256x192; ST: 8 mm



**Figure 6 (bottom left):** 42 year old male with aneurysmal coronary-cameral fistula.

Findings: ECG-gated cardiovascular MRI. Cine SSFP sequences on axial oblique view showing (A) the turbulent jet flow at its origin at the level of the right coronary sinus and (B) at the proximal aneurysmal portion as well (yellow arrows). End-diastole image (C) showing the course of the fistula and the turbulent flow within its lumen (yellow arrow). End-systole image at the same level (D) indicating the draining of the fistula into the left ventricle base with a flow jet (yellow arrow).

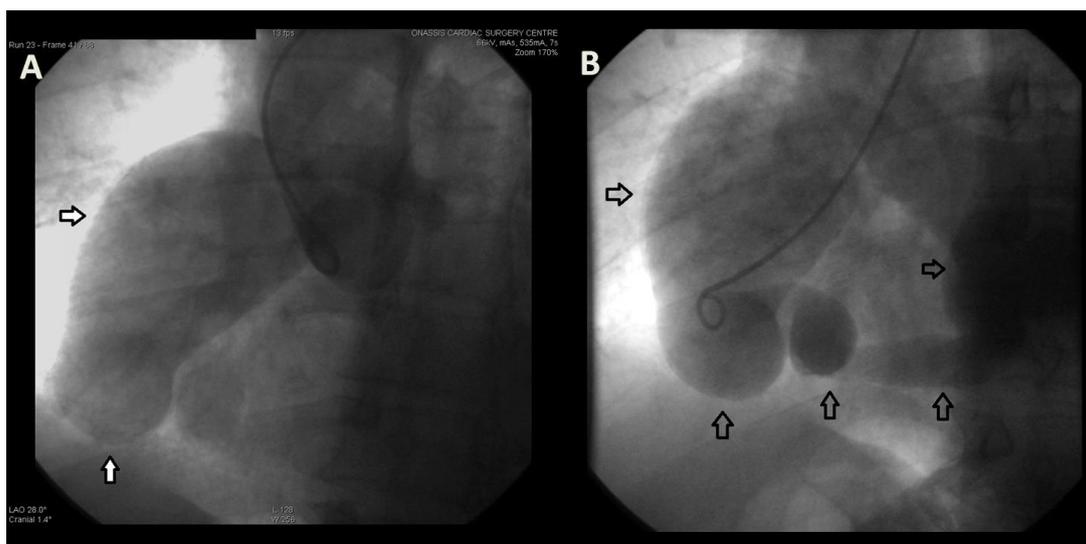
Technique: ECG-gated CMR, 1.5 T system Signa CV/i, GE Medical Systems, USA, with a 5-element cardiac phased- array coil, TR 3.5 ms, TE 1.7 ms, FA 55°, slice thickness 8 mm, in-plane resolution 1.2x1.8 mm.



**Figure 7:** 42 year old male with aneurysmal coronary-cameral fistula.

Findings: ECG-gated cardiovascular MRI. Contrast enhanced T1-weighted inversion recovery gradient echo (IR-GRE) sequences were acquired for late gadolinium enhancement. Data acquisition started 10min after Gadobutrol administration in short axis view (A and B), 3-chamber view (C) as well as 4-chamber view (D) showing no late gadolinium enhancement areas, excluding the presence of myocardial lesions that could be attributed to ischemia or volume overload related to the coronary-cameral fistula.

Technique: ECG-gated CMR, 1.5 T system Signa CV/i, GE Medical Systems, USA, with a 5-element cardiac phased- array coil. Contrast enhanced T1-weighted inversion recovery gradient echo (IR-GRE) sequences FOV: 340-400 mm; TR/TE: 6.0/3.8ms, ? : 25°; matrix: 256x192; ST: 8 mm, i.v administration of Gadobutrol (Gadovist®, Bayer) at a dose of 0.1mmol/kg. Nulling of the myocardium signal was achieved using a time to invert (TI) range between 220 and 300 ms.



**Figure 8:** 42 year old male with aneurysmal coronary-cameral fistula.

Findings: Left anterior oblique projection (A): Aortography of the ascending aorta with a Pigtail catheter in the aortic root. The giant right coronary artery aneurysm is seen (white arrows). Left anterior oblique projection (B): Angiogram of the right coronary artery with a Pigtail catheter inside the aneurysm of the right coronary artery. The whole course of the giant right coronary artery fistula is seen (empty arrows).

Technique: Angiographic system Philips, Integris Allura FD system, 66KV, 535mA

	<b>Classification of coronary artery anomalies</b>
<b>Anatomic location</b>	<ul style="list-style-type: none"> <li>• Origin</li> <li>• Course</li> <li>• Anomalous collateral vessels</li> <li>• Termination                             <ul style="list-style-type: none"> <li>- Coronary-vascular fistulae (CVF) (communication between a coronary artery and a major vessel)</li> <li>- Coronary-cameral fistulae (CCF) (communication between a coronary artery and a cardiac chamber)</li> </ul> </li> </ul>
<b>Etiology</b>	<ul style="list-style-type: none"> <li>• Congenital</li> <li>• Acquired                             <ul style="list-style-type: none"> <li>- Post-surgical</li> <li>- Post traumatic</li> </ul> </li> </ul>

**Table 1:** Classification of coronary artery anomalies<sup>3,4,5,6,7,8,9</sup>

<b>Etiology</b>	Most CCFs are congenital but they may be also acquired (post-surgical, post traumatic).
<b>Incidence</b>	The exact incidence is unknown but according to literature CCFs are seen in 0.1% of patients undergoing coronary angiograms.
<b>Gender ratio</b>	There is no gender predilection.
<b>Age predilection</b>	The most common CCFs are congenital and discovered during childhood.
<b>Risk factors</b>	Only in acquired cases, risk factors may be previous cardiac surgery / interventional manipulations and chest trauma
<b>Treatment</b>	<p>In less complicated cases</p> <ul style="list-style-type: none"> <li>• A transcatheter approach would be appropriate.</li> </ul> <p>In more complicated cases (i.e. multiple communications)</p> <ul style="list-style-type: none"> <li>• Surgical repair is followed                             <ul style="list-style-type: none"> <li>- Identifying the fistula, and closing the site of drainage with a patch or suture.</li> <li>- If the fistula enters the ventricle or if the feeding vessel is large, the coronary artery is opened, and the opening to the fistula is closed with a running suture. The arteriotomy is closed.</li> <li>- Large aneurysms, as in our case, may require excision and coronary artery bypass with the use of a venous graft.</li> </ul> </li> </ul>
<b>Prognosis</b>	Depends on the type of CCF and the cardiac conditions. There is usually a complete recover when discovered during childhood and repaired, surgically or even with a transcatheter approach.
<b>Findings on imaging</b>	<ul style="list-style-type: none"> <li>• An anomalous vascular structure may be initially detected on 2-D echocardiographic approach with echo color doppler use</li> <li>• Further imaging approach may be with cardiac CT and MRI, revealing in detail the origin, course and termination of the fistula.</li> </ul>

**Table 2:** Summary table of coronary-cameral fistulas

Entity	Echocardiography	Cardiac CT	Cardiac MRI
<b>Coronary-cameral fistula (CCF)</b>	May define (if the acoustic window is appropriate) the <ul style="list-style-type: none"> <li>dilatation of the affected coronary artery</li> <li>origin and termination into a cardiac chamber</li> <li>blood flow within fistula's lumen (pulse wave and color Doppler) and systolic and diastolic cardiac function</li> </ul>	Defines the <ul style="list-style-type: none"> <li>origin, course and termination of the fistula into a cardiac chamber</li> <li>fistula wall and analysis of probable collateral branches can be done with image reconstructions (MIP, MPR and VRT reformats)</li> </ul>	Defines the <ul style="list-style-type: none"> <li>origin, course and termination of the fistula into a cardiac chamber as well the blood flow within its lumen</li> <li>cardiac function and morphology (b-SSFP) and myocardial wall condition (LGE)</li> </ul>
<b>Coronary-vascular fistula (CVF)</b>	May define (if the acoustic window is appropriate) the <ul style="list-style-type: none"> <li>dilatation of the affected coronary artery</li> <li>origin and termination into a major vessel (flow within the fistula and detect the pathologic shunt (echo color Doppler))</li> </ul>	Defines <ul style="list-style-type: none"> <li>origin, course and termination of the fistula into a major vessel</li> <li>Fistula wall as well as that of the receiving major vessel can be studied with image reconstructions (MIP, MPR and VRT reformats)</li> </ul>	Defines <ul style="list-style-type: none"> <li>origin, course and termination of the fistula into a major vessel</li> <li>blood flow within its lumen and cardiac function and myocardium condition (b-SSFP, LGE)</li> </ul>
<b>Coronary artery aneurysm</b>	May define (if the acoustic window is appropriate) the <ul style="list-style-type: none"> <li>dilatation of the affected coronary artery alone without anomalous termination</li> </ul>	Defines the <ul style="list-style-type: none"> <li>origin and course of the dilated coronary artery, excluding an anomalous communication with any adjacent vessel or cardiac structure</li> </ul>	May define the <ul style="list-style-type: none"> <li>origin and course of such an aneurysm may be identified with CMR excluding other coronary artery anomalies.</li> </ul>
<b>Patent ductus arteriosus</b>	Echo pulse wave and color Doppler may <ul style="list-style-type: none"> <li>identify the abnormal communication between the aortic arch and pulmonary artery</li> </ul>	ECG-gated cardiac CT may <ul style="list-style-type: none"> <li>clearly identify the PDA presence and patency</li> <li>measure its diameter and length and exclude other kind of anomalous vessel communication</li> </ul>	CMR may <ul style="list-style-type: none"> <li>detect PDA (b-SSFP)</li> <li>measure the blood flow within with phase contrast velocity mapping, excluding other anomalous vascular communications</li> </ul>

**Table 3:** Differential diagnosis table for coronary-cameral fistulae

**ABBREVIATIONS**

- b-SSFP = balanced steady state free precession cine sequence
- CAF = coronary artery fistula
- Cardiac CT = cardiac computed tomography
- CCF = coronary-cameral fistula
- CMR = cardiac magnetic resonance
- CVF = coronary-vascular fistula
- ECG = electrocardiogram
- IR = inversion recovery sequence
- LCA = left coronary artery
- LGE = late gadolinium enhancement
- LV = left ventricle
- LVEDV = left ventricle end diastolic volume
- LVEF = left ventricle ejection fraction
- LVESV = left ventricle end systolic volume
- MIP = maximum intensity projection
- MPR = multiplanar reconstruction images
- PDA = patent ductus arteriosus
- RCA = right coronary artery
- RCC = right coronary cusp
- TI = time to invert
- VRT = volume rendering technique

**KEYWORDS**

Coronary artery fistula; Coronary-cameral fistula; cardiovascular CT; image reformats; cardiovascular MR; late gadolinium enhancement

**Online access**

This publication is online available at:  
[www.radiologycases.com/index.php/radiologycases/article/view/2305](http://www.radiologycases.com/index.php/radiologycases/article/view/2305)

**Peer discussion**

Discuss this manuscript in our protected discussion forum at:  
[www.radiolopolis.com/forums/JRCR](http://www.radiolopolis.com/forums/JRCR)

**Interactivity**

This publication is available as an interactive article with scroll, window/level, magnify and more features.  
 Available online at [www.RadiologyCases.com](http://www.RadiologyCases.com)

Published by EduRad



[www.EduRad.org](http://www.EduRad.org)