

Post Closure Evaluation of Coronary Artery Fistulae

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Abstracts: Coronary artery fistula (CAF) is a rare congenital anomaly in which a communication is present between a coronary artery and cardiac chamber. Conventional coronary angiography is an invasive and expensive procedure, and cannot provide three-dimensional data. ECG gated multidetector CT (MDCT) is useful for non-invasive evaluation of coronary artery fistulae. **Objectives:** In this paper, we describe ten post-closure cases of coronary artery fistulae. The aim of this study is to demonstrate the effectiveness of 128 slice-MDCT in demonstrating the precise anatomy and post closure follow-up of coronary artery fistula. **Methods:** 10 pediatric subjects underwent MDCT as well as conventional coronary angiography. Good quality images were obtained in all patients to analyze the coronary artery fistulae and their anatomic course. **Results:** MDCT not only provided precise details of post closure CAF but also clearly delineated anatomy of other coronary arteries. Catheter-related risks like bleeding at puncture site, hematoma formation were completely eliminated. **Advances in Knowledge:** MDCT may provide additional precise details of CAF. It also eliminates minor as well as more serious complications of catheter angiography and a small but definite incidence of mortality. The procedure is carried out as an outpatient procedure. Thus, MDCT is considered as a good alternative to echocardiography and coronary angiography for diagnosis as well as follow-up of CAF. [Patel D NJIRM 2014; 5(5):82-89]

Key Words: MDCT, Coronary Artery Fistulae, Follow-up

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Introduction: Coronary artery fistulae (CAF) are anomalous terminations of the coronary arteries. Many patients are asymptomatic; however, an awareness of the fistulas is important because they have been associated with various clinical features, including chest pain or heart failure in young patients. Correct diagnosis and follow-up of coronary artery fistula is important: - an early correction is indicated because of the high prevalence of late symptoms and complications. The number of incidentally found coronary artery fistulae has been increasing with more frequent use of multidetector computed tomography (MDCT) in chest and cardiac imaging. A special attention should be paid to the courses and terminations of coronary arteries to detect these potentially fatal anomalies in every CT study of the heart.

CAF was described in 1865 by Krause¹. It can be congenital or acquired. They account for approximately 0.2%–0.4% of congenital cardiac anomalies². There is no race or sex predilection for CAF.

Coronary artery fistulae are often caused by aberrancies of normal embryological development. Major sites of origin of the fistulae are from the right coronary artery (40-60%), left anterior

descending (30-60%), circumflex and a combination thereof¹¹. CAF predominantly drain into the right side of the heart (92%) into the right ventricle in 41%, the right atrium in 26%, the coronary sinus in 7%, the pulmonary in 17%, and superior vena cava in 1% of cases¹⁰. Its connection between coronary sinus; left atrium, and left ventricle is unusual.

The resultant physiologic derangement depends upon the site of origin, termination of the fistula and the size of the connection. The current study demonstrates that the anatomical features of CAF can be reliably established by MDCT, as opposed to conventional methods such as coronary angiography.

In this article, we present the pathophysiology, clinical features, imaging diagnosis, and treatment of different types of CAF - with special emphasis on follow-up after fistula closure with invasive and non-invasive modalities.

Materials and Methods: We report comparison of conventional versus MDCT coronary angiography in follow-up after closure of coronary artery fistulae in ten subjects. This study protocol was approved by the Institutional review board of our hospital.

Written informed consent was obtained from all the children’s parents.

MDCT Scanning Protocol: A retrospective ECG-triggered CT coronary angiography scan was performed on a 128 slice SOMATOM Definition AS+; (Siemens Healthcare, Forchheim, Germany). Sedation was achieved by administering oral chloral hydrate/ intravenous midazolam and ketamine according to the patient’s body weight and clinical condition as and when required. The scan range extended from the carina to the diaphragm. Tube voltage was 80kV, tube current was 100-120 mAs, pitch was 0.2 – 0.4. The CT data acquisition duration was 2-3 s. CT angiography acquisition with an automated bolus tracking technique was initialised when enhancement in the ascending aorta exceeded 100 HU.

The non-ionic iodinated contrast material (350mg /ml; Omnipaque (Iohexol); GE Healthcare, Shanghai, China) was administered with a dual-chamber mechanical power injector (Medtron, Germany) via a 22 or 20-gauge cannula inserted into an antecubital vein. The contrast material dose was tailored to the individual’s body weight (1.5ml /kg). After contrast injection, a given mass of saline chaser (half that of the contrast material) was administered at the same rate as the contrast material. Image reconstruction and reformation CT images were reconstructed

Conventional Angiography Protocol: Conventional angiography was performed using Philips AlluraXper FD10 cardiovascular X-ray system via the right femoral approach using 5F diagnostic catheters and non-ionic iodinated contrast material (350mg /ml; Omnipaque (Iohexol); GE Healthcare, Shanghai, China). The contrast material dose was tailored to the individual’s body weight. Sedation was achieved by administering intravenous midazolam and ketamine according to the patient’s body weight and clinical condition as and when required. Good quality images were obtained in all patients to analyze the coronary arteries and their anatomic course.

Subjects: 10 children [5 males; range 3 years to 15 years; 5 females; range 7 years to 17 years] with known case of coronary artery fistula and post

closure underwent MDCT coronary angiography and Conventional angiography or only MDCT. All children underwent routine blood investigations like serum Creatinine, HIV, HBsAg etc.

Results: MDCT and conventional coronary angiography findings were as described in Table 1.

Table 1: Comparison of MDCT and Conventional Coronary Angiography Findings

No.	Age	Sex	MDCT findings	Conventional angiography findings
1,	7 y	F	LAD* to RV* fistula. Closure device in situ. RCA* absent.	Same findings
2.	13y	F	LMCA* to RA* fistula. Closure device in situ. Dilated proximal part of LMCA Separate origins of fistulous track, LAD and LCx* arteries from dilated LMCA.	LMCA to RA fistula. Origin of LAD and LCx could not be delineated.
3.	11y	M	LCx to RA fistula Closure device in situ.	Same findings Patient developed local site hematoma (femoral).
4.	17y	F	RCA to RA fistula. Closure device in situ. RCA arising from proximal part of fistulous channel.	RCA to RA fistula. RCA origin could not be properly demonstrated.
5,	3y	M	RCA to RA fistula.	Same findings

			Closure device in situ.	
6.	7y	F	LCx to RA fistula. Closure device in situ.	Same findings
7.	5y	M	RCA to RA fistula Closure device in situ. Ramus intermedius was seen.	Same findings Patient developed local femoral site hematoma.
8.	10y	F	LMCA to RA fistula Closure device in situ. Origin of LAD and LCx from dilated LMCA.	LMCA to RA fistula Thrombosed distal fistulous track not seen completely. Origin of LAD and LCx could not be delineated.
9.	15y	M	LAD to RV fistula Closure device in situ.	Same findings
10.	8Y	M	RCA to RA fistula. Closure device in situ.	Same findings

*Abbreviations: LMCA – left main coronary artery, LAD – left anterior descending artery, LCx – left circumflex artery, RCA – right coronary artery, RA – right atrium, RV – right ventricle.

Figure 1: 3D Volume Rendered (VR) Image (1 A) And Conventional Coronary Angiography Image (1b, Left Anterior Oblique Projection- 40o) Showing Dilated Tortuous Left Anterior Descending Artery (LAD) Acting As Fistulous Channel, Culminating In Right Ventricle (RV).

Figure 1 A:

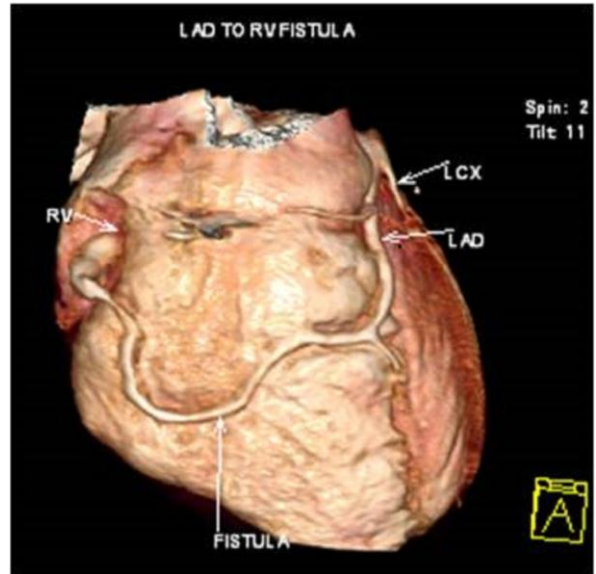


Figure 1 B:

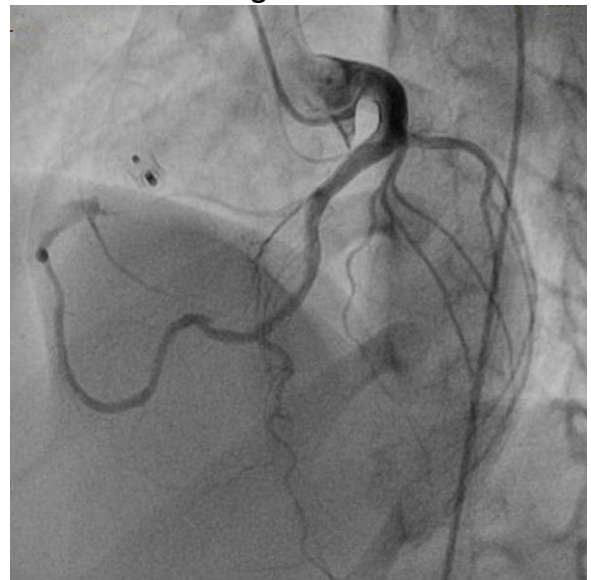


Figure 1C, Patient 1: LAD To RV Fistula -Curved MPR (Multiplanar Reformatted) Image Of The Same Patient Showing The Entire Fistulous Track. Closure Device Is Seen In Situ Near Its Culmination In Right Ventricle.



Figure 2A/2B, Patient 2: Coronal Oblique MIP (Maximum Intensity Projection) Image (2 A) And Axial MIP Image (2B) Showing Dilated Left Main Coronary Artery (LMCA) → RA Fistula. Thrombosed Distal Fistulous Track. Closure Device in Situ. No Artery Is Seen Arising From Right Coronary Cusp, Suggesting Absent Right Coronary Artery.

Figure 2 A:



Figure 2 B:

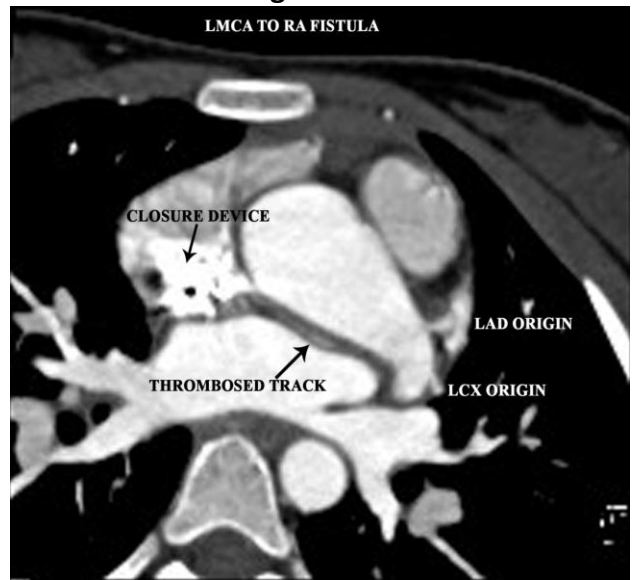


Figure 2C, Patient 2: Conventional coronary angiography image (left anterior oblique projection- 35°) of the same patient showing LMCA to RA fistula.

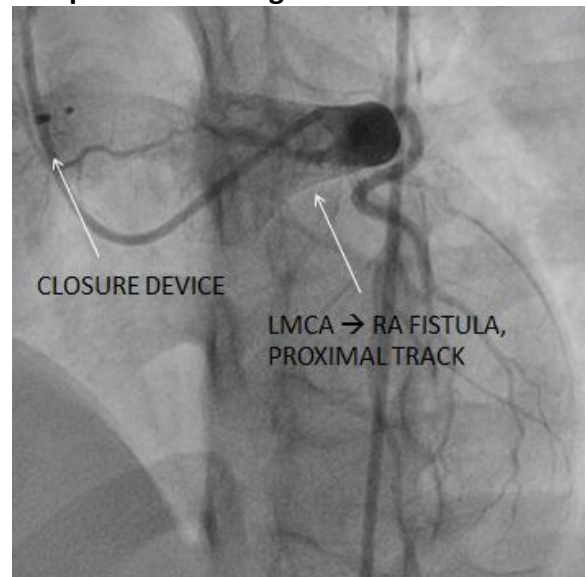


Figure 3A/3B, Patient 8: Axial MIP Image (3A) And Conventional Coronary Angiography Image (3B, Left Anterior Oblique Projection- 43o) Of The Same Patient Showing Patent LMCA To RA Fistula, Closure Device In Situ And Origin Of LAD.

Figure 3 A:



Figure 3 B:

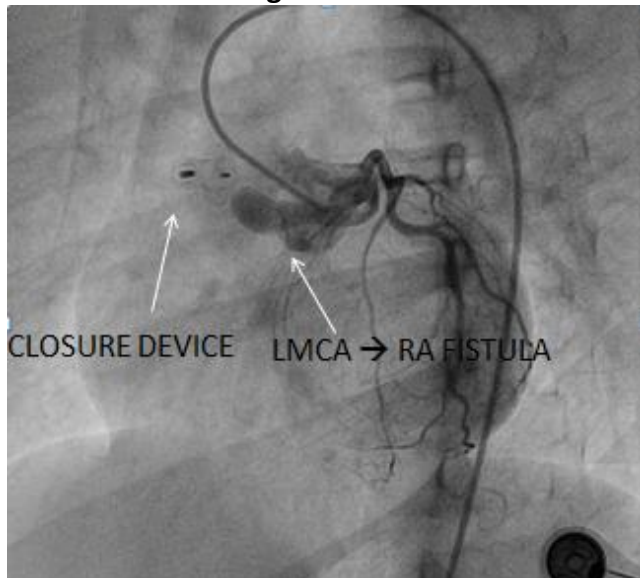


Figure 4, Patient 4: Curved MPR Image Showing Right Coronary Artery To Right Atrium Fistula With Distally Thrombosed Ectatic Fistulous Track And Closure Device In Situ.

Figure 4:



Figure 5, Patient 5: Curved MPR Image Showing Right Coronary Artery To Right Atrium Fistula In Yet Another Patient with Thrombosed Ectatic Fistulous Track.



Figure 6, Patient 6:
Curved MPR Image Showing Left Circumflex Artery to Right Atrium Fistula and Closure Device In Situ

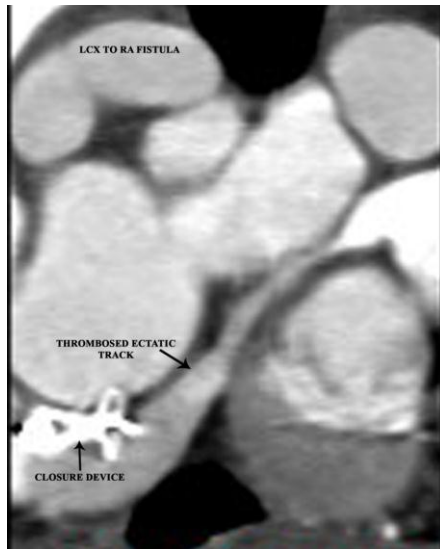


Figure 7, Patient 10:
Curved MPR Image Showing Right Coronary Artery To Right Atrium Fistula And Closure Device In Situ.



Discussion: CAF are the most common congenital coronary anomalies affecting hemodynamic parameters³. A fistula exists if a substantive communication arises bypassing the myocardial capillary phase and communicates with a low-pressure cardiac cavity (atria or ventricle) or with a branch of the systemic or pulmonary systems.

Anomalies of the coronary arteries result from a rudimentary persistence of an embryologic coronary arterial structure, a failure of normal coronary development, a failure of the normal atrophic process of development, or the misplacement of a connection of an otherwise normal coronary artery.

Over time, the coronary artery leading to the fistulous track progressively dilates, which, in turn, may progress to frank aneurysm formation, intimal ulceration, medial degeneration, intimal rupture, atherosclerotic deposition, calcification, side-branch obstruction, mural thrombosis and rarely, rupture¹¹.

When the fistula drains into the right side of the heart, the volume load is increased in this side as well as in the pulmonary vascular bed, the left atrium and the left ventricle. When the fistula drains into the left atrium or the left ventricle, although there is volumeoverloading of these chambers, there is no increase in the pulmonary blood flow. A left-to right shunt exists in over 90% of cases. The size of the shunt is determined by the size of the fistula and the pressure difference between the coronary artery and the chamber into which the fistula drains. However, the shunt ratio is generally small regardless of age; in many cases, a shunt is not detectable. Large shunts are particularly prevalent when the fistula terminates in the atrial chambers¹².

Clinical Features: Coronary artery fistulae are usually asymptomatic in the first two decades, especially when they are haemodynamically small. Indeed, a small number may close spontaneously. After this, the frequency of both symptoms and complications increases. Complications include 'steal' from the adjacent myocardium causing myocardial ischemia, thrombosis and embolism, cardiac failure, atrial fibrillation, rupture, endocarditis/endarteritis and arrhythmias. Thrombosis within the fistula is rare but may cause acute myocardial infarction, and atrial and ventricular arrhythmias⁴. Spontaneous rupture of the aneurysmal fistula causing haemopericardium has also been reported⁵.

Diagnosis, Treatment & Follow-Up: Many fistulae are small and found incidentally during coronary angiography. Coronary angiography can reliably demonstrate the proximal part of the CAF and allows evaluation of the size or number of fistulas. However, coronary fistulas drain into low-pressure chambers of the heart. These drainage sites may not be well-visualized at conventional angiography because of significant dilution of the contrast medium⁸.

CT is a useful, non-invasive, and accurate imaging technique for the detection as well as follow-up of major coronary artery anomalies. Use of multiplanar reformation may demonstrate sites of origin and termination of abnormal blood vessels. Multidetector CT has been shown to provide a high-resolution anatomic image by using electrocardiographically gated reconstruction methods. It shows enlarged fistulae and allows evaluation for aneurysmal dilatation or thrombus formation in the vessel. Volume-rendered images acquired from three-dimensional CT data sets provide an excellent overview of the cardiac and vascular anatomy and help surgeons understand the anatomic complexity before surgery⁸. Thus, multidetector CT is considered a good alternative to echocardiography and coronary angiography⁶.

CT can be performed in a single breath-hold. Also, submillimeter reconstruction imparts a higher temporal and spatial resolution to multidetector CT. New multidetector CT scanners can demonstrate coronary arteries as small as the fifth-order branches. Potential applications of CT coronary angiography include identification of anomalous origin and course of the coronary arteries, assessment of the complexity of the fistula, and preoperative evaluation⁸.

Distal vessel entry depiction by MDCT allows an assessment for the presence or absence of obstruction, which determines the likelihood of a coronary artery steal presentation.

Cardiac catheterization is the best diagnostic method for identification of CAFs, but it is invasive and has been reported to have a morbidity of 1.5% and a mortality risk of 0.15%. Therefore, MDCT may provide additional precise details of CAFs,

enabling more optimal therapeutic planning in patients where echocardiography and angiography are unable to provide adequate anatomical and physiological information¹³.

Spontaneous closure of the fistula secondary to spontaneous thrombosis has been reported, although it is uncommon (1%–2% of cases). Therefore, it is suggested that there is no urgency to close CAF immediately in asymptomatic patients and that careful periodic evaluation and close follow-up for several years are useful⁷.

In general, patients with CAF who undergo a closure procedure have an excellent prognosis. In the early postoperative period, rare instances of ST changes, myocardial infarction, or arrhythmias have been reported⁹. Long-term follow-up is essential due to the possibility of postoperative recanalization, persistent dilatation of the coronary artery and ostium, thrombus formation, calcification, arrhythmias, and myocardial infarction. Patients remain at risk for developing endocarditis until the flow is totally abolished. Therefore, they should receive antibiotic prophylaxis for any dental, gastrointestinal, or urologic procedure⁷.

Patients in which closure is performed by transcatheter techniques need to be closely followed up for complications such as residual shunts, new fistula formation, and formation of thrombus, coronary aneurysm and coronary artery stenosis.

Residual or recurrent shunts after transcatheter closure have been reported in 10–20% of patients and these may require further procedures to achieve complete occlusion¹⁴.

Conclusion: In our study we have found that for follow-up of post closure CAF patients, MDCT angiography is an excellent non-invasive tool. MDCT may provide additional precise details of CAF. Catheter-related risks, including bleeding at puncture site, haematoma formation, occurrence of arteriovenous fistula are completely eliminated. It also eliminates more serious complications of catheter angiography like coronary artery dissection, stroke and a small but definite

incidence of mortality. The procedure is carried out as an outpatient procedure, and hence, hospitalization is not needed.

References:

1. Krause W. Über den Ursprungeiner accessorischen a. coronaria aus der a. pulmonalis. *Z Ratl Med* 1865; 24: 225–227.
2. Coronary Artery Fistulas: CT Findings-Navid A. Zenoos, Reza Habibi, Leena Mammen, J. Paul Finn, Robert C. Gilkeson, *RadioGraphics* 2009; 29:781–789 • Published online 10.1148/rg.293085120.
3. Gowda RM, Vasavada BC, Khan IA. Coronary artery fistulas: clinical and therapeutic considerations. *Int J Cardiol* 2006; 107(1): 7–10.
4. Ramo OJ, Totterman KJ, Harjula AL: Thrombosed coronary artery fistula as a cause of paroxysmal atrial fibrillation and ventricular arrhythmia. *Cardiovasc Surg* 1994, 2:720-722.
5. Bauer HH, Allmendinger PD, Flaherty J, Owlia D, Rossi MA, Chen C: Congenital coronary arteriovenous fistula: spontaneous rupture and cardiac tamponade. *Ann Thorac Surg* 1996, 62:1521-1523.
6. YUN Hong, ZENG Meng-su, YANG Shan, JIN Hang and YANG Xue - Congenital coronary artery fistulas: dual-source CT findings from consecutive 6624 patients with suspected or confirmed coronary - *Chin Med J* 2011;124(24):4172-4177
7. Said SA, Lam J, van der Werf T. Solitary coronary artery fistulas: a congenital anomaly in children and adults. *Congenit Heart Dis* 2006; 1(3): 63–76.
8. Levin DC, Fellows KE, Abrams HL. Hemodynamically significant primary anomalies of the coronary arteries: angiographic aspects. *Circulation*. 1978;58:25–34
9. Gupta-Malhotra M. Coronary artery fistulas. Available from: <http://emedicine.medscape.com/article/895749-overview>, 2010.
10. Nakayama Y, Shikawa A, Ayusawa Y, Hosoda S et al. (2010). Surgical repair of complicated coronary arteriovenous fistula and coronary artery aneurysm in an elderly patient after 26 years of conservative therapy. *Heart Vessels*, Vol.26, No.1 pp. 111-116, ISSN: 1615-2573
11. Dodd JD, Ferencik M, Liberthson RR, Nieman K et al. Evaluation of efficacy of 64-slice multidetector computed tomography in patients with congenital coronary fistulas. *J Comput Assist Tomogr* 2008; 32:265–270
12. Zhu XY, Zhang DZ, Han XM, Cui CS, et al. (2010). Transcatheter Closure of Congenital Coronary Artery Fistulae: Immediate and Long-Term Follow-Up Results. *ClinCardiol*, Vol.32, No.9, pp.506–512, ISSN: 1932-8737

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