



## Cardiothoracic Imaging

## Imaging of coronary artery fistulas by multidetector CT angiography using third generation dual source CT scanner

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## ABSTRACT

Coronary artery fistulas are rare cardiac conditions which constitute a subgroup of anomalies of the coronary arteries. Though majority are asymptomatic, they may be associated with high prevalence of late symptoms and complications. Accurate identification of the fistulas, their hemodynamic significance and associated conditions generally influence management strategies. Dual source computed tomographic evaluation is valuable in delineating its precise morphology with identification and characterization of associated anomalies, thereby assisting in mapping the ideal treatment option.

## 1. Introduction

Coronary artery fistula (CAF) represents an abnormality in the termination of a coronary artery. It is defined as a direct pre-capillary connection between a branch of a coronary artery and the lumen of a cardiac chamber, coronary sinus or superior vena cava, or a pulmonary artery or pulmonary vein close to the heart [1]. It is a rare anomaly, prevalence has been reported as 0.002% in the general population and accounts for approximately 0.2% – 0.4% of congenital cardiac anomalies [2]. Majority of CAFs reported in literature are congenital and appear to represent persistence of embryonic intra-trabecular spaces and sinusoids. The rare acquired fistulas are usually iatrogenic as a complication of coronary angioplasty, coronary artery bypass surgery, or after cardiac transplantation and myocardial biopsy [3]. Rarely, they may be secondary to trauma, Takayasu arteritis or chest irradiation [4–6]. No race or sex predilection for CAF has been observed.

Though mostly asymptomatic and incidentally detected, coronary artery fistulas may be associated with chest pain, arrhythmias, endocarditis, thrombosis, myocardial infarction, stroke or very rarely sudden death. They usually drain into low pressure areas and their origin and proximal course is optimally delineated on conventional coronary angiography; however their distal evaluation may be sub-optimal [7]. Large fistulas may also be seen on echocardiography; however it provides limited information and is operator dependent [8]. Multi-detector computed tomography (MDCT) is very useful in delineating the exact anatomy of the fistula; correctly identifying its origin, course, and termination, besides providing information about

associated anomalies [8]. With the advent of third generation dual source computed tomography (DSCT) scanners and use of advanced dose reduction strategies, images can be obtained in a very short time having excellent spatial and temporal resolution with minimal radiation burden.

## 2. Clinical presentation/pathophysiology

Mostly, they are incidentally detected and are asymptomatic in adult patients unlike that seen in paediatric population. Majority of symptomatic CAFs originate from the right coronary artery than left coronary artery. Symptomatic patients often present with exertional dyspnoea and fatigue. The severity of symptoms is largely determined by the degree of shunt which in turn is dependent on the calibre of the fistula as well as the site of origin and distal drainage. Patients may present with myocardial steal phenomenon, features of which include arrhythmias, angina and myocardial infarction. Long-standing lesions may present with various complications including thrombo-embolic events, congestive heart failure, endocarditis, aneurysmal dilatation and rarely rupture [9–13].

## 3. Imaging modalities

Transthoracic echocardiography generally remains the initial investigation of choice for majority of cardiac conditions. In CAFs, dilated coronary artery may be seen and the distal drainage may be ascertained by using colour flow mapping. However, its utility may be limited in

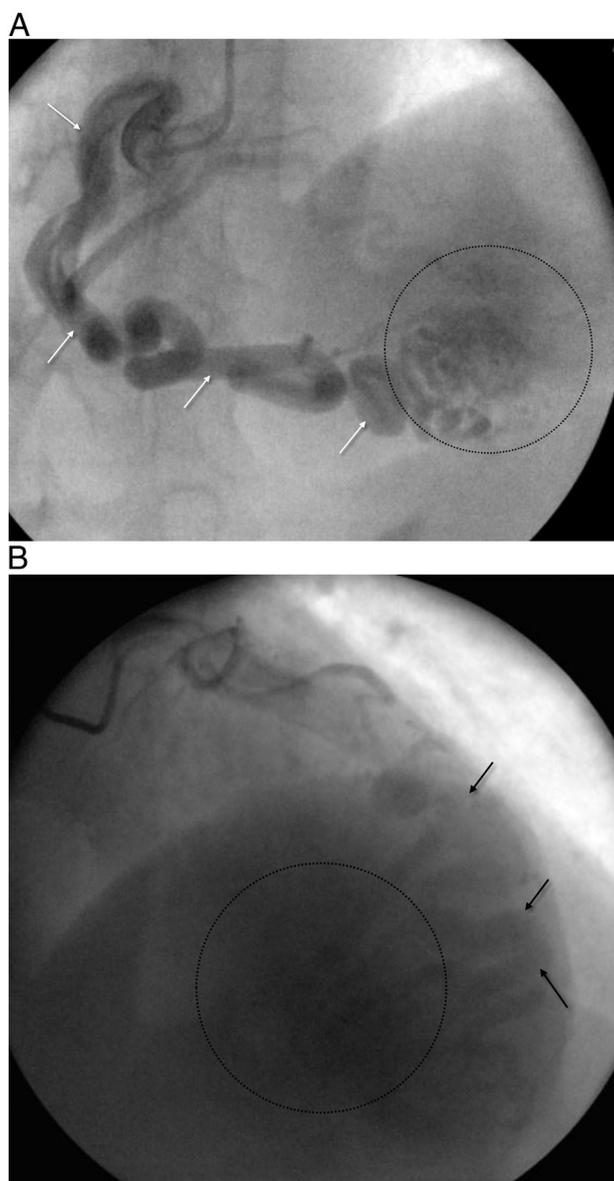
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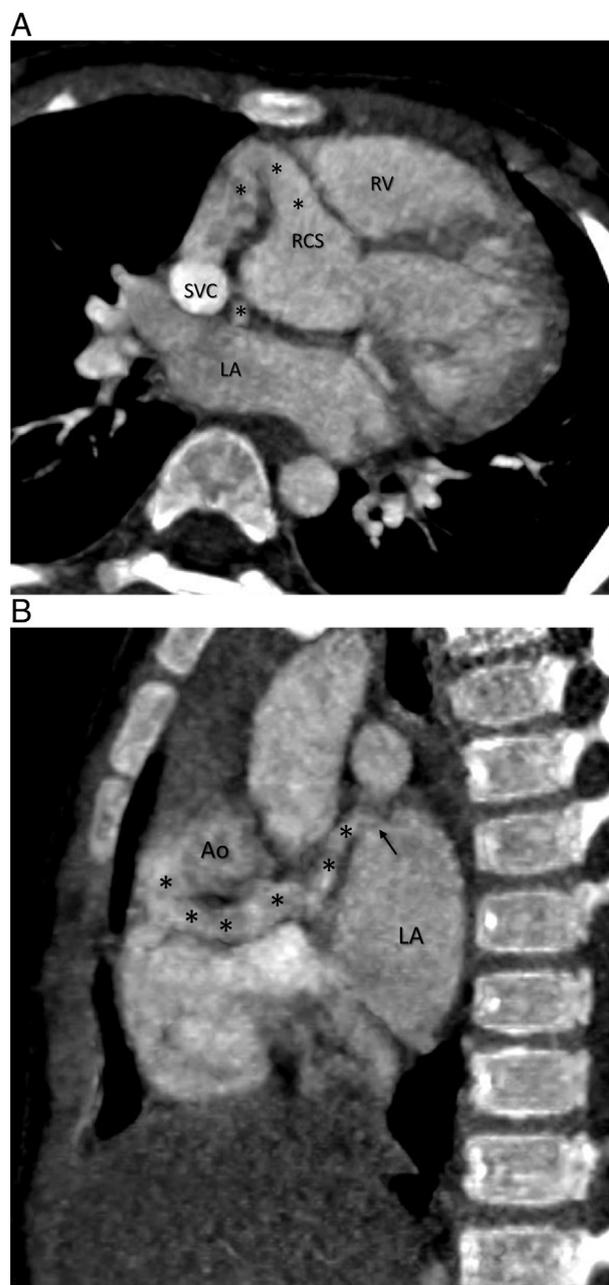


**Fig. 1.** (A) Right coronary angiogram reveals dilated and tortuous right coronary artery (indicated by white arrows) with distal fistulous communication with the right ventricle; the distal drainage site and pattern is however not very clearly delineated (dotted circle) (B) Similarly, a left coronary angiogram, in a case of left anterior descending artery to right ventricle fistula, depicts the dilated and tortuous left anterior descending artery (indicated by black arrows) with poor delineation of distal drainage point (dotted circle).

cases of small shunts and fistulas to pulmonary arteries, where the coronaries may be of normal size. Use of microbubbles to augment the colour Doppler signals helps in delimiting the site and extent of CAFs [8].

Catheter angiography is invasive in nature. Although it delineates the origin and proximal course of CAFs well, demonstration of distal drainage may be suboptimal (Fig. 1). This is attributed to the contrast dilution that occurs at the distal drainage sites, usually low pressure chambers of the heart [7]. Previously, cardiac catheterization was usually performed pre-operatively to confirm the anatomy and in planning the surgical treatment. However, with advancements in non-invasive imaging modalities, nowadays it is only used as an adjunct to endovascular management in these lesions.

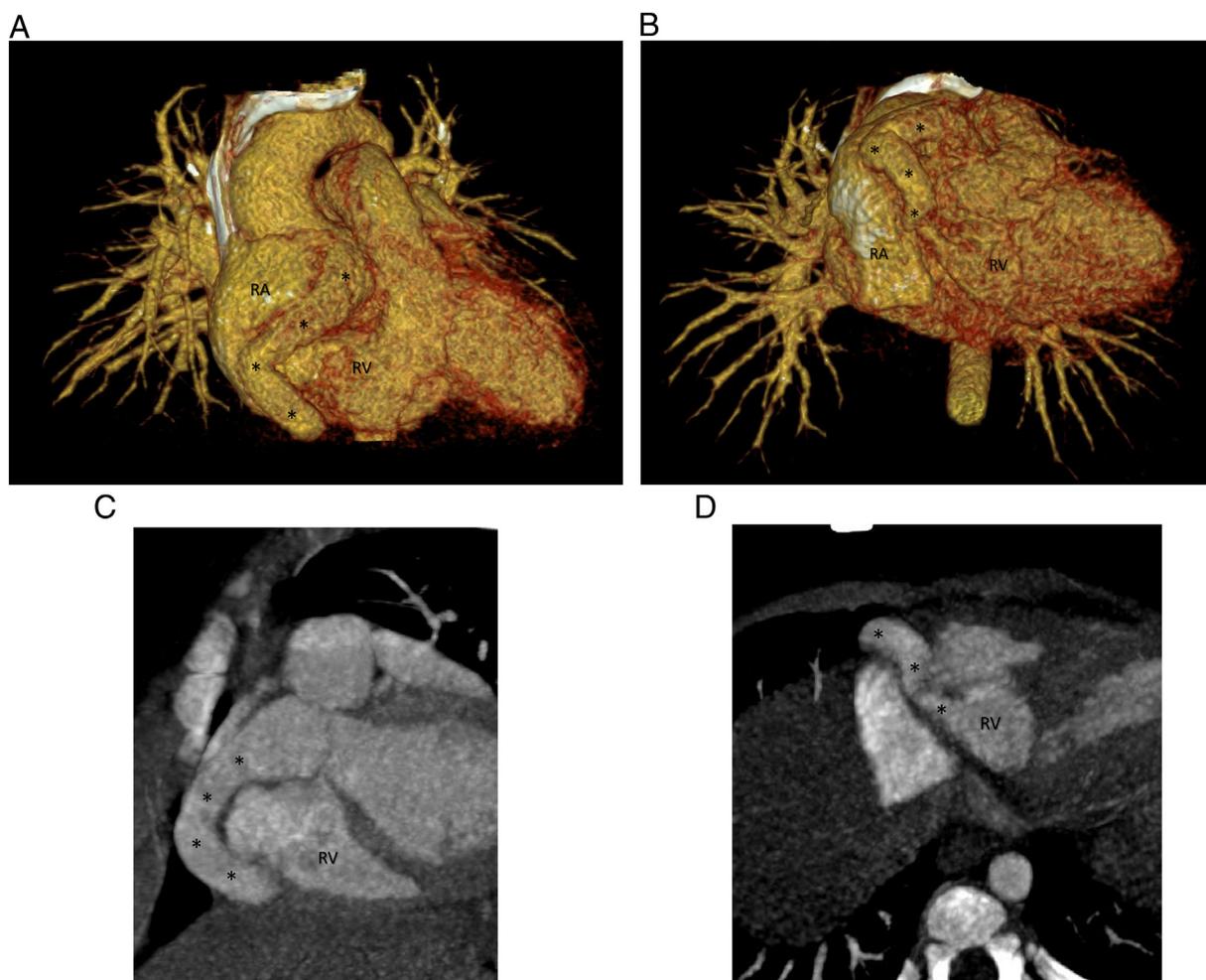
Magnetic resonance (MR) imaging has also been used in the pre-operative evaluation of CAFs. Newer MRI sequences allow



**Fig. 2.** Maximum intensity projections in the axial (A) and sagittal (B) reconstructions reveal a dilated right coronary artery (indicated by \*) draining into the left atrium (LA). Point of drainage indicated by a black arrow in Fig. 2B. (RCS: right coronary sinus; RV: right ventricle; SVC: superior vena cava; Ao: Aorta).

improvement in image quality along with better anatomical demarcation. Cine MR sequences exquisitely demonstrate the flow dynamics, especially the turbulence observed at fistulous communication site, while the black-blood sequences permit excellent visualization of the vessel lumen and its wall [14]. MRI has evolved into an imaging substitute to assess anatomic, flow-related and functional aspects of the lesion with no ionizing radiation burden [15–17]. However, MRI has its own inherent disadvantages including long acquisition times with frequent need for sedation, issues related to cost and availability in emerging countries and limited temporal and spatial resolution.

MDCT is considered to be a good alternative to echocardiography and catheter angiography for evaluation of these anomalies [18–20]. With the increasing use of MDCT, an increase in prevalence rate of



**Fig. 3.** Volume rendered images (A & B) and maximum intensity projection images in the oblique coronal (C) and axial view (D) reveals dilated right coronary artery (indicated by \*) coursing in the right atrioventricular groove and finally draining into the right ventricle (RV). (RA: right atrium).

these anomalies has been noted, owing to better sensitivity of the volumetric data acquisition. Current-generation DSCT with ECG gating provides high resolution images. Moreover, DSCT is much faster and can be performed in a single breath-hold with higher temporal and spatial resolution than those of magnetic resonance imaging (MRI). Volume rendered images acquired from three-dimensional CT data sets provide an excellent overview of the cardiac and vascular anatomy which help surgeons and interventionists understand the anatomic complexity before surgery [7]. The major limitation of CT examination is the risk of radiation exposure; the same can be substantially lowered by current-generation DSCT scanners along with advanced dose reduction techniques including automatic anatomy based & ECG based tube current modulation, use of iterative reconstruction methods and shielding.

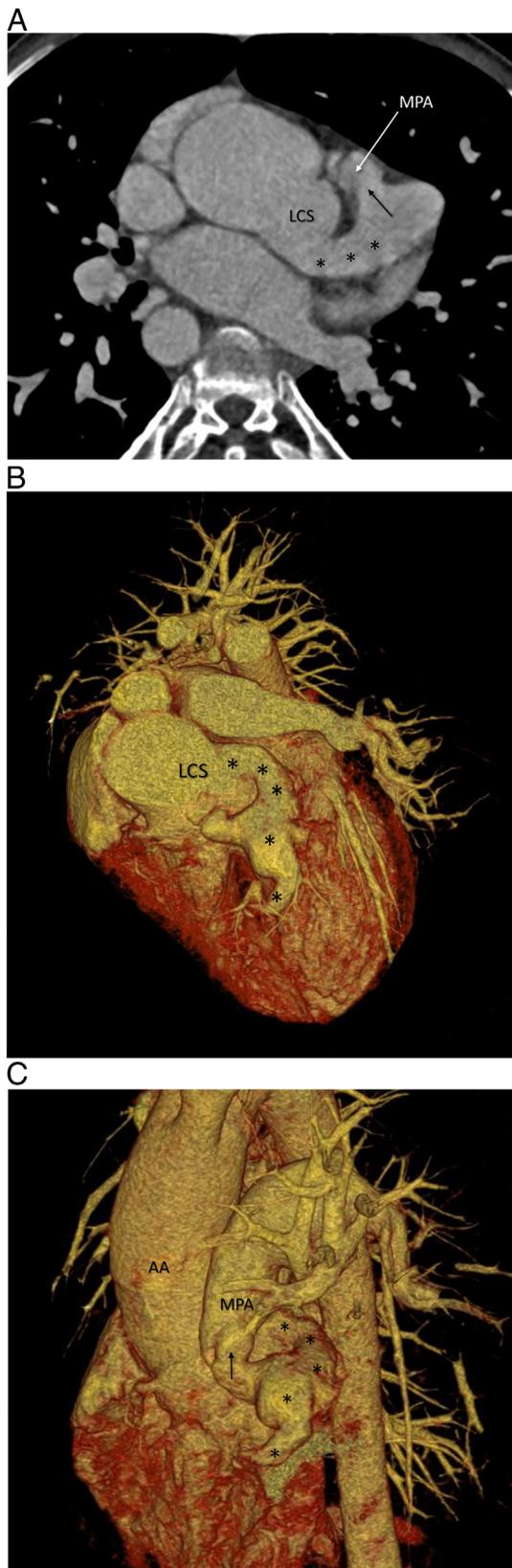
### 3.1. MDCT protocol

Retrospective ECG gated scanning with thin detector collimation was performed on a SOMATOM FORCE (Siemens Healthcare, Forchheim, Germany) CT scanner to obtain an isotropic dataset. The scanner is a third generation 384 (192 × 2) dual source CT scanner with two X-ray tubes and their corresponding detectors at an angular offset of 90 degrees. They rotate simultaneously capturing image data in half the time as compared to single source scanners. The scanner has a temporal resolution of as low as 66 ms. Along with increasing the speed of acquisition, the use of advanced dose reduction techniques have also reduced the radiation burden by more than half.

Non-ionic iodinated contrast (1.0–1.5 mL/kg) was administered via a peripheral intravenous line using a power injector at rates varying from 3.5–4.5 mL/s. Tube potential of 80 kV with automatic tube current modulation (CAREDose; Siemens Medical Solutions, Forchheim, Germany) and a reference tube current-time product of 270 mAs/rotation was used. Advanced dose reduction techniques such as anatomy based tube current modulation (CareDose; Siemens Medical Solutions, Forchheim, Germany), low kV and use of ECG guided current modulation (MinDose; Siemens Medical Solutions, Forchheim, Germany) where the tube current is reduced to 4% of its nominal value during the non-diagnostic phases of the cardiac cycle, allowed for performance of very low dose scans in all patients.

Images were reconstructed using advanced model-based iterative reconstruction (ADMIRE, strength level 3; Siemens Medical Solutions, Forchheim, Germany) with a medium soft tissue kernel (Bv40) and a slice thickness of 0.6 mm at an increment of 0.4 mm (field of view, 200 mm; pixel matrix, 512 × 512). Axial sections were analysed along with coronal, sagittal and curved multiplanar reformats (MPR), followed by volume rendered (VR) and maximum intensity projection (MIP) images on a dedicated workstation (Syngo.via; Siemens Medical Solutions, Forchheim, Germany).

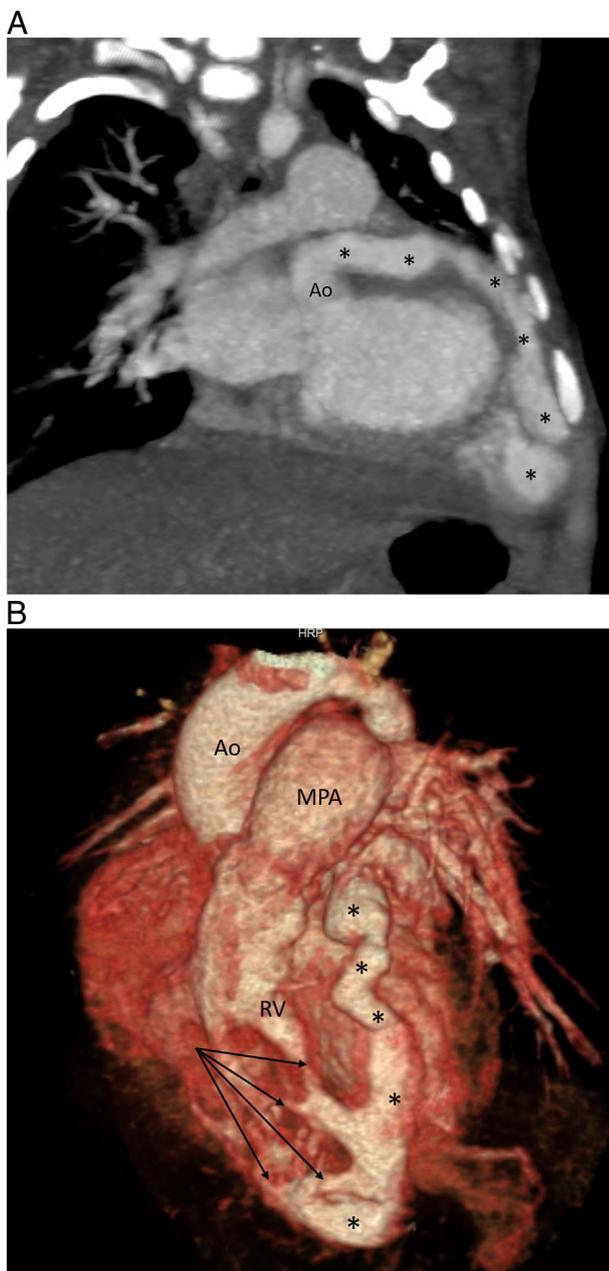
A previous study using a dual source CT scanner in the evaluation of CAF employed a similar protocol, albeit a higher flow rate of 5 mL/s was given and intravenous esmolol was used in patients whose heart rate exceeded 80 beats/min [21]. No form of heart rate control was required in any case in the current study.



**Fig. 4.** Axial maximum intensity projection image (A) and volume rendered images (B & C) show the dilated left anterior descending artery (indicated by \*) communicating with the main pulmonary artery (MPA). Point of communication indicated by black arrows in Fig. 4A and C. (LCS: left coronary sinus; AA: ascending aorta).



**Fig. 5.** Maximum intensity projection images in the axial view (A), oblique coronal reconstruction (B) and volume rendered image (C) depict the dilated & tortuous left anterior descending artery (indicated by \*) draining into the right ventricle (RV). (LCS: left coronary sinus).



**Fig. 6.** Oblique coronal maximum intensity projection image (A) and volume rendered image (B) show the dilated and tortuous left anterior descending artery (indicated by \*) communicating with the right ventricle (RV) at multiple points. Point of communications indicated by black arrows in Fig. 6B. (Ao: aorta; MPA: main pulmonary artery).

## 4. Imaging findings

### 4.1. Origin

They may arise from any of the major three coronary arteries. Several case series report a higher prevalence of CAFs involving the right coronary artery (Figs. 2 and 3), with estimates as high as 90% [22]. However, this was contradicted by a review of 12 case series including 227 patients which revealed half of the CAFs to be involving the left coronary artery (Figs. 4 and 5), 38% involving the right coronary artery and remaining 12% involving both left and right coronary arteries [23]. Moreover, CAFs behave differently with respect to their proximal versus distal origin, size and drainage, which have therapeutic implications. Sakakibara et al. classified CAFs into two types; Type A or

the proximal type where the proximal part of the involved coronary artery is dilated up to the fistula origin and the distal artery is normal, and Type B or the distal type where the entire length of the coronary artery is dilated and it terminates as a fistula usually into the right heart [24]. Fistulas having a more proximal origin have a higher probability to be dilated, show atherosclerotic changes, and give rise to myocardial steal symptoms. Lesions having a more distal origin are likely to be smaller, but can be tortuous and thus pose problems for coil embolization.

### 4.2. Drainage sites

They most commonly drain into right ventricle (Figs. 3, 5 and 6), right atrium, pulmonary artery (Fig. 4), coronary sinus, left atrium (Fig. 2), left ventricle, and superior vena cava in 41%, 26%, 17%, 7%, 5%, 3% and 1% of cases respectively [25]. In cases where the fistulous communication is to a low-pressure chamber, it may result in increased tortuosity and dilatation of the fistula. In some cases, it may also result in the formation of an aneurysm (Fig. 7) which poses a risk of rupture and can be catastrophic. Moreover, an enlarged calibre at the distal drainage site also portends a higher clinical probability of coronary steal symptoms.

### 4.3. Associations

Associated congenital cardiovascular anomalies are seen in 5% – 30% of cases [26]. Most frequently associated anomalies include tetralogy of Fallot, atrial septal defects, patent ductus arteriosus, ventricular septal defects, and pulmonary atresia with an intact ventricular septum [27]. Few rare reports also suggest associations with bronchiectasis and hereditary haemorrhagic telangiectasia [28,29].

The imaging assessment of coronary artery fistulas is summarized in Table 1 [30,31].

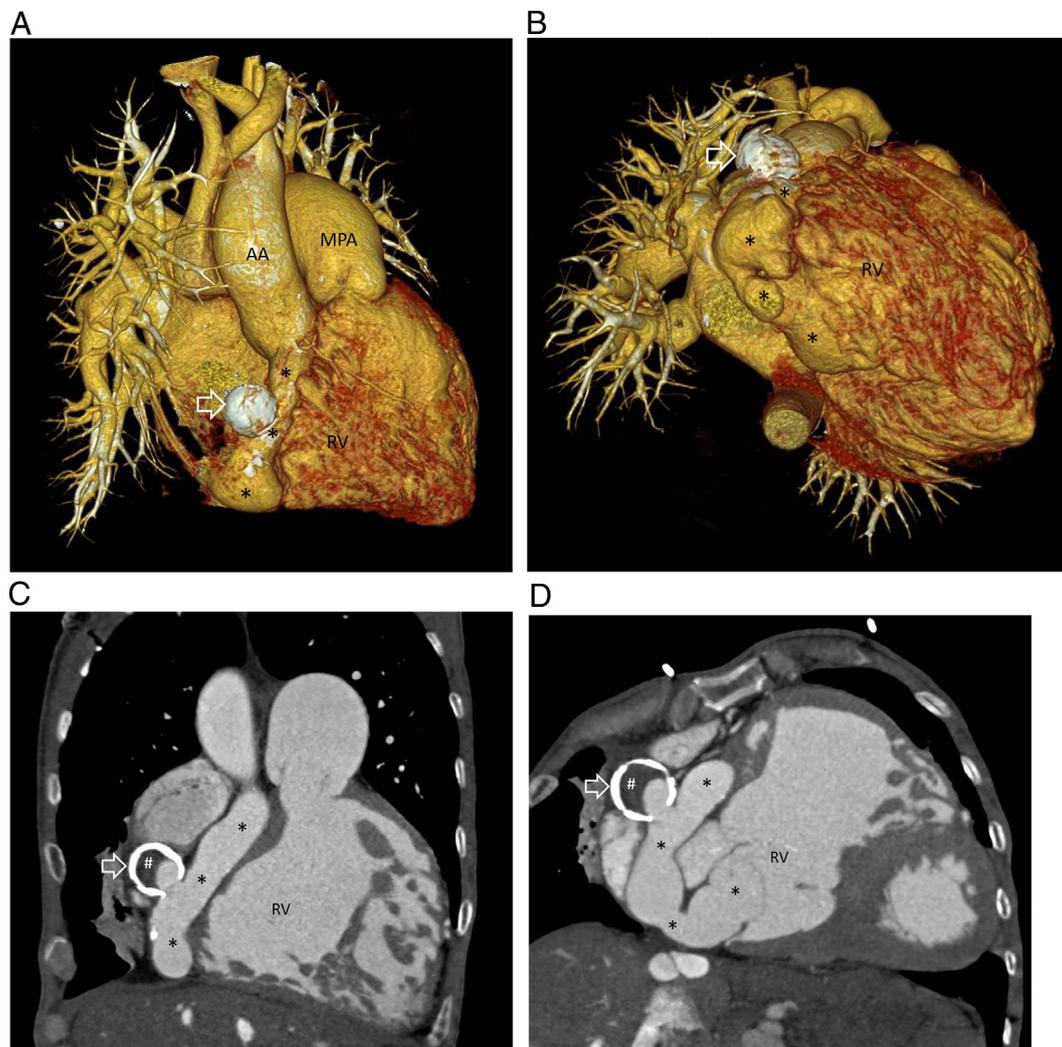
### 4.4. Factors to be considered in the decision making process regarding management of CAFs

Important variables influencing the decision making process include the size of the fistula, proximal or distal location of the fistulous communication, anatomy of the fistulous communication, number of sites of communication (single vs. multiple), presence of any significant complication or symptom which can be attributed to the fistula (e.g. angina, arrhythmias, shortness of breath, heart failure, endocarditis), age of the patient and presence or absence of any associated condition warranting a surgical or endovascular intervention [32].

## 5. Management

Spontaneous closure of fistula is rare but well known [33]. Potential complications related to untreated large fistula may include congestive heart failure, pulmonary hypertension, myocardial ischemia, bacterial endocarditis, thrombosis or rupture which can be catastrophic. Moreover, untreated larger fistulas may even predispose to premature coronary artery disease in the affected vessel. Considering the morbidity of these complications, closure of the fistula is sometimes advocated even in asymptomatic individuals. Hence the management strategy in these cases needs to be individualized. Largely the indications for fistula closure include significant left to right shunt, myocardial ischemia, a positive treadmill test, perfusion defect on stress myocardial perfusion imaging, aneurysmal dilation, mural thrombus and prevention of endoarteritis or rupture.

Surgery has long been the accepted treatment of choice for fistula closure [34,35]. Surgical management is preferred in complex cases with multiple fistulas and drainage sites with multiple feeders. Complications of surgery include myocardial infarction, arrhythmia, transient ischemic changes, and stroke. Transcatheter closure of coronary



**Fig. 7.** Volume rendered images with right atrium (RA) subtracted (A & B) and oblique maximum intensity projection images (C & D) reveals grossly dilated & tortuous right coronary artery (indicated by \*) coursing in the right atrioventricular groove and draining into the right ventricle (RV). There is an eccentric sacular aneurysm (indicated by thick white arrow) arising from the right coronary artery showing rim calcification & eccentric peripheral thrombus within (indicated by # in Fig. 7C & 7D) (AA: aorta; MPA: main pulmonary artery).

**Table 1**  
Imaging assessment of Coronary artery fistula.

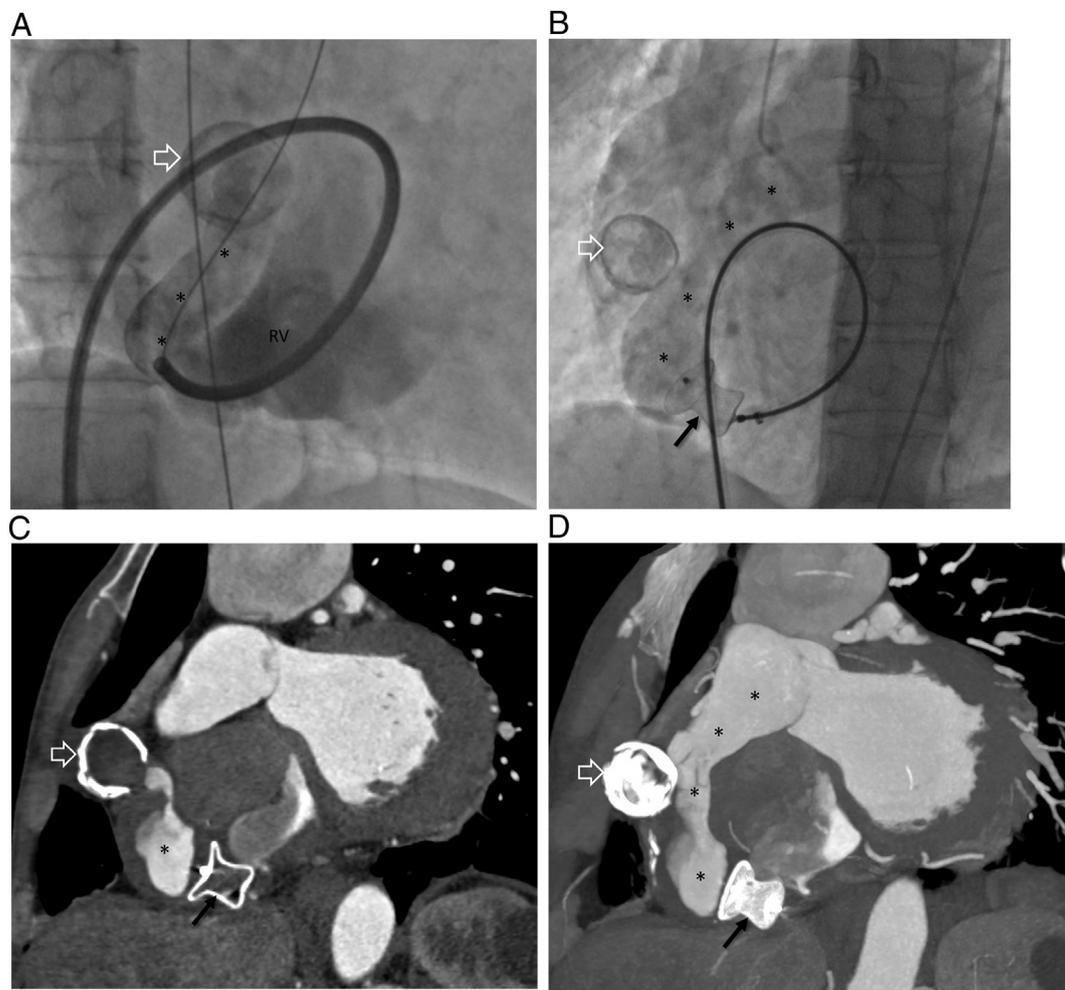
Site of origin	Right coronary artery
Segmental involvement	Left coronary artery, including left anterior descending artery, left circumflex artery or ramus intermedius
Drainage site	Type A: proximal part of the involved coronary artery is dilated up to the fistula origin and the distal artery is normal Type B: entire length of the coronary artery is dilated and it terminates as a fistula, usually into the right heart Cardiac chambers including right ventricle, right atrium, left atrium and left ventricle
Morphology	Vessels including pulmonary artery, coronary sinus and superior vena cava Size of the fistula Proximal or distal location of the fistulous communication Anatomy of the fistulous communication Number of sites of communication Presence of aneurysm or thrombus
Other associated intracardiac defects	Atrial septal defects, patent ductus arteriosus, ventricular septal defects, pulmonary atresia

arterial fistulas was first described by Reidy et al. in 1983 using a detachable balloon. Several studies have now shown that transcatheter closure is feasible, safe and effective with comparable results to that of surgery in these cases [36–41]. Moreover, the transcatheter approach is associated with avoidance of thoracotomy and cardiopulmonary bypass, decreased morbidity and shorter recovery time. Nowadays, percutaneous transcatheter closure whenever feasible is considered the treatment of choice and is generally preferred in cases with single fistula, well-defined drainage to a single site, easy accessibility to the

coronary artery that supplies the fistula and absence of large lateral branches (Fig. 8). Complications of percutaneous closure, although rare, may include coil embolization to the coronary bed or to the fistula draining bed, coronary or fistula dissection, transient myocardial ischemia or transient atrial arrhythmias.

## 6. Conclusion

CAFs are rare group of termination anomalies with varied clinical



**Fig. 8.** In the same patient as Fig. 7, catheter angiogram (A) with the catheter tip placed into the dilated right coronary artery across its drainage point into right ventricle (RV) reveals opacification of the artery (indicated by \*) and the patent part of the saccular aneurysm (indicated by white arrow) along with opacification of the RV. An Amplatzer vascular plug (indicated by black arrow in Fig. 8B) was deployed at the distal drainage point. Post deployment right coronary artery catheter angiogram reveals non-opacification of RV. On follow up CT angiogram, oblique multiplane reformatted image (C) and maximum intensity projection image (D) shows the vascular plug in situ (black arrow) with complete thrombosis of the saccular aneurysm (white arrow). Right coronary artery is patent along its course (indicated by \* in Fig. 8D).

significance. Their identification and delineation is important for advocating appropriate interventional or surgical therapy. Detection of CAFs has been increasing with more frequent use of MDCT in chest and cardiac imaging. Special attention should be paid to the courses and terminations of the coronary arteries to detect these potentially fatal anomalies. With advent of current-generation DSCT scanners along with advanced dose reduction techniques, MDCT is now regarded to be the modality of choice to evaluate such complex coronary anatomy and their anomalies, besides delineation of associated conditions.

#### Conflicts of interest

The authors declare no conflicts of interest.

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