Curr Cardiovasc Imaging Rep (2015) 8: 23
DOI 10.1007/s12410-015-9339-8

Coronary Artery Anomalies: a Pictorial Review

Joel McLarry[1] · **Maros Ferencik**[1] · **Michael D. Shapiro**[1]

Published online: 15 May 2015
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Abstract Coronary artery anomalies range in prevalence from 0.2 to 2.3 % of the population. They range from benign incidental findings to an important cause of sudden cardiac death (SCD). In fact, coronary anomalies are the second leading cause of SCD in athletes and are responsible for ~30 % of SCD in the young. Clinically, anomalous coronary arteries arising from the opposite sinus and anomalous left coronary artery arising from the pulmonary artery are the most important as they are associated with the highest risk of mortality. Several high-risk features and their pathophysiology are reviewed. Multiple imaging modalities have been utilized to study coronary artery anomalies; however, coronary computed tomography angiography (CTA) is uniquely suited to characterize coronary artery anomalies as it allows for clear elucidation of origin, course, and termination in relationship to other relevant anatomy with high spatial resolution. This paper will provide an overview of the wide spectrum of coronary artery anomalies and variants, review the most relevant coronary CTA imaging features for each, and differentiate benign from malignant varieties.

Keywords Anomalous coronary artery · Computed tomography · Coronary CTA · Sudden cardiac death · Anomalous coronary artery arising from the opposite sinus

This article is part of the Topical Collection on *Cardiac Computed Tomography*

✉ Michael D. Shapiro
shapirmi@ohsu.edu

[1] Knight Cardiovascular Institute, Oregon Health & Science University, UHN62, 3181 SW Sam Jackson Park Rd, Portland, OR 97239, USA

Introduction

Collectively, anomalous coronary arteries and variants are estimated to have a prevalence of 0.2–2.3 % [1•, 2, 3••, 4]. However, the true prevalence of these conditions is unknown since detection is subject to referral bias and the work-up is usually prompted by suspicion for coronary artery disease or a coronary anomaly. Importantly, coronary anomalies are the second most common cause of mortality in competitive athletes (after hypertrophic cardiomyopathy) and are associated with ~30 % of sudden cardiac death (SCD) in young adults [5, 6]. Historically, conventional coronary angiography (CCA) has been considered the gold standard for detecting coronary artery anomalies. However, CCA is an invasive procedure and it only detects about half of coronary artery anomalies [7, 8]. Other modalities such as cardiac MRI (CMR) elucidate the proximal coronary artery but lack the spatial resolution needed to define the entire course. Conversely, coronary computed tomography angiography (CTA) is well suited to characterize the coronary origin, course, and termination with superb spatial and temporal resolution. Additionally, it has the advantage of defining the anomalous coronary arteries relative to other important anatomic structures, such as the great vessels. There is increasing recognition of the utility of coronary CTA for this purpose. Recommendations for the use of coronary CTA in the evaluation of anomalous coronary arteries have been incorporated into ACC/AHA Guidelines and Appropriate Use Criteria [9, 10].

Variants vs Anomalies

The distinction between a coronary anomaly and a variant is defined by population prevalence. By definition, coronary artery anomalies are present in <1 % of the population and normal variants are found in >1 % of the population [11]. However, several studies have demonstrated a higher prevalence of certain coronary "anomalies" [4, 12, 13] (Table 1). Fortunately, many anomalies are benign and serve simply as incidental findings without prognostic relevance. The current discussion will focus on the most clinically relevant coronary anomalies. Clearly, individuals who interpret coronary CTA studies need to be familiar with the entire spectrum of coronary anomalies in order to differentiate those that are benign from those that are malignant.

Prevalence of Coronary Anomalies in Congenital Heart Disease

This review focuses on isolated coronary artery anomalies, not in the setting of concomitant congenital heart disease.

Table 1 Prevalence of coronary artery anomalies and specifically ACAOS by CT angiography

	n	Prevalence of all coronary artery anomalies	Prevalence of ACAOS	Prevalence of specific ACAOS subgroups
CTA				
Krupinski [23••]	7115	0.76 % (only ACAOS reported)	0.76 %	LCA from RCS 0.182 %
				RCA from LCS 0.084 %
				LCX from RCS 0.309 %
				LCA from NCS 0.028 %
Nasis [13]	9774	1.14 % (only anomalies of origin reported)	1.09 %	RCA from LCS 0.368 %
				LCA from RCS 0.726 %
Opolski [3••]	8522	0.844 % (only ACAOS reported)	0.844 %	LCA from RCS 0.129 %
				LAD from RCS 0.106 %
				LCX from RCS 0.375 %
				RCA from LCS 0.235 %
Namgung [1•]	8864	1.14 %	0.47 %	RCA from LCS 0.463 %
				LCA from RCS 0.068 %
				LCX from RCS 0.00 %
Szymczyk [24]	726	1.1 % (only ACAOS)	1.1 %	LCX from RCS (0.6 %)
				RCA from LCS 0.3 %
				LCA from RCS 0.1 %
				LCX from RCA 0.1 %
Park JH [25]	1582	1.14 %	0.7 %	Not reported
Villines [22]	577	2.1 % (only anomalies of origin reported)	2.1 %	LCA from RCS 0.35 %
				LCX from RCS 0.69 %
				RCA from LCS 1.04 %
Zhang [26]	1879	1.3 %	0.905 %	RCA from LCS 0.639 %
				LCA from RCS 0.053 %
				LCA from NCS 0.160 %
				LAD from RCS 0.000 %
				LCX from RCS 0.053 %
Cheng [27]	3625	0.99 %	Not reported	Not reported
von Ziegler [2]	748	2.3 %	2.139 %	RCA from LCS 1.070 %
				LCA from RCS 0.134 %
				LCX from RCS 0.936 %
Kosar [28]	700	2.139 %	1 %	RCA from LCS 0.571 %
				LCA from RCS 0.286 %
				LCX from RCS 0.143 %
Duran [29]	725	5.793 %	0.551 %	LCA from RCS 0.138 %
				LCX from RCS 0.413 %
Sato [30]	1153	0.43 % (only anomalies of origination reported)	0.347 %	RCA from LCS 0.260 %
				LCX from RCS 0.087 %

ACAOS anomalous origin of a coronary artery from the opposite sinus

However, coronary anomalies are found much more commonly in the setting of congenital heart disease with a prevalence as high as 36 % in single ventricle patients and 15 % overall in a diverse group of congenital heart disease subtypes [14].

Anomalies of Origin and Course

The majority of coronary anomalies fall within this group. This group is further divided into three subcategories: absent left main coronary artery, anomalous coronary ostium outside of the aortic sinuses, and anomalous origin of a coronary artery from the opposite sinus (ACAOS).

Absent Left Main Coronary Artery

Absent left main coronary artery is characterized by separate ostia for the left anterior descending artery and the left circumflex. It is associated with a higher proportion of left dominant coronary circulation and myocardial bridging. The prevalence of this benign anomaly is estimated at 0.4 % in one series [15].

Anomalous Coronary Ostium Outside of the Aortic Sinuses

The spectrum of coronary ostial anomalies and variants outside of the aortic sinuses is broad, ranging from clinically benign conditions to those that result in early mortality, often within the first year of life if corrective surgery is not pursued early. Some variants, such as superior takeoff of the right coronary artery, are common (8.7 %) and are unlikely to be of clinical significance [1•]. Other anomalies such as coronary origins from the left ventricular outflow tract, ascending aorta (Fig. 1), and brachiocephalic or subclavian arteries are found less commonly, and their clinical significance is uncertain.

Patients with anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA) typically present in the first few months of life with angina-like episodes and/or cardiomyopathy [16]. This condition occurs in 1 in 300,000 live births and, if untreated, is associated with a 1-year mortality ranging from 35 to 90 % as a result of progressive heart failure or life-threatening arrhythmia [16, 17]. ALCAPA pathophysiology results in myocardial ischemia as a result of coronary steal. The right coronary with a normal course is perfused at aortic diastolic pressure which then flows down a pressure gradient through collaterals to the left coronary artery and ultimately to the pulmonary artery whose maximum pressure is the pulmonary artery systolic pressure (typically less than half the aortic diastolic pressure). Rare individuals with ALCAPA can develop very robust collaterals and thus present later in childhood or adulthood with angina or SCD [16, 18]. ALCAPA can be repaired with direct coronary re-implantation or intrapulmonary or extrapulmonary baffling of the left coronary artery. Direct coronary re-implantation appears to have the least post-operative complications but is not always feasible when the length of the coronary artery is insufficient [19]. Intrapulmonary baffling commonly leads to suprapulmonary stenosis (16 of 21 patients) or baffle leaks (11 of 21 patients) [19].

Anomalous origin of the right coronary artery from the pulmonary artery (ARCAPA) (Fig. 2) is found less commonly than ALCAPA with an estimated prevalence of only 0.002 % in the general population. It can manifest with a presentation similar to ALCAPA with heart failure or sudden cardiac death in infancy. However, patients with ARCAPA typically present later (average age at diagnosis 22.8 years) with signs of ischemia or during an evaluation for a murmur [20] (Fig. 2).

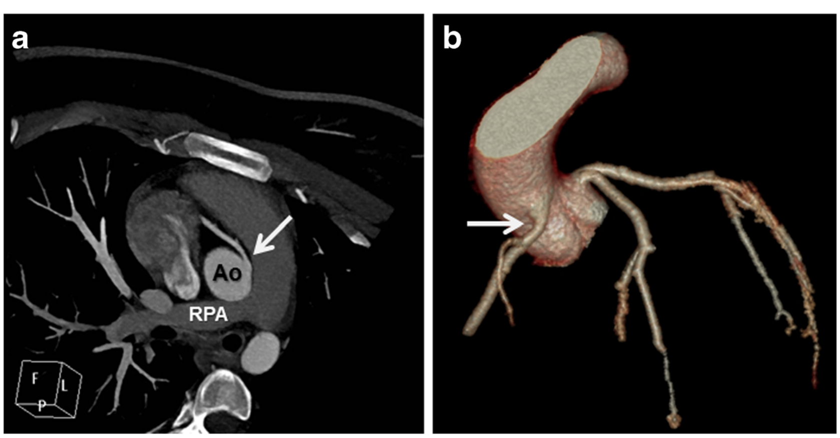

Fig. 1 A 71-year-old male underwent non-invasive coronary evaluation for chest pain. **a** An oblique axial maximum intensity projection image demonstrates the superior origin of the right coronary artery (*arrow*) from the ascending aorta at the level of the right pulmonary artery. **b** A three-dimensional volume-rendered image demonstrates the superior takeoff of the right coronary artery above the right sinus of Valsalva (*arrow*) with normal origin of the left coronary artery. *Ao* aorta, *RPA* right pulmonary artery

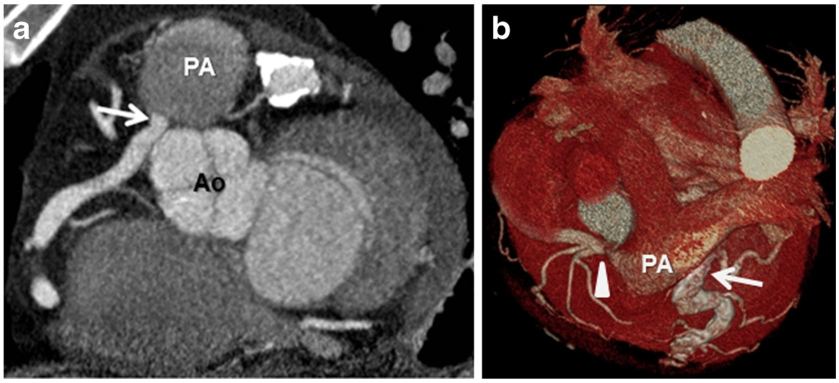

Fig. 2 A 52-year-old male presents with dyspnea on exertion. An evaluation led to a coronary CTA. **a** The maximum intensity projection image demonstrates the anomalous right coronary artery arising directly from the pulmonary artery (ARCAPA) (*arrow*). **b** The three-dimensional volume-rendered image demonstrates a markedly dilated and tortuous left anterior descending artery (*arrow*) that supplies extensive collaterals to the anomalous right coronary artery (*arrowhead*) arising from the pulmonary artery. *PA* pulmonary artery, *Ao* aorta

ACAOS

ACAOS comprises a large group of coronary anomalies that is of utmost clinical importance. This condition has received a great deal of attention as an important cause of SCD in competitive athletes and young adults. For most clinicians, it represents the entity that is evoked during a discussion of anomalous coronary arteries. The prevalence estimates of ACAOS by echocardiography, coronary CTA, CMR, CCA, and autopsy range widely from 0.15 [21] to 2.1 % [2, 22]. The prevalence of coronary anomalies and specifically ACAOS has been reported extensively (Table 1) [1•, 2, 3••, 13, 22, 23••, 24–30]. Since patients undergoing diagnostic tests that identify ACAOS typically have symptoms prompting the referral, the true population prevalence of ACAOS is expected to be lower than reported prevalence estimates. Indeed, the few large studies that have prospectively evaluated asymptomatic patients found the incidence of ACAOS to be much lower at 0–0.17 % [31–33]. Younger cohorts with ACAOS are commonly asymptomatic. However, as these patients age, chest pain, myocardial infarction, and arrhythmia become more common [3••]. Significant interest has been focused on identifying the anatomical and pathophysiological features that predict these adverse outcomes.

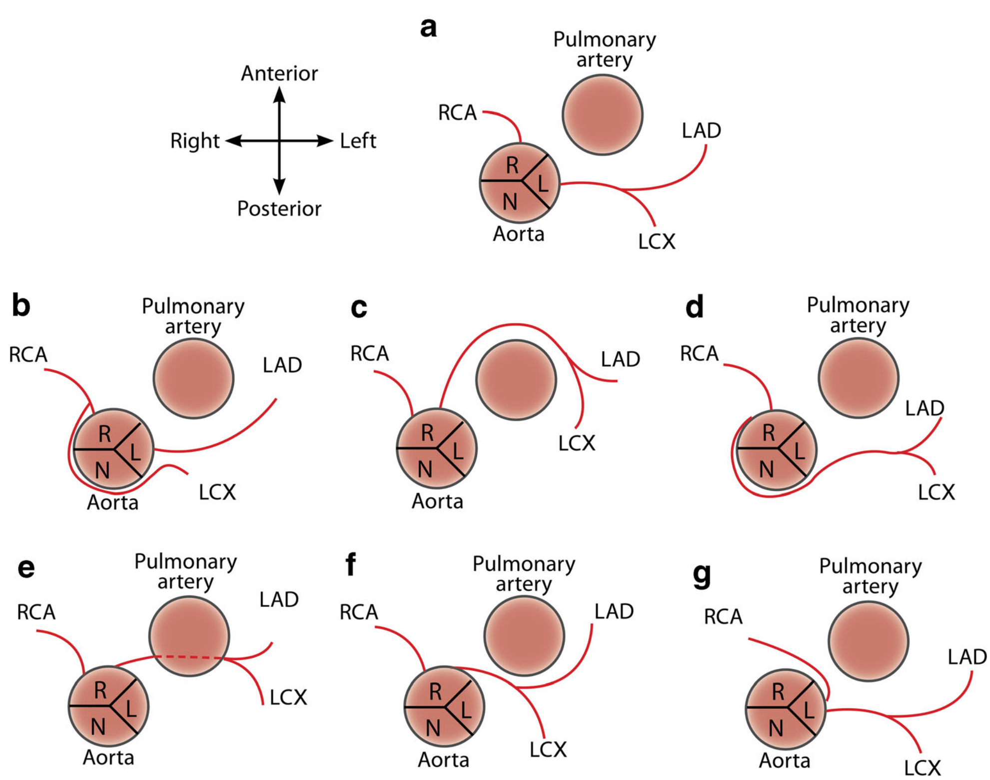

Fig. 3 Normal coronary artery origin and course contrasted with several varieties of ACAOS. **a** Normal coronary artery origin and course. **b** Anomalous origin of the LCX from the RCA with a posterior (retroaortic) course; this is more commonly encountered than ACAOS of the LCX from the right sinus of Valsalva. **c** ACAOS of the LM with an anterior (prepulmonic) course. **d** ACAOS of the LM with a posterior (retroaortic) course. **e** ACAOS of the LM with a subpulmonic (intramyocardial) course. **f** ACAOS of the LM with an interarterial course. **g** ACAOS of the RCA with an interarterial course. *LCX* left circumflex coronary artery, *RCA* right coronary artery, *LAD* left anterior descending artery

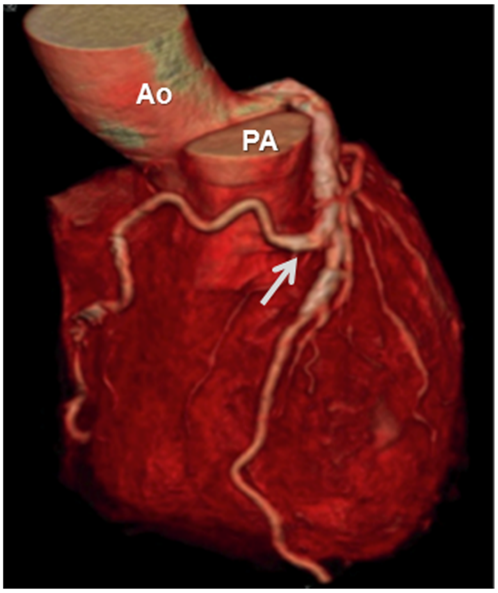

Fig. 4 A three-dimensional volume-rendered image demonstrates anomalous right coronary artery arising from the left anterior descending artery (*arrow*) as it courses anterior (prepulmonic) to the pulmonary artery. This coronary anomaly is considered benign and is not associated with a significant risk for sudden cardiac death or ischemia. *Ao* aorta, *PA* pulmonary artery

Extensive classification systems have been developed [12] but ACAOS is typically grouped into four main categories (Fig. 3): left main coronary artery from the right sinus of Valsalva, left anterior descending artery from the right sinus of Valsalva (Fig. 4), left circumflex from the right sinus of Valsalva or, more commonly, right coronary artery (Fig. 5) [34•], and right coronary artery from the left sinus of Valsalva (Table 2). The proximal course of the anomalous coronary is further categorized into interarterial (between the aortic root and pulmonary artery) (Fig. 6), anterior (prepulmonic) (Fig. 7), intraseptal (subpulmonic) (Fig. 8), and posterior (retroaortic) (Fig. 9) subtypes. A single coronary that supplies the left anterior descending, left circumflex,

Table 2 Classification scheme of anomalous origin of a coronary artery from the opposite sinus

• LAD from the right sinus of Valsalva
Interarterial
Intraseptal (subpulmonic)
Anterior (prepulmonic)
• LCX from the right sinus of Valsalva/proximal RCA
Posterior (retroaortic)
• LMCA or RCA from the opposite sinus of Valsalva
Interarterial
Intraseptal
Posterior
Anterior

LAD left anterior descending artery, *LCX* left circumflex coronary artery, *RCA* right coronary artery, *LMCA* left main coronary artery

and right coronary arteries is extremely rare (Fig. 10). It shares some overlapping features with ACAOS, including a similar risk for ischemia and sudden cardiac death depending on its origin and course.

High-Risk Anatomic Features of ACAOS

ACAOS is a heterogeneous group of coronary anomalies with variable clinical expression. In fact, most ACAOS are not associated with a high risk of myocardial ischemia/SCD. The malignant variety is almost exclusively limited to ACAOS with an interarterial course (Figs. 6 and 11). High-risk anatomical features of ACAOS with an interarterial course associated with myocardial ischemia and SCD include acute angle of origin, a slit-like coronary ostium, and an intramural course. Anomalous left coronary arteries have a higher risk of SCD compared to anomalous right coronary arteries [6, 35, 36]. Not surprisingly when the anomalous coronary artery is dominant, higher rates of SCD have

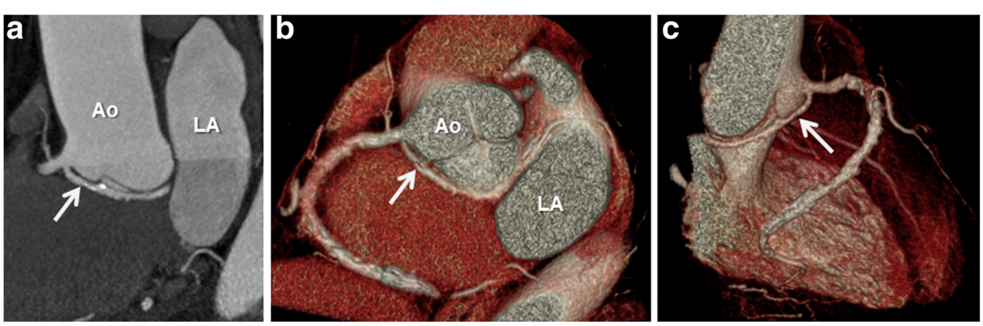

Fig. 5 A 54-year-old male with multiple medical problems presented with intermittent, non-exertional chest pain and underwent coronary CTA. **a** A multiplanar reformatted thin maximum intensity projection image demonstrates the anomalous origin of the left circumflex (*arrow*) from the proximal right coronary artery with evidence of calcified atherosclerotic plaque. **b**, **c** The three-dimensional volume-rendered images demonstrate the posterior course (*arrow*) of the anomalous circumflex between the aorta and the left atrium. This anomaly does not carry an increased risk for sudden cardiac death but is associated with a high prevalence of atherosclerosis [34•]. *Ao* aorta, *LA* left atrium

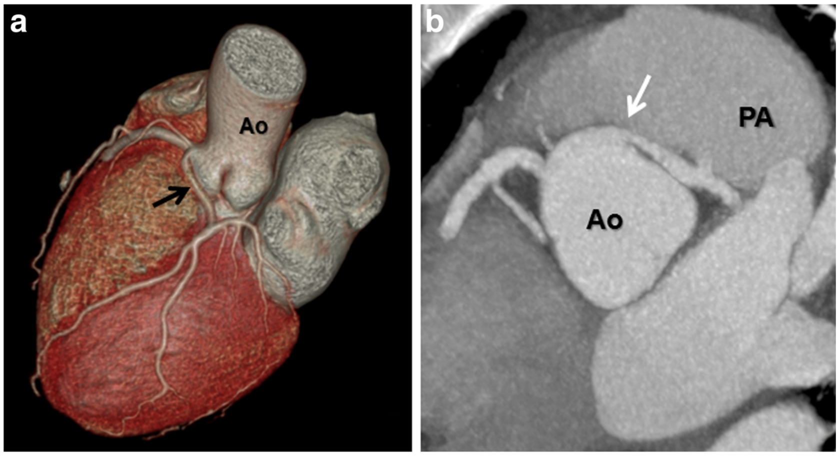

Fig. 6 A 55-year-old female presented with exertional syncope. The evaluation culminated in a coronary CTA that demonstrated an anomalous left main coronary artery arising from the right sinus of Valsalva and coursing between the aorta and pulmonary artery. **a** The three-dimensional volume-rendered image demonstrates the malignant interarterial course of the anomalous left main (*arrow*) coronary artery between the aorta and pulmonary artery (subtracted to visualize the left main). **b** The maximum intensity projection image demonstrates the acute angulation, slit-like ostium, and the anomalous course of the left main coronary artery (*arrow*) between the aorta and pulmonary artery. The interarterial form of ACAOS is associated with an intramural course that leads to coronary intussusception with exertion, luminal compression, critical reduction of flow, and an increased risk for sudden cardiac death. *Ao* aorta, *PA* pulmonary artery

been observed [37]. Although anomalous right coronary arteries (Fig. 11) are associated with a lower rate of SCD, they may be more prone to ischemia as evidenced by a higher proportion of patients presenting with chest pain compared to anomalous left coronary arteries [23••].

Mechanism of Sudden Cardiac Death in ACAOS

The physiological underpinning(s) for myocardial ischemia/SCD in ACAOS has been a matter of debate. Table 3 lists a number of proposed mechanisms for reduced coronary flow in the setting of ACAOS [37, 38]. As previously mentioned, an interarterial coronary course is the subtype of ACAOS that is associated with the worst prognosis [12, 37, 39–41]. In particular, a left main coronary artery with an interarterial course is associated with the highest risk of sudden cardiac death. However, since the true prevalence is unknown, so is the actual risk of sudden death among affected individuals. The association between an interarterial course and SCD led many to hypothesize that the great arteries critically compress the anomalous coronary, leading to ischemia and fatal arrhythmia [42]. However, since coronary diastolic pressure is greater than pulmonary artery diastolic pressure (in the absence of pulmonary hypertension), mechanical compression of the coronary artery is unlikely. Using intravascular ultrasound (IVUS), Angelini et al. [43] demonstrated that an interarterial course is associated with a proximal segment of the coronary anomaly that runs within the aortic wall (intramural course). During pharmacologic stress, proximal intussusception of the anomalous artery at the

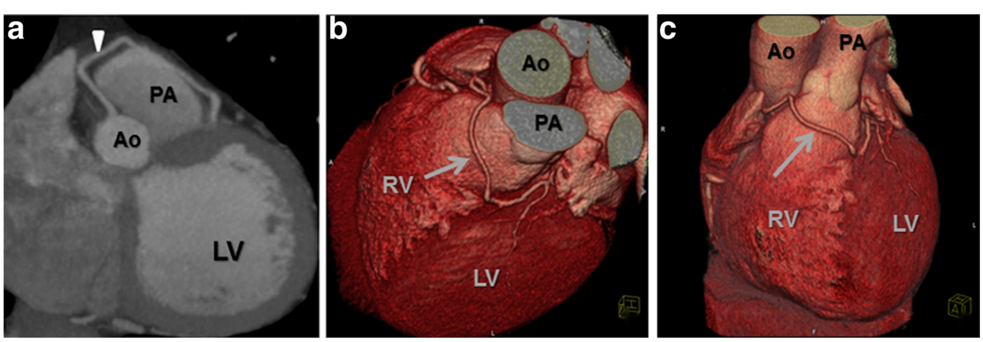

Fig. 7 Anomalous left main coronary artery arising from the right sinus of Valsalva and coursing anterior to the pulmonary artery discovered incidentally on coronary CTA during evaluation for atypical chest pain in a 47-year-old woman. **a** The maximum intensity projection image demonstrates the left main coronary artery arising from the right sinus of Valsalva with a course anterior to the pulmonary artery (prepulmonic) (*arrowhead*). **b**, **c** The three-dimensional volume-rendered images demonstrate the benign course anterior to the pulmonary artery (*arrows*) without acute angulation or a slit-like coronary ostium. This ACAOS is considered benign. *LV* left ventricle, *RV* right ventricle, *Ao* aorta, *PA* pulmonary artery

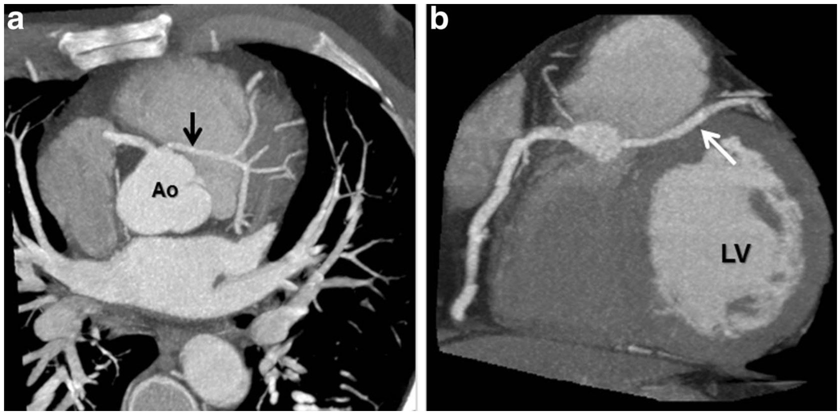

Fig. 8 A 57-year-old male presented with atypical chest pain and subsequently underwent coronary CTA. The CT demonstrated an anomalous left main coronary artery originating from the right sinus of Valsalva with an intraseptal (subpulmonic) course. **a** The maximum intensity projection image demonstrates the anomalous left main coronary artery arising from the right sinus and coursing within the interventricular septum (*arrow*). **b** The oblique coronal image demonstrates the intraseptal course of the anomalous coronary artery (*arrow*) as it passes below the pulmonary artery. This ACAOS is considered benign. *Ao* aorta, *LV* left ventricle

aortic root wall occurs, leading to significant coronary stenosis. The degree of coronary hypoplasia and lateral compression of the coronary lumen, both found within the intramural segment, predicts ischemia as well. A large degree of interindividual variation exists with regard to the extent of coronary hypoplasia, lateral compression, and ultimately stenosis length. This variation likely, in large part, explains the heterogeneity in clinical presentation of ACAOS. Other investigators have investigated anomalous right coronary artery from the left cusp with fractional flow reserve (FFR) during dobutamine stress [44••]. Of 33 patients, 3 demonstrated an abnormal FFR (≤0.80) and all affected patients had an interarterial course and a slit-like coronary ostium. Their findings with FFR were corroborated by the degree of stenosis assessed by IVUS. Interestingly, 9 patients had a positive non-invasive stress test, but none of them had a significant stenosis by IVUS or FFR.

Anomalies of Intrinsic Coronary Artery Anatomy

Left Main Coronary Atresia

Left main coronary atresia is a rare condition associated with absence of a left coronary ostium/left main trunk (Fig. 12) [45]. The blood supply to the left anterior descending coronary artery and left circumflex originates through collaterals from the right coronary artery. Left main coronary atresia is almost universally symptomatic with only one patient reported in the literature that remained asymptomatic throughout life. Prognosis is poor without coronary artery bypass grafting [46].

Myocardial Bridging

Myocardial bridges are coronary anatomic variants in which a segment of the epicardial coronary artery takes an intramyocardial course. Myocardial bridges most commonly

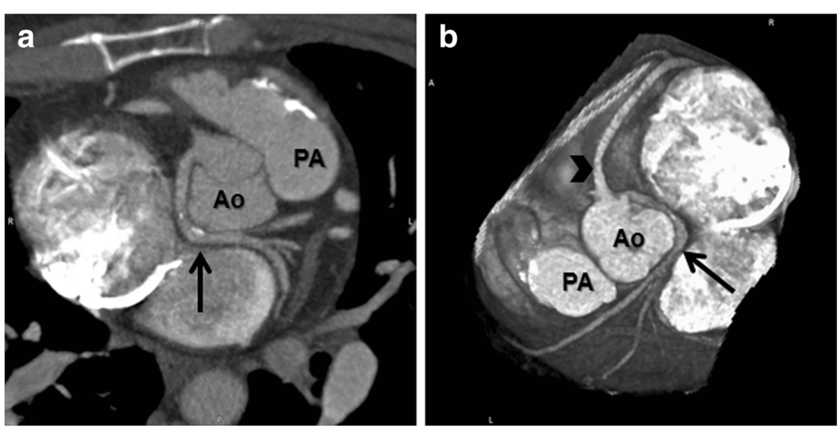

Fig. 9 A 35-year-old man was referred for coronary CTA after a non-gated chest CT suggested ACAOS. **a** The maximum intensity projection image demonstrates the left main coronary artery arising from the right sinus of Valsalva with a posterior (retroaortic) course (*arrow*). Calcified atherosclerotic plaques in the left main coronary artery are present. **b** The three-dimensional volume-rendered image demonstrates the relationship of the anomalous left main coronary artery (*arrow*) to the right coronary artery (*chevron*). This ACAOS is considered benign. *Ao* aorta, *PA* pulmonary artery

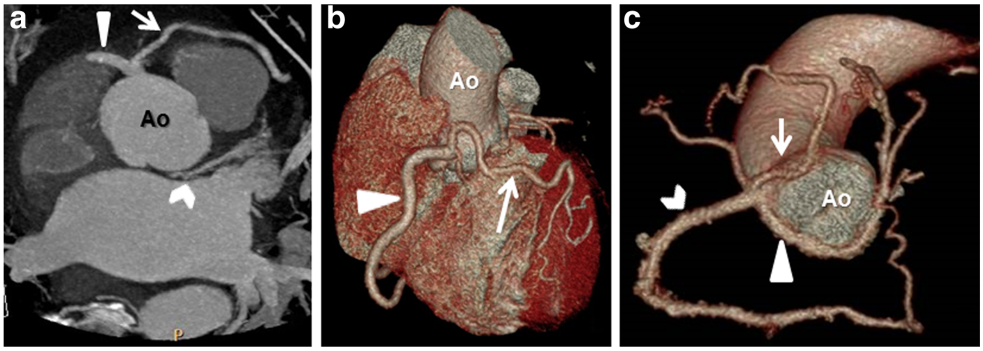

Fig. 10 A 67-year-old male presented with chest pain and was found to have a pulmonary embolism. Incidental note was made of a coronary anomaly that was further evaluated with coronary CTA. **a** A maximum intensity projection image demonstrates the single coronary ostium from the right coronary cusp, branching into the right coronary artery (*arrowhead*), left circumflex (with a posterior course) (*chevron*), and left anterior descending artery (with an anterior course) (*arrow*). **b** A three-dimensional volume-rendered image demonstrates the normal RCA course (*arrowhead*) and the anomalous origin of the LAD from the RCA (*arrow*). **c** Another three-dimensional volume-rendered image demonstrates the single coronary ostium with all three branch arteries in view with an anterior course of the left anterior descending artery (*arrow*), a posterior course of the left circumflex (*arrowhead*), and a normal RCA course (*chevron*). *Ao* aorta

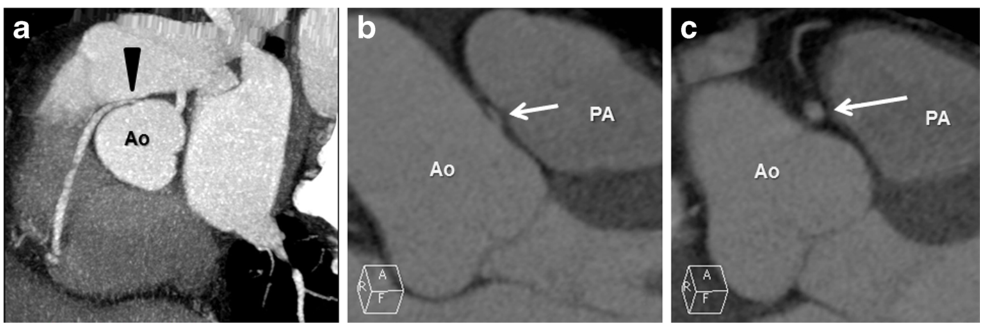

Fig. 11 A 71-year-old female presented with exertional chest pain and underwent coronary CTA which demonstrated ACAOS of the right coronary artery. **a** An oblique sagittal maximum intensity projection image demonstrates the acute angle of takeoff and a slit-like orifice of the anomalous right coronary artery (*arrowhead*) as it courses between the aorta and the pulmonary artery. **b** The oblique coronal multiplanar reformat demonstrates lateral luminal compression of the anomalous right coronary artery (*arrow*), suggestive of an intramural course. **c** The luminal shape and diameter are restored distally (*arrow*) as it exits the intramural portion. This ACAOS with an interarterial course is considered malignant and is associated with increased risk of sudden cardiac death. *Ao* aorta, *PA* pulmonary artery

occur in the mid-segment of the left anterior descending artery (Fig. 13) [47]. Most patients are asymptomatic; a small subgroup develops ischemia. The cause of ischemia is uncertain as the majority of coronary blood flow occurs during diastole and compression of the myocardial bridge occurs during systole. Interestingly, the tunneled segment of the coronary artery exhibits less atherosclerosis compared to anatomically normal coronary arteries [48]. However, the coronary segment proximal to the myocardial bridge demonstrates a higher rate of plaque development [49•], implicating sheer forces as the likely culprit. However, the mechanism for proximal atherosclerosis is still incompletely understood.

The prevalence of myocardial bridges is difficult to define precisely for many of the same reasons alluded to earlier

Table 3 Proposed mechanisms for reduced coronary flow in ACAOS with an interarterial course

High-risk anatomical feature of ACAOS	Mechanism for myocardial ischemia
Presence of a coronary ostial ridge	May function as a valve, restricting flow during exertion
Acute angulation at coronary takeoff	During exercise, expansion of the aortic and pulmonary roots increases angulation of the ostium leading to coronary kinking
Slit-like orifice of a coronary ostium	Becomes compressed by exercise-induced aortic dilation
Intramural coronary course	Lateral luminal compression that worsens during exertion

ACAOS anomalous origin of a coronary artery from the opposite sinus

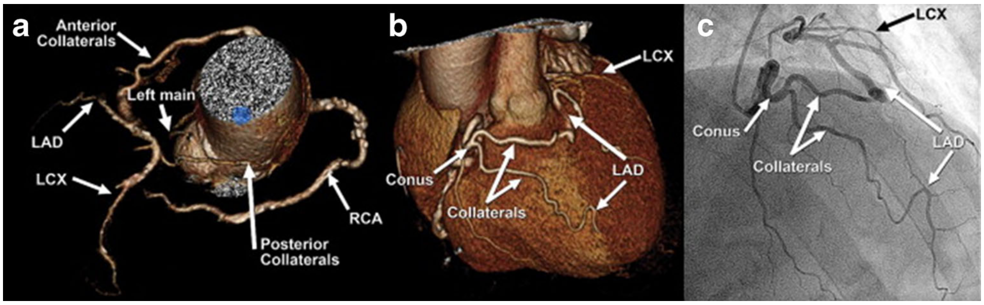

Fig. 12 A 66-year-old male presented with new-onset exertional chest pain. His exercise stress test was positive for ischemia and he was referred for coronary CTA. **a** A three-dimensional volume-rendered image of the coronary tree demonstrates an atretic left main coronary artery. Collateral vessels arising from the right coronary artery supply the left coronary artery system. **b** A three-dimensional whole heart volume-rendered image demonstrates collateral vessels (*arrows*) from the conus branch supplying segments of the left anterior descending artery (*arrows*). Image reprinted with permission [45]. *RCA* right coronary artery, *LCX* left circumflex coronary artery, *LAD* left anterior descending artery. c The invasive coronary angiogram confirms the atretic left main coronary artery. A selective injection of the conus branch demonstrates collaterals to the mid- and distal LAD with retrograde filling of the proximal vessel and LCX

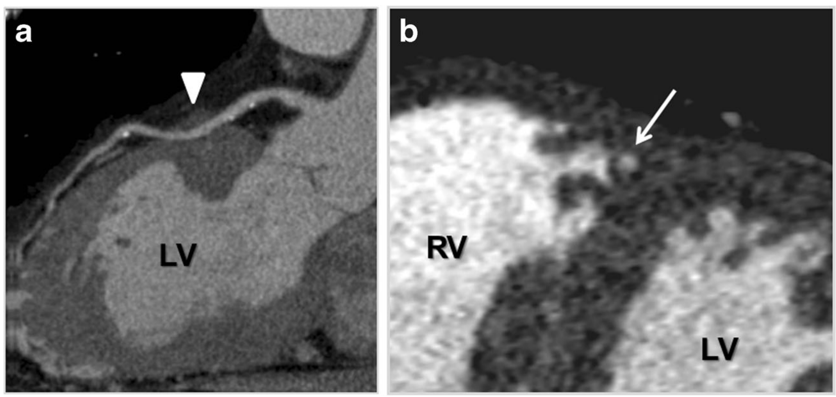

Fig. 13 A 67-year-old female presented with atypical, non-exertional chest pain and underwent coronary CTA. **a** An oblique sagittal image demonstrates a left anterior descending artery bridge as it courses deeply into the myocardium (*arrowhead*). **b** A cross-sectional thin multiplanar reformat of the left anterior descending artery bridge demonstrates its depth (*arrow*) within the myocardium. *LV* left ventricle, *RV* right ventricle

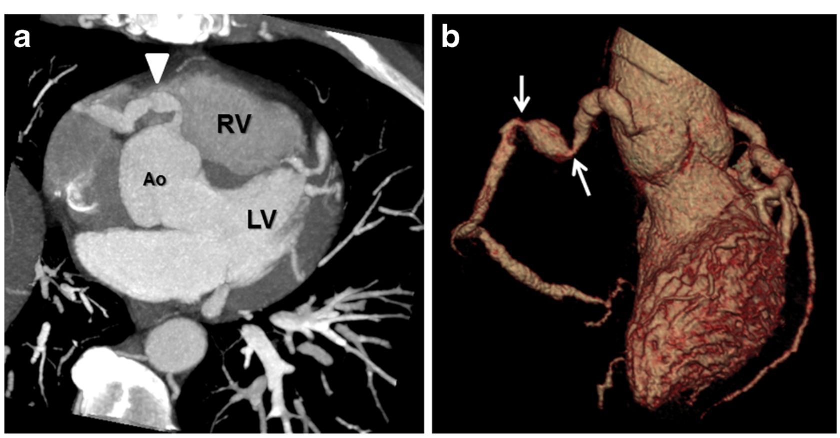

Fig. 14 A 37-year-old male with Kawasaki disease underwent coronary CTA. **a** A maximum intensity projection image demonstrates multiple sequential aneurysms, most notably in the proximal right coronary artery without atherosclerosis (*arrowhead*). **b** A three-dimensional volume-rendered image demonstrates the proximal RCA aneurysms with discrete stenoses between aneurysmal segments (*arrows*). *Ao* aorta, *LV* left ventricle, *RV* right ventricle

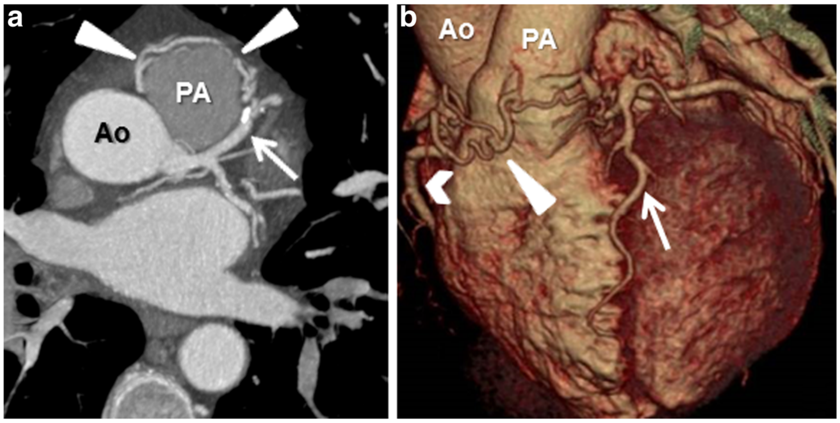

Fig. 15 A 65-year-old female presented with dyspnea with subsequent evaluation by coronary CTA. **a** An axial maximum intensity projection image demonstrates the left anterior descending artery (*arrow*) supplying feeder vessels (*arrowheads*) to the pulmonary artery. **b** A three-dimensional volume-rendered image illustrates the feeder vessels (*arrowhead*) to the pulmonary artery from the left anterior descending artery (*arrow*) and the right coronary artery (*chevron*). *PA* pulmonary artery, *Ao* aorta

regarding selection bias. However, until recently, myocardial bridges were diagnosed by either autopsy or CCA. CCA demonstrates only modest sensitivity in diagnosing myocardial bridges as compared to autopsy series [50, 51]. Coronary CTA has produced similar prevalence estimates relative to autopsy series, ranging from approximately 14 to 30 % [52–55], secondary to its ability to visualize not only the coronary lumen but the overlying myocardium as well.

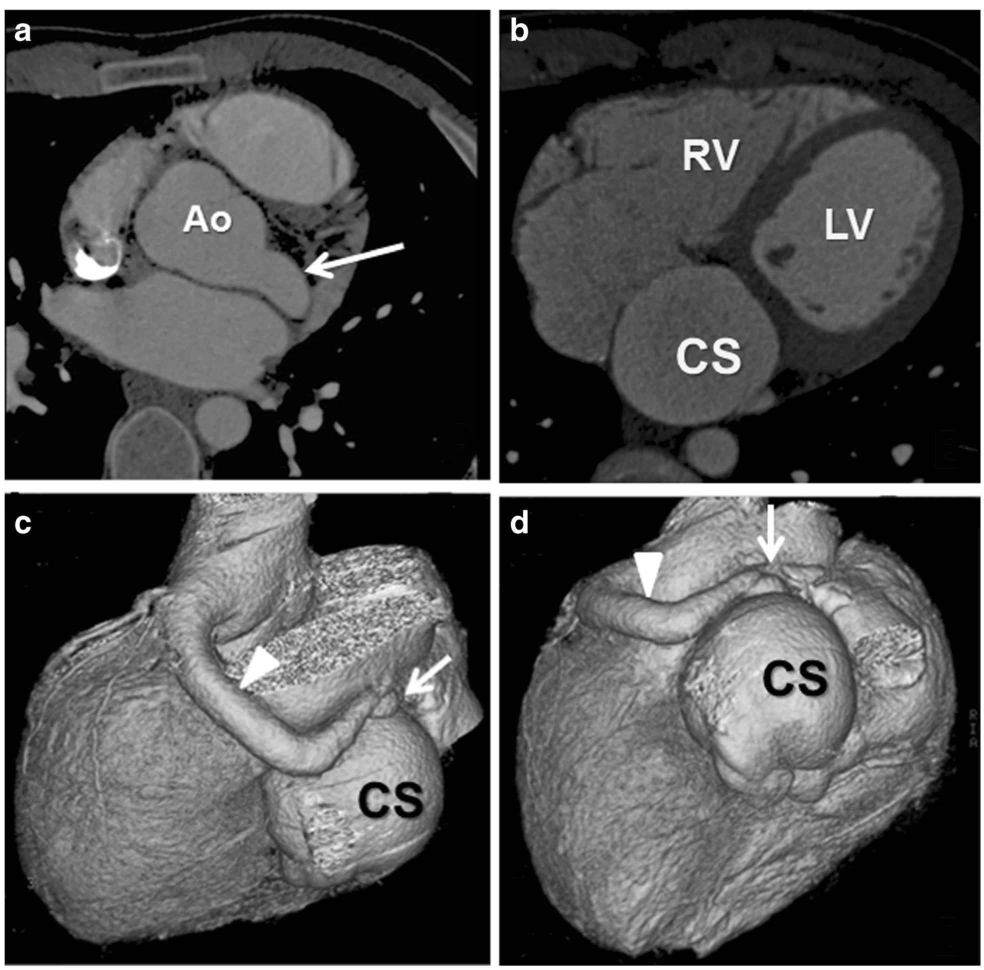

Fig. 16 A 68-year-old male presented with dyspnea and an abnormal transthoracic echocardiogram. He was subsequently referred for a coronary CTA. **a** An axial maximum intensity projection image at the level of the aortic root demonstrates a massively dilated proximal left circumflex (*arrow*). **b** A more caudal slice demonstrates an aneurysm secondary to a left circumflex to coronary sinus fistula. **c**, **d** Three-dimensional volume-rendered images demonstrate the dilated left circumflex (*arrowhead*) and fistulous connection (*arrow*) to the coronary sinus. *CS* coronary sinus, *Ao* aorta, *LV* left ventricle, *RV* right ventricle

Coronary Aneurysms

Coronary aneurysms are defined as dilated coronary segments that are 1.5 times the diameter of the reference segment. The prevalence of coronary aneurysms was reported to be 1.4 % in a population referred for coronary angiography. This same cohort demonstrated a high prevalence (83 %) of obstructive coronary artery disease [56]. Indeed, atherosclerosis is the most common etiology for coronary aneurysms in the USA. Worldwide, Kawasaki disease remains the most common cause of coronary aneurysms. Approximately 25 % of patients with untreated acute Kawasaki disease develop coronary artery aneurysms (Fig. 14), with giant aneurysms at risk of developing mural thrombi and/or rupture [57]. Coronary CTA can be extremely useful at defining these aneurysms and associated complications. Other causes of coronary artery aneurysms include congenital, iatrogenic, traumatic, mycotic, embolic, and connective tissue diseases.

Anomalies of Termination

Coronary Artery Fistulas

Coronary artery fistulas are abnormal communications between a coronary artery and the pulmonary artery (Fig. 15) or coronary sinus (coronary arteriovenous fistula) (Fig. 16), or between a coronary artery and any cardiac chamber (coronary cameral fistula). Similar to ALCAPA, coronary drainage into a lower-pressure structure can lead to a steal phenomenon and ischemia. Small- to moderate-sized fistulas are generally asymptomatic and associated with a good prognosis. About half of affected adults develop dyspnea (high-output heart failure) and/or chest pain. Children are rarely symptomatic. Treatment is reserved for fistulas that either cause symptoms or are associated with large aneurysms. Management strategies for hemodynamically significant coronary artery fistulas include transcatheter approaches, surgical ligation, and/or medical therapy [58].

Conclusion

Coronary anomalies are an important contributor to SCD, particularly in athletes and young adults, and thus represent an important clinical entity. While most coronary anomalies are benign, it is important to be aware of potentially malignant varieties, as identification is likely to inform patient management. ACAOS with an interarterial course is the most common of the malignant coronary anomalies. Historically, CCA has been considered the gold standard for evaluation of coronary anomalies. However, defining the origin and course of the anomalous coronary remains challenging with this technique. With refinements in technology, coronary CTA has emerged as an accurate and robust non-invasive method to define the full spectrum of coronary artery anomalies. Given the three-dimensional nature of this technique, the origin, course, and termination of anomalous coronary arteries are easily delineated relative to other important anatomic structures, such as the great vessels. Given its utility for the evaluation of patients with anomalous coronary arteries, recommendations for coronary CTA have been incorporated into appropriate use criteria and national guidelines.

Compliance with Ethics Guidelines

Conflict of Interest J McLarry declares no conflicts of interest.

M Ferencik has received research grants from the American Heart Association (13FTF16450001).

MD Shapiro declares no conflicts of interest.

Human and Animal Rights and Informed Consent This article does not contain any studies with human or animal subjects performed by any of the authors.

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- • Of importance
- •• Of major importance

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