Coronary CTA assessment of coronary anomalies

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Abstract. Coronary anomalies occur in <1% of the general population and can range from a benign incidental finding to the cause of sudden cardiac death. The coronary anomalies are classified here according to the traditional grouping into those of origin and course, intrinsic arterial anatomy, and termination. Classic coronary anomalies of origin and course include those in which a coronary artery originates from the contralateral aortic sinus or the pulmonary artery with anomalous course. Single coronary artery anomalies, in which single coronary artery branches to supply the entire coronary tree, are also included in this category. Anomalies of intrinsic arterial anatomy are a broad class that includes myocardial bridges, coronary ectasia and aneurysms, subendocardial coursing arteries, and coronary artery duplication. Coronary anomalies of termination are those in which a coronary artery terminates in a fistulous connection to a great vessel or cardiac chamber. In the case of those anomalies associated with a risk of sudden cardiac death, the relevant imaging features on CT angiography (CTA) associated with poorer prognosis are reviewed. Recent guidelines and appropriateness criteria favor the use of coronary CTA for the evaluation of coronary anomalies. Although invasive angiography has historically been used to diagnose coronary anomalies, multidetector CT imaging techniques have now become an accurate noninvasive alternative. Cardiac CTA provides excellent spatial and temporal resolution, allowing accurate anatomical assessment of these anomalies.

Introduction

Because of its ability to accurately depict the anatomy of the heart and thorax, coronary computed tomography...
angiography (CTA) has been deemed appropriate for evaluation of coronary anomalies. In the 2010 guidelines from the American College of Cardiology and American Heart Association (ACC/AHA) for the management of adults with congenital heart disease, the use of coronary CTA or magnetic resonance angiography is a Class I recommendation.

Figure 1  Absent left main: 65-year-old woman presenting with dyspnea on exertion. Thin maximum intensity projection image (A) and volume rendered 3-dimensional reconstruction (B), showing the left anterior descending artery (arrow) and left circumflex artery (arrowhead) arising separately from the left sinus of Valsalva.

Figure 2  Bland-White-Garland Syndrome (anomalous left coronary artery originating from pulmonary artery; ALCAPA): 1-year-old infant presenting with episodic nighttime diaphoretic spells was found to have severe systolic failure on echocardiogram referred for coronary evaluation. (A) Maximum intensity projection image depicting left anterior descending artery (LAD) originating from the pulmonary artery (PA), and (B) 4-chamber view showing significant left ventricular chamber enlargement. Ao, aorta; LA, left atrium; LV, left ventricle; RV, right ventricle; RVOT, right ventricular outflow tract.

Figure 3  Anomalous right coronary artery arising from pulmonary artery (ARCAPA): 52-year-old woman presenting with angina. Coronal and axial maximum intensity projection images (A and B) and corresponding 3-dimensional volume-rendered image (C) showing a dilated and aneurysmal right coronary artery (white arrow) originating from the main pulmonary artery. The left coronary artery originates as usual from the left aortic sinus (black arrow).
recommendation for initial screening of congenital coronary anomalies of ectopic origin in centers with expertise in such imaging.1,2

In this pictorial essay the classification scheme and coronary CTA features of a comprehensive array of coronary anomalies are reviewed, and the relative strengths and weaknesses of coronary CTA relative to cardiac catheterization and other imaging modalities are discussed.

Definitions: Coronary anomalies versus coronary variants

Any anatomic or morphologic finding found in >1% of the general population is defined as normal. A normal variant describes an alternative, relatively unusual finding that is observed in >1% of the same population.3 An anomaly is a morphologic feature seen in <1% of the general population.3,5

Figure 4  Left circumflex artery from the left ventricular outflow tract: 55-year-old man presenting with hypertension and prior coronary bypass surgery underwent 64-slice multidetector CT to evaluate chest pain. (A) Oblique coronal left ventricular outflow view of the aortic valve (arrow 1) and anterior mitral leaflet (arrow 2). Notice how the anomalous circumflex artery arises from within the left ventricular outflow tract inferior to the aortic valve leaflet (arrow 1). As it leaves the left ventricular outflow from a subaortic origin, it passes over the great cardiac vein (arrow 4) in the posterior atrioventricular ring. The right pulmonary artery (RP) is labeled. (B) Surface-rendered 3-dimensional reconstruction of the left ventricle and aortic root, viewed from behind. The anomalous origin of the left anterior descending coronary artery (LAD; arrow 1) is just to the left of the origin of the right coronary artery (arrow 2) from the anterior aortic sinus of Valsalva. Notice the severe stenosis (arrowhead) of the proximal LAD. The LAD follows a course with that of the great cardiac vein and gives off marginal branches to the posterior wall of the left ventricle. The circumflex artery (arrow 3) arises anomalously from the left ventricle (LV), immediately inferior to the posterior left (pl) aortic sinus of Valsalva, and then extends into the posterior atrioventricular ring (arrow 4). Ao, aorta; LV, left ventricle; RA, right atrium.

Figure 5  Right coronary artery (RCA) from above sinotubular junction: 48-year-old man with hyperlipidemia for evaluation of angina symptoms. Right coronary artery (arrow) is seen originating from above the sinotubular junction. (A) Three-dimensional view of the coronary tree and surface-rendered image displaying origin site of RCA above right sinus of Valsalva. (B) Coronal thin maximum intensity projection image shows the origin of RCA from the ascending aorta above the sinotubular junction.
Normal variants of coronary artery anatomy are benign entities with limited clinical significance. In contradistinction, coronary artery anomalies range from benign entities to those associated with a high risk of sudden cardiac death. In the United States, coronary artery anomalies are the second most common cause of sudden death in competitive athletes after hypertrophic cardiomyopathy.

The vast array of coronary anomalies has propagated multiple complex classification schemes although the traditional schematic divides them into anomalies of (I) origin and course, (II) intrinsic coronary anatomy, and (III) termination. Although describing every anomaly is beyond the scope of this review, characteristic coronary CTA examples and features from each category are discussed.

I. Anomalies of origin and course

Most coronary anomalies comprise this group, some of which are of considerable clinical importance. The 3 main subcategories include (A) absent left main (LM) artery, (B) anomalous ostium outside of the aortic sinuses, and (C) anomalous ostium at an improper sinus.

A. Absent left main

Absent LM, or split origin of the left coronary artery (LCA), is typically a benign entity in which the left circumflex (LCX) and left anterior descending (LAD) arteries each have separate ostia from the left coronary sinus of Valsalva. The vast array of coronary anomalies has propagated multiple complex classification schemes although the traditional schematic divides them into anomalies of (I) origin and course, (II) intrinsic coronary anatomy, and (III) termination. Although describing every anomaly is beyond the scope of this review, characteristic coronary CTA examples and features from each category are discussed.

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Figure 6  Right coronary artery from left sinus of Valsalva with interarterial course (Type 2A): 14-year-old boy with presyncope after exercise and abnormal stress test. (A) Volume-rendered 3-dimensional image showing anomalous origin of the right coronary artery (RCA) from the left sinus with interarterial course. (B) Multiplanar-reformatted image in oblique axial view shows acute take-off of the RCA from the aorta. (C) Multiplanar-reformatted image in oblique coronal view shows slit-like opening of the origin of the RCA, suggestive of possible intramural course. Coronary CT angiography imaging performed with prospective electrocardiogram-triggered high-pitch spiral acquisition (Somatom Definition FLASH; Siemens Healthcare, Forchheim, Germany) with estimated radiation dose of 0.3 mSv.

Figure 7  Left coronary artery from right sinus of Valsalva with intraseptal course (Type 2B): 53-year-old woman with atypical chest pain. (A) Volume-rendered 3-dimensional image with pulmonary trunk obscuring the left main (LM) because of the intraseptal course. (B) Volume-rendered 3-dimensional image with pulmonary trunk removed to show the course of the LM. (C) Multiplanar-reformatted image in an oblique sagittal view shows the intraseptal course of the LM that courses below the pulmonic and aortic valves, between the right and left ventricular outflow tracts. LAD, left anterior descending artery; LCX, left circumflex artery; RCA, right coronary artery.
population and is associated with a higher incidence of left coronary dominance and myocardial bridging. Clinically, it is important to recognize this anomaly to prevent complications in patients undergoing cardiac surgery who require selective coronary cardioplegia such as aortic valve replacement. Although inability to separately cannulate the LCX or LAD may result in the erroneous assumption that the vessel is absent or occluded on invasive coronary angiography, the anatomic display of coronary CTA makes this diagnostic error uncommon.

B. Anomalous coronary ostium outside of the aortic sinuses

This subcategory includes a large subset of coronary anomalies, including origin of the LCA from the pulmonary artery (ALCAPA). ALCAPA is rare, affecting roughly 1 in 300,000 births, and typically results in significant myocardial ischemia due to a “steal phenomenon.” Blood flow is directed from the higher pressure right coronary artery (RCA) system to the lower-pressure pulmonary system via RCA-to-LCA collateral vessels. Hence, there is retrograde flow in the left coronary circuit routing blood toward the lower resistance pulmonary circulation. In summary, blood flows from aorta → RCA → collateral vessels → LCA → low pressure pulmonary circuit. The resultant chronic myocardial ischemia is theorized to cause cardiomyopathy and heart failure.

Approximately 90% of patients with ALCAPA die within the first year of life as the result of heart failure, ischemia, and ventricular arrhythmias. There have been

Figure 8  Left circumflex from the right sinus of Valsalva with posterior course (Type 3): 54-year-old man with hypertension and hyperlipidemia presenting with anginal symptoms. (A) 2D thin maximum intensity projection and (B) 3D volume rendered image demonstrating the left circumflex (LCX) arising from the right sinus of Valsalva, adjacent to the right coronary artery (RCA), with a posterior course behind the great vessels to reach its normal anatomic position. LAD, left anterior descending artery.

Figure 9  Right coronary artery from noncoronary sinus of Valsalva: 73-year-old man with history of coronary artery disease presenting with chest discomfort. (A) Axial multidetector CT images showing the anomalous origin of the right coronary artery (RCA) from the posterior aortic coronary sinus (PCS) and the origin of the left main coronary artery (LMCA) from the left coronary sinus (LCS). There is no artery arising from the right coronary sinus (RCS). (B) Cranial 3-dimensional volume-rendering coronary-tree reconstructions and maximum intensity projection 3-dimensional globe view showing the anomalous origin of the RCA from the PCS. (C) Cranial virtual angioscopy view showing the ostium of the RCA (arrow) at the PCS. There is no ostium at the RCS. The normal ostium of the left main coronary artery (arrow) is present at the LCS. LAD, left anterior descending artery.
several reported cases of late presentation of ALCAPA in adults. This delayed presentation is probably because of the extensive collateral circulation provided by the RCA. A rarer anomaly, in which the RCA originates from the pulmonary artery (ARCAPA), was previously reported by our laboratory.12

Coronary CTA evaluation of ALCAPA and ARCAPA (Fig. 2 and Fig. 3; Movie 2) necessitates assessment of the origin of the anomalous artery from the pulmonary artery, the collateral vessels network, and ventricular size and function.

Additional sites of anomalous origin at locations above or below the aortic sinuses include the left or right ventricular outflow tracts, the ascending aorta, and the brachiocephalic or subclavian arteries (Fig. 4 and Fig. 5; Movies 3 and 4). The clinical sequelae of these coronary anomalies remain uncertain.

C. Anomalous location of coronary ostium at improper sinus

This subcategory comprises anomalies associated with important clinical sequelae including coronary ischemia and sudden cardiac death, depending on the anomalous coronary artery’s course. The course may be anterior to the pulmonary artery (Type 1); interarterial, between the aorta and pulmonary artery (Type 2A) (Fig. 6; Movies 5 and 6); septal or intramyocardial (between the aorta and pulmonary artery but with an intraseptal component) (Type 2B) (Fig. 7); or posterior to the great vessels (Type 3) (Fig. 8). Type 2A is considered to be a malignant subtype, with a high risk of sudden cardiac death, especially when the LCA originates from the right coronary sinus.

Theories about this anomaly’s malignant potential include ischemia from compression of the vessel by the great arteries.
vessels at the interarterial component of the artery, acute take-off from the aortic wall, an associated ostial ridge, a slit-like ostium, intramural (inside the wall of the aortic root) course, superimposed spasm, intimal plaque and clotting\(^{13}\) (Fig. 6; Movies 5 and 6).

Coronary CTA is an excellent tool to identify the origin and course of the anomalous coronary, enabling assessment of the angle of take-off, size, and course of the anomalous vessel (Figs. 6–8).

Origination of the RCA from the left sinus of Valsalva can be a benign entity. However, a malignant interarterial course, an intramural origin, or evidence of ischemia on stress testing may necessitate surgical intervention. Other anomalies with unclear clinical significance include origination of the RCA or LCA from the noncoronary (posterior) sinus of Valsalva (Fig. 9 and Fig. 10; Movies 7 and 8).

**D. Single coronary artery**

Single coronary artery is among the rarest of coronary artery anomalies\(^{14,15}\) yet this subgroup has its own coding system that is defined by the originating sinus and relation between the 3 main coronary arteries. The first letter in the 3-letter coding scheme refers to the sinus of Valsalva from which the single coronary artery originates [ie, left (L) or right (R)]. Thereafter a Roman numeral (I, II, or III) describes the anatomical course of the artery. In group I, the artery follows the anatomic course of LCA or RCA (Fig. 11; Movies 9 and 10). In group II, one coronary artery arises from the proximal part of the normally located other coronary artery (Fig. 12; Movies 11 and 12). In group III, the LAD and LCX each arise separately from the normally positioned RCA (Fig. 13; Movie 13). The last letter refers to the relation of the coronary arteries to the great vessels [ie, anterior (A), posterior (P), between the great vessels (B), and combined (C)].\(^{14–16}\)

**Figure 12** Single coronary artery (Type L-IIA): 45-year-old man presenting with chest pain in the setting of an intermediate likelihood of coronary artery disease. (A) Thin maximum intensity projection oblique axial image showing single coronary artery arising from the left sinus of Valsalva with right coronary artery arising from the mid left anterior descending. (B and C) Volume-rendered 3-dimensional images showing origin from the left coronary sinus with origin of the right coronary artery (RCA) from the mid left anterior descending artery (LAD) and coursing anterior to the main pulmonary artery.

**Figure 13** Single coronary artery (Type R-IIIC): 35-year-old woman presenting with atypical angina symptoms. (A) Volume-rendered 3-dimensional image showing large single right coronary artery arising from right coronary sinus which trifurcates proximally to give rise to an anterior branch that supplies the mid and distal left anterior descending artery (LAD) territory, a posterior branch that supplies the left circumflex (LCX) and proximal LAD territory, and normal course of the right coronary artery (RCA). There is also a very small septal branch that courses between the great vessels. (B) Volume-rendered 3-dimensional image in the superior-anterior view displaying the proximal LAD supplied by the posterior branch and the mid and distal LAD supplied by the anterior branches from the single coronary artery. (C) Posterior view showing the course of the posterior branch that supplies the circumflex and proximal LAD territories.
A single coronary artery is thought to be associated with a risk of sudden cardiac death, even in patients without a malignant arterial course. Demonstration of significant ischemia in a symptomatic patient therefore warrants further clinical attention.\textsuperscript{15}

II. Anomalies of intrinsic coronary anatomy

A. Congenital ostial stenosis or atresia

Most cases of ostial atresia involve the LM artery (Fig. 14), with subsequent LM developmental failure. The LAD and LCX subsequently fill in retrograde fashion via collaterals from the RCA circulation. This anomaly is distinguished from single coronary artery in that the flow is centripetal (from smaller peripheral vessels to larger central vessels) and often from multiple small collaterals in ostial atresia as opposed to centrifugal (from larger to progressively smaller vessels) in single coronary artery.\textsuperscript{17} This collateral circulation is usually incapable of providing adequate myocardial perfusion, resulting in failure to thrive, early myocardial ischemia and infarction, and a poor prognosis without surgical revascularization.\textsuperscript{18}

B. Coronary ectasia and aneurysm

Coronary artery ectasia or aneurysm is defined as an arterial segment with a diameter $>$1.5 times the normal adjacent artery segment. Coronary artery ectasia also commonly involves $>$50\% of the total length of the coronary artery. The most common worldwide cause of coronary artery aneurysm is Kawasaki’s disease, although

Figure 14  Left main atresia: 66-year-old man without significant past medical history presented with new onset exertional chest pain and positive exercise stress test. (A) Volume-rendered 3-dimensional image from superior projection showing the small, atretic left main and small anterior and posterior collateral vessels originating from the right coronary artery (RCA) to supply the left coronary artery system. (B) Volume-rendered 3-dimensional image shows collateral vessels from the conus branch crossing anteriorly to supply segments of the left anterior descending artery (LAD). (C) Corresponding invasive coronary angiogram still frame with selective injection of the conus branch shows collaterals to the LAD with retrograde filling of the proximal LAD and circumflex (LCX) arteries.

Figure 15  Coronary artery aneurysm: 19-year-old woman with Behçet disease and history of peripheral artery aneurysm. (A) Volume-rendered 3-dimensional image of the heart showing large coronary aneurysm (black arrow) involving the proximal left anterior descending artery. (B) Oblique sagittal image showing the significant amount of the intraluminal thrombus (asterisk) present within the left anterior descending artery aneurysm.
Atherosclerotic coronary artery disease accounts for most cases in the United States. In one study of nearly 5000 angiograms, significant coronary artery dilation or ectasia was noted in 1.4% of patients, with more frequent RCA than LCA involvement. Coronary CTA allows a more accurate assessment of aneurysm size and degree of thrombus and calcification than invasive angiography (Fig. 15; Movies 14 and 15).

C. Myocardial bridging
Myocardial bridging refers to the intramyocardial course of a portion of the normally epicardially positioned coronary artery. Although typically a benign finding, myocardial bridging has been associated with angina thought to be secondary to myocardial ischemia, myocardial infarction, arrhythmias, and sudden cardiac death. The cause of this relation is unclear, although it may be that the presence of an intramyocardial bridge leads to increase shearing forces at the segment proximal to the bridge which predispose to atherosclerotic plaque formation. This plaque may be responsible for the observed symptoms. Coronary CTA can clearly delineate the intramyocardial segment of the coronary artery (Fig. 16; Movie 16). Electrocardiographic-gated reconstruction can be performed to evaluate the degree of luminal narrowing during systole.

D. Duplicated arteries
The most common of this group of anomalies is a duplication of the LAD (Fig. 17), in which there is a short LAD that travels in the anterior interventricular groove, terminating prematurely before reaching the apex, and running parallel to a longer LAD that continues to supply the apical wall.

E. Subendocardial coronary course
In rare cases, a coronary artery may pursue a subendocardial course after penetrating the myocardium. This anomaly

![Figure 16](image1.png)
![Figure 17](image2.png)
can be viewed as an intermediate stage in coronary malposition from intramyocardial to coronary-cameral fistula.4

E. Coronary crossing

Although coronary arteries and their branches typically run parallel to one another, published case reports have described anomalous crossing of coronary branches and, less commonly, of the LAD and LCX (Fig. 18; Movies 17 and 18). This is typically seen as a benign incidental finding on conventional angiography or coronary CTA.

III. Anomalies of coronary termination

Coronary artery fistulas

Coronary artery fistulas are communications between ≥1 coronary arteries and a cardiac chamber (coronary-cameral) (Fig. 19; Movie 19), the pulmonary artery (Fig. 20; Movies 20 and 21), or a venous structure such as the coronary sinus or superior vena cava. They are identified in roughly 0.15% of patients undergoing angiography.22 Most cases involve the RCA, and the feeding artery is typically dilated and tortuous, given that it drains into a lower pressure chamber (typically right atrium or right ventricle). Coronary artery fistulas can result in myocardial ischemia via a hemodynamic steal phenomenon.

Strengths and weakness of coronary CTA in coronary anomalies

The strengths of coronary CTA over invasive coronary angiography are its noninvasive nature and minimal risk that it imposes to the patient. Coronary CTA enables accurate assessment of the entire heart, coronary artery system, and thorax, providing important 3-dimensional information about the spatial relations of the anomalous vessels and surrounding intraluminal and extraluminal anatomy and disease, and thereby contributing clinically important prognostic information. A limitation of coronary CTA compared with imaging modalities such as magnetic resonance angiography is its use of ionizing radiation. However, spatial resolution and acquisition speed are significantly improved with coronary CTA compared with magnetic resonance angiography, and newer techniques of radiation dose reduction allow radiation exposures of <1 mSv, while retaining excellent spatial resolution (Fig. 6; Movies 5 and 6).

Conclusions

Coronary CTA provides excellent spatial and temporal resolution, 3-dimensional depiction, and a large field of
view that provides reference landmarks for accurate anatomical assessment of coronary anomalies.

**Supplementary data**

Supplementary data related to this article can be found online at doi:10.1016/j.jcct.2011.06.009.

**References**


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