Different Paths to a Beating Heart: A Pictorial Review of Coronary CT Angiography of Congenital Coronary Artery Anomalies

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Learning objectives

1. Review coronary artery anatomy and normal anatomical variants.
2. Provide a list of the most common congenital coronary artery anomalies.
3. Discuss the role of coronary CT angiography in the diagnosis of such anomalies.
4. Describe pertinent imaging findings on coronary CT angiography with multiplanar three-dimensional reconstructions.
5. Discuss the clinical significance of an accurate diagnosis in the management of these anomalies.
Background

Coronary Artery Anomalies (CAA) are a rare condition that can be found as an isolated finding or in conjunction with congenital heart disease. Many of the CAA are considered to be of no clinical significance. Only 20% of them have been found to be clinically significant, usually presenting with angina, myocardial infarction or sudden cardiac death. CAA is the second most common cause of sudden cardiac death among young athletes, after hypertrophic cardiomyopathy. Historically, conventional angiography was the gold standard for evaluating CAA. However, it has been recently demonstrated that Coronary Computed Tomography Angiography (CCTA) improves diagnostic accuracy and has emerged as the standard for diagnosis [7].

Anatomy:

There are two main coronary arteries that arise from the coronary sinuses in the aortic root: the right coronary artery (RCA) arises from the right coronary sinus and the left coronary artery (LCA) arises from the left coronary sinus. There is a posterior coronary sinus which usually does not give rise to a coronary artery, hence termed the noncoronary sinus.

The RCA arises inferior to the LCA and courses through the right atrioventricular groove giving off several branches. The first branch after its origin is the sinus-atrial nodal branch, which supplies the SA node. It then gives off the right marginal branch that supplies the right border of the heart. The RCA then courses to the posterior surface of the heart giving off the atrioventricular nodal branch and, in most people, the posterior descending artery (PDA). The LCA bifurcates into the left anterior descending artery (LAD) and left circumflex artery (LCX). The LAD courses along the anterior interventricular groove supplying both ventricles and the anterior two thirds of the interventricular septum. The LCX courses along the left atrioventricular groove giving off obtuse marginal branches that supply the left ventricle.

Anatomical Variants:

Several anatomic variants of the branching pattern of the coronary arteries exist. The dominance of the coronary circulation is determined by which artery gives rise to the PDA. The most common pattern is a right dominance pattern, observed in 85% of the population, in which the PDA is a branch of the RCA. The left dominant pattern is seen in 7-8% of population, and in such cases the PDA arises from the LCX. The remaining 7% of the population has been described as being co-dominant [2].
Classification Scheme:

CAA can be classified based on the anatomy (anomalies of origin, course or termination) or by the clinical significance (hemodynamically significant or not hemodynamically significant). Hemodynamically significant anomalies have been associated with shunting, ischemia or sudden cardiac death and include: atresia, origin from the pulmonary artery, interarterial course and congenital fistula [6]. Not hemodynamically significant anomalies include: duplication, origin anomalies, systemic termination, origin from the aorta in anomalous position, prepulmonic, transseptal or retroaortic course, among others.

The anatomy classification scheme divides them by [2]:

1. **Anomalies of origin**: High origin, multiple ostia, single coronary artery, anomalous origin of coronary artery from pulmonary artery.
2. **Anomalies of course**: Myocardial bridging, duplication of arteries.
3. **Anomalies of termination**: Coronary artery fistula, extracardiac termination, coronary arcade.
Findings and procedure details

I. Anomalies of Origin

A. High Origin

High origin CAA refers to the RCA or LCA arising 10mm above the sinotubular junction of the ascending aorta (Fig. 1 on page 9, Fig. 6 on page 13). It occurs most commonly on the RCA than the LCA, and is seen in patients with bicuspid aortic valve [6]. The importance of diagnosing a high origin CAA is that it may difficult coronary artery catheterization. Additionally, if aortic valve surgery or aortotomy is going to be performed, the ascending aorta should not be clamped inferior to the high origin coronary artery because it may lead to cessation of cardiac activity [6].

B. Multiple Ostia

Multiple ostia can occur when the RCA and the conus branch arise from separate locations at the coronary sinus. It can also be seen in patients were the LAD and LCX arise from separate locations and there is no LCA (Fig. 2 on page 9). Multiple ostia represent a challenge for angiography and a conus branch that arises separately from the RCA is at risk for injury on heart surgery if not previously identified. If the LAD and LCX arise separately it provides a small benefit if proximal atherosclerosis occurs [2].

C. Single Coronary Artery

The anomalous origin of a single coronary artery from the coronary sinus may have the initial course of either the RCA or LCA, and then it divides into two branches that follow the RCA and LCA course. It has been described that if the single coronary artery has an interarterial course (between the aorta and pulmonary artery) it increases the risk for sudden cardiac death. Another possible complication of a single coronary artery is the inability to form collateral vessels if proximal stenosis occurs [2].

D. Anomalous Origin of Coronary Artery from Pulmonary Artery

The most common presentation of this disease is the Bland Garland White Syndrome or ALCAPA (Anomalous Left Coronary Artery arising from Pulmonary Artery) in which the LCA arises from the pulmonary artery (PA) and the RCA arises from the right coronary sinus. This condition occurs in 1/300,000 births and can have two presentations
depending on the development of collaterals between the RCA and LCA. The infantile type shows no collateral vessels (Fig. 3 on page 10) and presents in the first weeks of life with failure to thrive, dyspnea, pallor or chest pain [7]. The adult onset type is known to have collateral vessels between the RCA and LCA (Fig. 4 on page 11) presenting with myocardial infarction, left ventricular dysfunction or mitral regurgitation [4]. As pulmonary resistance falls, the blood flows from the aorta to the RCA, into the LCA in a retrograde fashion via collaterals, and then to the PA creating a coronary steal phenomenon in which there is a left to right shunt [4]. On CT imaging findings the LCA is seen arising from the main pulmonary artery. Other imaging findings include dilated and tortuous appearance of the LCA and RCA, as well as dilated intercoronary collateral arteries.

II. Anomalies of Course

A. Myocardial Bridging

Myocardial bridging occurs when the coronary arteries pass along a tunnel of myocardium instead of running on the epicardial surface (Fig. 5 on page 12). Myocardial bridging can be incomplete in which the arterial segment is partially surrounded by myocardium or complete in which the arterial segment is completely surrounded by the myocardium [7]. Myocardial bridging is most commonly seen on the middle segment of the LAD. This condition is often clinically silent, but some patients may present with symptoms of angina, myocardial infarction, arrhythmias or death. Symptoms are believed to be the result of the direct compression on the coronary artery during systole or by the formation of atherosclerosis proximal and distal to the tunneled segment which is protected from the development of atherosclerosis [3].

B. Interarterial Course

Interarterial course refers to a coronary artery passing between the aorta and the main pulmonary artery (Fig. 6 on page 13). When a coronary artery arises from the opposite cusp or from the contralateral coronary artery it takes an interarterial course between the aorta and pulmonary arterial trunk. This is the most common clinically significant coronary artery anomaly; it is the second cause of sudden cardiac death among young athletes [6]. An interarterial course is more commonly seen when the LCA arises from the right coronary cusp and also carries a higher mortality rate when compared to an interarterial course of the RCA [7]. Several theories have been proposed to explain the association between sudden cardiac death and interarterial course. On physical exertion, there may be direct compression of the interarterial artery by the aorta and pulmonary artery. It has also been proposed that interarterial arteries have a "slit-like" orifice that is more prone to occlusion [6].
C. Transseptal Course

When coronary arteries have an abnormal origin including origin from the contralateral coronary artery or opposite coronary sinus, they may have a transseptal course along the upper interventricular septum (Fig. 7 on page 14). Several characteristics can help to differentiate the transseptal vs interarterial course. The transseptal course artery is covered in myocardium, in contrast to the interarterial course artery which courses along the epicardial surface. The transseptal course artery has a downward configuration referred to as the hammock sign. The most common arteries having a transseptal course are the LCA and LAD [6].

D. Retroaortic Course

A coronary artery arising from the RCA or the right coronary cusp and supplying the opposite side of the heart (LCX or LCA) may take a retroaortic course, between the posterior aspect of the aorta and the interatrial septum (Fig. 8 on page 15, Fig. 9 on page 16). It is important to diagnose this anomaly prior to aortic valve replacement surgery due to possible damage to the artery encircling the aortic valve [6].

E. Prepulmonic Course

Coronary arteries found anterior to the pulmonary artery or right ventricular outflow tract are classified as having a prepulmonic course. This course is most commonly seen with LCA, but can also be seen if the RCA has an abnormal origin from the LAD or LCA. An association with prepulmonic course of coronary arteries and Tetralogy of Fallot has been established, which makes important recognizing this coronary artery anomaly prior to corrective surgery via median sternotomy [6].

III. Anomalies of Termination

A. Coronary Artery Fistula

Coronary artery fistula is the abnormal termination of a coronary artery in a cardiac chamber, coronary venous system, pulmonary artery or pulmonary vein (Fig. 9 on page 16, Fig. 10 on page 17). If the coronary artery terminates in a lower pressure chamber it can cause a "steal phenomenon" resembling a left to right shunt [2, 6]. Coronary artery fistulas involve the RCA in 60% of cases and the left coronary system in 40% of cases [2]. On most cases, the fistula connects with the right sided heart chambers inducing the hemodynamics of a left to right shunt [2]. On CT imaging, the coronary artery...
fistula has a dilated and tortuous appearance secondary to the shunt which leads to an increase in blood flow [2, 6].
Images for this section:

**Fig. 1:** Axial view (a) and volume-rendered images (b, c) of a coronary CTA showing high origin of the right coronary artery above the level of the sinotubular junction (yellow and black arrows).

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**Fig. 2:** Coronary CTA axial view (a) and volume-rendered images (b, c) demonstrating left anterior descending coronary artery (red arrow) and left circumflex coronary artery (yellow arrow) both arising independently from the left coronary sinus. There is no left main coronary artery.

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**Fig. 3:** Chest x-ray of a newborn (a) presenting with massive cardiomegaly and cephalization of the pulmonary vasculature. Coronary CTA (b, c) shows anomalous origin of the left coronary artery (red arrow) from the pulmonary artery (ALCAPA). ALCAPA is resulting in myocardial ischemia represented by marked hypoattenuation of the subendocardium (yellow arrows).

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Fig. 4: Maximum intensity projections (a, b) and volume-rendered image (c) of a coronary CTA showing anomalous origin of the left coronary artery (yellow arrows) off the pulmonary trunk 1.6 cm above the pulmonary valve level. Markedly dilated coronary arteries are seen (red arrow) with numerous epicardial intercoronary collaterals (black arrows).

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Fig. 5: Reconstruction of the left anterior descending coronary artery (a, b) and axial view (c) of a coronary CTA showing a myocardial bridge (yellow arrow and circle) between the takeoff of the first and second diagonal branches of the left anterior descending coronary artery.

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Fig. 6: Reconstruction of the left main coronary artery (a), volume-rendered image (b), and axial oblique view (c) of coronary CTA demonstrating anomalous origin of the left main coronary artery above the sinotubular junction (yellow arrows), above the right coronary cusp, showing an interarterial course.

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Fig. 7: Sagittal oblique (a, b) and axial (c) views of a coronary CTA demonstrating the left coronary artery (red arrow) coursing into the interventricular septum. Image (b) is showing the left coronary artery in a transseptal course embedded in the anterior interventricular septum (red arrow). Also note the left anterior descending coronary artery coursing along the epicardial fat (yellow circle).

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**Fig. 8:** Axial view (a) and volume-rendered image (b) of a coronary CTA demonstrating anomalous origin of the left circumflex coronary artery (red arrow) arising from the right coronary cusp, with a retroaortic course.

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**Fig. 9:** Coronary CTA coronal view (a) and sagittal view (b) showing a proximal branch of the left circumflex coronary artery with a retroaortic course (red arrow) and a fistulous connection (yellow arrow) distally with the right lower lobe pulmonary artery about the level of the hilum (more specifically at the level of the distal truncus basalis).
**Fig. 10:** Maximum-intensity projection (a) and volume-rendered image (b) of coronary CTA showing dilated and tortuous left circumflex coronary artery (LCX) draining anomalously (yellow arrow) into the dilated coronary sinus (CS).

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Conclusion

Coronary artery anomalies are a rare condition that can be found as an isolated finding or in conjunction with a congenital heart disease. Many of the coronary artery anomalies have been found to be of little or no clinical significance, yet coronary artery anomalies remain the second cause of sudden cardiac death among young athletes. Coronary Computed Tomography Angiography (CCTA) has become the gold standard for diagnosing and classifying coronary artery anomalies as anomalies of origin, course or termination. Among the hemodynamically significant anomalies are the coronary atresia, origin from a pulmonary artery, interarterial course and coronary artery fistula which may lead to shunting, ischemia or sudden cardiac death. Using CCTA we can adequately diagnose these conditions, resulting in prompt treatment in order to prevent complications.
References


