



Congenital Coronary Artery Anomalies

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DEVELOPMENT AND VARIATION IN CORONARY ARTERIES

The embryonic heart undergoes a remarkable transformation during the first trimester of pregnancy. Initially, the myocardium is thin, and immature vascular plexus develop on its outer surface. This plexus fueled by migrating progenitor cells from both endocardium and sinus venosus, eventually connect with the aorta, receiving blood flow and transforming into primitive coronary arteries. Through a process of regional growth and regression, this vascular plexus is sculpted into the mature coronary artery system we know today. However, any delays in connection to blood flow during this critical development can disrupt downstream vessel formation, leading to anomalous coronary artery anatomy (1).

Within the spectrum of coronary artery development, some clinically insignificant variations are considered normal. These variations, essentially the remnants of the dynamic interplay between blood flow and vessel maturation, can present a wide range of geometries. Understanding these variations, including the compensatory adaptations of neighboring coronary branches, is crucial for interpreting coronary artery images and reasoning myocardial perfusion.

CLASSIFICATION AND CLINICAL SIGNIFICANCE

Congenital coronary artery anomalies can be broadly classified into three categories (2):

1. Anomalies of origin and course: This includes situations where the coronary arteries arise from unexpected locations or follow unusual pathways.
2. Anomalies of intrinsic anatomy: These involve abnormalities within the vessel wall itself, such as myocardial bridging, where a segment of coronary artery dips into the heart muscle.

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3. Anomalies of termination: These occur when a coronary artery ends in an abnormal location, such as a fistula draining blood to an unintended structure.

The prevalence of coronary artery anomalies can vary depending on the imaging technique used. Invasive coronary angiography (ICA), traditionally considered the gold standard, may underestimate their presence. Studies using coronary CT angiography (CCTA) report a prevalence as high as 7.9%, compared to 2.1% with ICA (2, 3).

Understanding the clinical significance of these anomalies is paramount. They are implicated as a leading cause of sudden cardiac death in young athletes (4). Hemodynamically significant anomalies, such as atresia, origin from the pulmonary artery, unusual inter-arterial course, and fistulas, pose a particular threat. Especially the inter-arterial course anomaly is regarded as a malignant anomaly (2, 5). These anomalies can be misinterpreted as coronary artery occlusion during ICA, potentially leading to unnecessary and risky recanalization procedures. Additionally, they can complicate coronary artery bypass surgery and catheter placement during ICA (6).

CCTA: UNVEILING THE HIDDEN

While ICA has historically been the gold standard, advancements in CT technology have positioned CCTA as the preferred method for evaluating coronary artery anomalies. Unlike ICA, which primarily provides a lumenogram, CCTA offers a wealth of information, including three-dimensional relationships within and around the coronary arteries, detailed visualization of the coronary artery lumen, and assessment of arterial wall morphology.

This comprehensive view allows CCTA to readily identify anomalies even in cases that may not cause evident luminal narrowing, such as inter-arterial course and myocardial bridging. Furthermore, recent CCTA models offer a significant advantage in terms of radiation exposure, typically delivering a lower dose than ICA. Additionally, CCTA appears to provide a more accurate picture of the true prevalence of congenital coronary artery anomalies (2, 5).

Given these advantages, CCTA is increasingly recognized as the preferred modality for identifying patients at risk from malignant coronary artery anomalies. It is now recommended as the first-line investigation for patients suspected of having these anomalies (7-9).

A CLOSER LOOK; DUAL LAD ARTERY

One of the most common anomalies in the first category is the presence of dual left anterior descending (LAD) arteries. Studies using CCTA suggest a prevalence of up to 4% (10). Due to its relatively high occurrence, the dual LAD anomaly has been extensively studied, with ten distinct subtypes classified based on the origin, shape, and location of the proximal and distal segments of the LAD arteries.

Among these subtypes, type 4 presents a particularly rare and potentially dangerous scenario. It involves a short proximal LAD and an aberrant distal LAD arising from the right coronary artery (11). This unusual course of the distal LAD increases the risk of damage during cardiac surgery and can also be misdiagnosed as a short-segment LAD occlusion during ICA,

potentially leading to unnecessary recanalization procedures. A recent case report by Jang et al. (12) highlights the potential clinical significance of type 4 dual LAD, presenting a patient with coronary artery disease involving a combined diagonal artery stenosis originating from the proximal LAD segment.

CONCLUSION

Congenital coronary artery anomalies can pose a significant health risk. Advanced imaging techniques, particularly CCTA, are revolutionizing our ability to identify these anomalies and guide appropriate clinical decision-making. As our understanding of these variations continues to evolve, CCTA will undoubtedly play a pivotal role in ensuring optimal patient care.

Conflicts of Interest

The author has no potential conflicts of interest to disclose.

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