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Review

Coronary artery anomalies: clinical overview, diagnostic strategies, and management

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ABSTRACT

Coronary artery anomalies (CAAs) represent a heterogeneous group of congenital abnormalities involving the origin, course, or termination of the coronary arteries. Although most CAAs are asymptomatic and discovered incidentally, certain variants particularly those with an interarterial or intramural course carry a substantial risk of myocardial ischemia, arrhythmia, or sudden cardiac death. With the advent of multidetector computed tomography and cardiac magnetic resonance imaging, recognition of these anomalies has markedly increased. This mini review summarizes the classification, epidemiology, pathophysiological mechanisms, clinical significance, and current diagnostic and management strategies for CAAs, emphasizing their relevance in contemporary cardiology practice.

Keywords: Coronary artery anomalies, coronary angiography, computed tomography angiography, cardiac magnetic resonance, anomalous aortic origin of coronary artery

INTRODUCTION

Coronary artery anomalies (CAAs) are congenital malformations involving deviations in the origin, course, or termination of coronary vessels, occurring in approximately 0.2-1.3% of the general population on invasive angiography and up to 5% on computed tomography coronary angiography (CTCA).[1,2] Most variants are benign; however, certain configurations, such as anomalous aortic origin of the coronary artery (AAOCA) with an interarterial course between the aorta and pulmonary trunk, have been associated with myocardial ischemia and sudden cardiac death, especially in young athletes.[3,4] Advances in cardiac imaging have led to increasing detection, yet optimal management remains controversial. This review provides a concise overview of CAAs with a focus on diagnostic principles and clinical implications.

CLASSIFICATION

CAAs are typically categorized into three broad groups: (1) anomalies of origin and course, (2) anomalies of intrinsic coronary anatomy, and (3) anomalies of termination.[5] Anomalous origin and course include aberrant origins from the opposite sinus (ACAOS), high take-off from the ascending aorta, single coronary artery, or anomalous origin from the pulmonary artery (ALCAPA or ARCAPA). Among these, left coronary artery arising from the pulmonary artery (ALCAPA) represents a malignant variant with early-onset ischemic cardiomyopathy.[6] Anomalous course refers to interarterial, intramural, retroaortic, prepulmonic, or septal trajectories.[7] The interarterial subtype is of greatest clinical concern because of potential vessel compression during exercise.[8] Anomalies of termination include coronary fistulas draining into cardiac chambers or great vessels, which may lead to volume overload or coronary steal phenomena.[9,10]

EPIDEMIOLOGY

The true prevalence of CAAs varies according to imaging modality and population studied. Autopsy and angiographic series estimate an incidence of approximately 1%, whereas CTCA-based studies report higher frequencies due to improved spatial resolution. In a large multicenter CT study, prevalence reached 5.6%, with most anomalies classified as benign variants.[11] Notably, AAOCA has been identified in up to 0.3% of individuals and accounts for 14-19% of sudden cardiac deaths in young athletes.[12] These data highlight the importance of recognizing high-risk morphologies despite overall rarity.

PATHOPHYSIOLOGY AND CLINICAL SIGNIFICANCE

The clinical manifestations of CAAs range from asymptomatic incidental findings to exertional angina, syncope, arrhythmia, myocardial infarction, or sudden cardiac death.[12,13] Pathophysiological mechanisms include slit-like ostium and acute take-off angle, causing dynamic obstruction during systole; intramural course within the aortic wall, leading to luminal compression; interarterial course, predisposing to external compression between the great vessels during exertion; and coronary steal in fistulous communications.[14-16] In ALCAPA, perfusion of the left ventricle from the low-pressure pulmonary system results in chronic ischemia and

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progressive ventricular dysfunction.[6] Conversely, benign variants, such as retroaortic or prepulmonic courses, typically lack hemodynamic consequences.[17]

DIAGNOSTIC EVALUATION

Echocardiography

Transthoracic and transesophageal echocardiography serve as first-line tools, especially in pediatric patients, offering real-time visualization of coronary origins and proximal courses. However, spatial resolution is limited in adults or complex anatomies.[18]

Computed Tomography Coronary Angiography (CTCA)

CTCA has emerged as the gold standard for anatomical delineation due to its superior three-dimensional resolution, enabling precise mapping of coronary origins, trajectories, and relationships with adjacent structure. The modality is particularly useful in differentiating interarterial and intramural components, which are difficult to appreciate on conventional angiography.[19,20]

Cardiac Magnetic Resonance Imaging (CMR)

CMR complements CT by providing both anatomical and functional assessment, allowing evaluation of ischemia and myocardial viability without radiation exposure[16].

INVASIVE CORONARY ANGIOGRAPHY AND FUNCTIONAL TESTING

Conventional angiography remains essential when physiological significance must be assessed. Techniques such as fractional flow reserve (FFR) or instantaneous wave-free ratio (iFR) measurements can quantify ischemia in anomalous vessels.[21]

MANAGEMENT STRATEGIES

Therapeutic decision-making depends on anatomical configuration, symptom status, and evidence of ischemia. Low-risk or incidental variants are usually managed conservatively with regular follow-up and activity modification.[22] High-risk configurations particularly AAOCA with an interarterial or intramural course and ALCAPA warrant surgical correction. Surgical options include unroofing of the intramural segment, reimplantation into the appropriate sinus, coronary artery bypass grafting, or ostioplasty.[23,24] Percutaneous closure is reserved for selected coronary fistulas.[25] Recent consensus statements recommend individualized management guided by multimodality imaging, physiological assessment, and patient-specific risk evaluation.[26] Despite increasing recognition, randomized trials are lacking, and most recommendations rely on expert consensus.[27]

CONCLUSION

Coronary artery anomalies encompass a diverse spectrum of congenital abnormalities with variable clinical significance. While most are benign, certain high-risk variants pose substantial risk for ischemic events and sudden cardiac death. Advances in CT and MR imaging have revolutionized detection, yet management continues to rely heavily on anatomical risk stratification and multidisciplinary judgment. Early recognition, appropriate imaging, and tailored treatment remain essential to improving patient outcomes. Future research should focus on prospective multicenter registries and long-term outcome data to better define optimal management pathways.

ETHICAL DECLARATIONS

Referee Evaluation Process

Externally peer-reviewed.

Conflict of Interest Statement

The authors have no conflicts of interest to declare.

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Author Contributions

All of the authors declare that they have all participated in the design, execution, and analysis of the paper, and that they have approved the final version.

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