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CASE REPORT

# A clinical report on the anomalous aortic origin of the right coronary artery

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## Abstract

Anomalous aortic origin of the right coronary artery (AAORCA) is a rare congenital anomaly where the right coronary artery originates abnormally from the aorta, posing risks such as myocardial ischemia and sudden cardiac death, particularly in young athletes. Symptoms range from being asymptomatic to exertional signs such as dizziness or chest pain. Diagnosis is typically made using coronary computed tomography angiography, the gold standard for detailed assessment. Treatment depends on symptom severity, ranging from medical management to surgical options such as unroofing, reimplantation, or bypass grafting. This report presents a case of a 9-year-old patient diagnosed with AAORCA, exhibiting mild symptoms in the absence of exertional intolerance. This case emphasizes the importance of imaging, regular follow-ups, and early diagnosis. Personalized treatment and further research are crucial for improving outcomes and reducing the risk of sudden cardiac death in AAORCA patients.

***Keywords:*** Anomalous aortic origin of the right coronary artery; Congenital abnormality; Sudden cardiac death

## 1. Introduction

Anomalous aortic origin of the right coronary artery (AAORCA) is a rare congenital anomaly characterized by the abnormal origin of the right coronary artery (RCA) from an inappropriate aortic sinus, typically the left sinus of Valsalva.[1] This condition, affecting approximately 0.008 – 0.32% of the population, poses significant clinical risks, including myocardial ischemia, sudden cardiac death, and other complications, particularly in young individuals and athletes.[2,3] AAORCA can present with two primary courses:

An interarterial course, where the RCA passes between the aorta and pulmonary artery, and an intramural course, where the artery runs within the aortic wall. Both variations are associated with increased risk for ischemia or infarction, especially during exertion, due to external compression or luminal narrowing (Figure 1).[3]

Clinical manifestations of AAORCA can range from asymptomatic to more severe symptoms such as chest pain, syncope, palpitations, or exertional dyspnea. Sudden cardiac death is a catastrophic presentation, often occurring during or after exercise. Diagnosis can be challenging as symptoms may resemble those of other coronary artery diseases.[4] Initial non-invasive screening, such as echocardiography, is commonly used in pediatric patients, with transesophageal echocardiography offering better visualization of coronary origins. However, coronary computed tomography angiography (CCTA) is considered the gold standard for diagnosing AAORCA, providing detailed anatomical insights into coronary artery origins and their relation to surrounding structures. Cardiac magnetic resonance (CMR) imaging is useful for assessing myocardial perfusion and viability, particularly in ischemic cases. Invasive coronary angiography is employed when non-invasive methods fail to provide sufficient information.[5]

The management of AAORCA is tailored based on the severity of symptoms, presence of ischemia, or high-risk anatomical features, such as an interarterial or intramural course. A shared decision-making tool is crucial for managing AAORCA, guiding the physician and patient/family in selecting the most appropriate treatment based on the individual's specific anatomical features, symptoms, and risk factors. Medical management includes the use of β-blockers to reduce myocardial oxygen demand and avoiding strenuous physical activity in high-risk patients. Surgical options include the unroofing procedure, which corrects the intramural course to ensure proper luminal flow, reimplantation to reposition the RCA to the correct sinus of Valsalva, and coronary bypass grafting, particularly for cases unsuitable for unroofing or reimplantation.[6] Percutaneous coronary interventions are rarely employed but may be considered for select cases of stenosis relief. For asymptomatic or low-risk individuals, regular follow-up with imaging is recommended for ongoing surveillance.[7]

The prognosis of AAORCA varies widely, but early diagnosis and effective, personalized management can substantially reduce the risk of sudden cardiac death and improve quality of life. Surgical interventions typically yield favorable outcomes, alleviating ischemic symptoms and restoring coronary flow. Recent advancements in imaging and surgical techniques have enhanced the understanding and management of AAORCA. However, challenges persist in identifying optimal strategies for asymptomatic

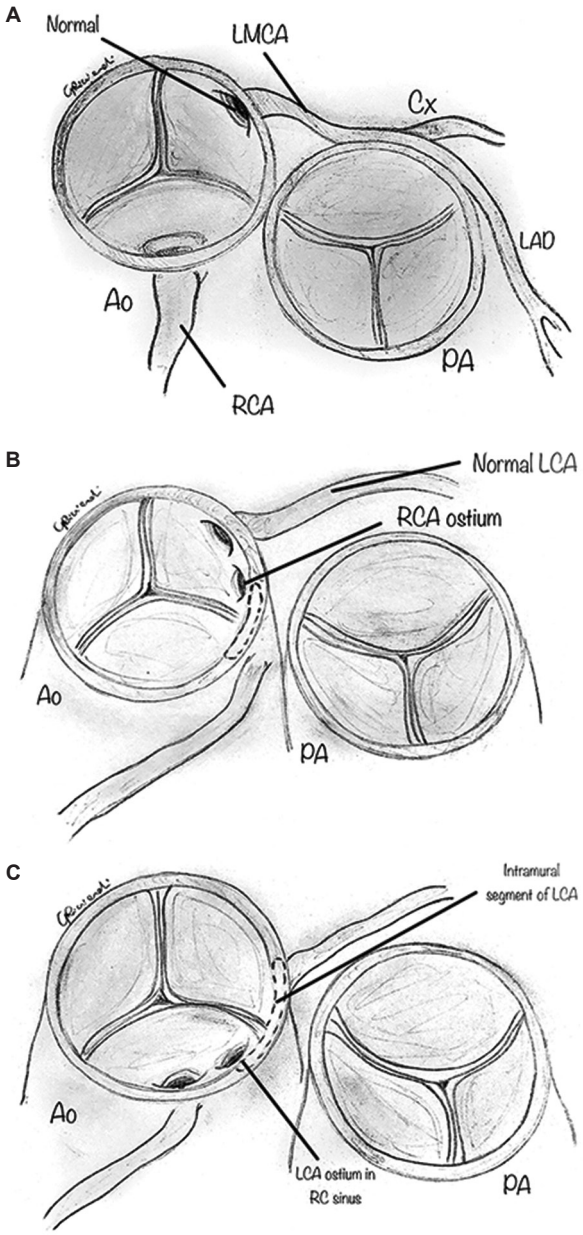


**Figure 1.** Diagram showing cross section aortic view: (A) Normal coronary artery anatomy: the left main coronary artery gives origin to the left anterior descending and circumflex coronary artery, which arise from a common stem from the left aortic sinus; the RCA arises from the right aortic sinus; (B) the most common type, that is, anomalous aortic origin of the RCA from the left sinus (AAORCA), with an intramural segment; (C) the most lethal type, that is, anomalous aortic origin of the left coronary artery from the left sinus (AAOLCA), with an intramural segment. Reproduced from Padalino *et al.*[4]; under the terms of the creative commons attribution license (CC BY).
Abbreviations: RCA: Right coronary artery; AAORCA: Anomalous aortic origin of the right coronary artery; AAOLCA: Anomalous aortic origin of the left coronary artery

patients and determining the long-term outcomes of both surgical and non-surgical approaches. AAORCA, while rare, is a potentially life-threatening coronary anomaly that demands a high degree of clinical suspicion, particularly in young individuals presenting with exertional symptoms. Advanced imaging techniques and tailored management strategies are vital in preventing adverse events. Further research is needed to refine risk stratification methods and develop effective long-term management plans.[8]

## 2. Case presentation

A 9-year-old African American female presented to the clinic with primary concerns of fatigue and occasional dizziness. An echocardiogram performed during the initial evaluation revealed AAORCA, which was subsequently confirmed on a computed tomography (CT) scan. The patient was physically active and had participated in school sports without experiencing chest pain, shortness of breath, or syncope. She reported no difficulty keeping up with her peers during physical activity. She occasionally experienced dizziness and felt fatigued after school, often taking multiple naps. Her nutritional intake was appropriate, though her fluid consumption was suboptimal. Her medical history was significant only for asthma. She denied any aggravating factors for her symptoms. Her family history is negative for congenital heart disease, cardiomyopathy, or sudden cardiac death before the age of 50.

On presentation, the patient appeared alert and in no acute distress. Her vital signs included a blood pressure of 108/65 mmHg, pulse of 111 beats/min, and a temperature of 36.6°C (97.8°F). She measured 151.5 cm in height and weighed 37.9 kg, resulting in a body mass index of 16.5 kg/m$^2$. The physical examination findings were unremarkable. Bowel sounds were normoactive, and the abdomen was soft, non-tender, and non-distended, with no evidence of hepatosplenomegaly. The lungs were clear to auscultation, and the heart rate and rhythm were regular with a normal S1 and S2, without murmurs, friction rubs, or gallops. Her skin was warm, with normal capillary refill, and there were no signs of cyanosis. While she reported occasional dizziness, she had no other complaints and appeared otherwise well. An echocardiogram revealed an AAORCA, which was subsequently confirmed by a CT scan (Figure 2). An electrocardiogram demonstrated a left atrial rhythm but was otherwise normal in rate and rhythm. An exercise stress test showed no abnormal findings.

The CT angiography (Figure 3) identified a right-dominant coronary circulation. The RCA arises from a separate origin obliquely and eccentrically to the right of the left coronary cusp. It courses approximately 1.7 cm between the aorta, main pulmonary artery, and right ventricular outflow tract, with a suspected intramural course. The RCA is ovoid at its origin, measuring 1.9 × 1 mm (Z-score = −2.7). Approximately 2 cm distal to its origin, the RCA measures 1.9 × 1.6 mm (Z-score = −2). The left coronary artery left anterior descending artery and left circumflex artery are normal in both course and caliber.

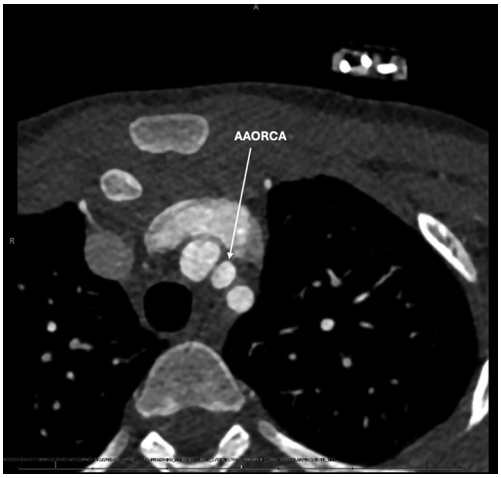


**Figure 2.** Axial computed tomography image demonstrating anomalous aortic origin of the right coronary artery. The right coronary artery arises anomalously from the left coronary sinus of Valsalva and follows an interarterial trajectory, coursing between the aortic root and pulmonary trunk. This high-risk anatomical variant is associated with dynamic compression during periods of increased cardiac output, predisposing to myocardial ischemia and sudden cardiac death, particularly in young athletes or during strenuous exertion.

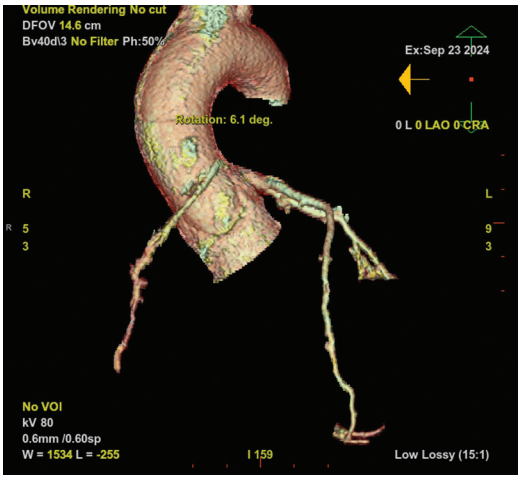


**Figure 3.** Volume-rendered three-dimensional computed tomography angiography reveals an anomalous aortic origin of the right coronary artery from the left sinus of Valsalva. The right coronary artery follows an interarterial course between the aortic root and pulmonary trunk, a high-risk anatomical variant predisposing to dynamic compression and myocardial ischemia during exertion. The reconstruction further demonstrates a right-dominant coronary circulation, evidenced by the right coronary artery's posterior extension and its contribution to the posterior descending artery and posterolateral ventricular branches.

The differential diagnosis for AAORCA includes conditions with overlapping symptoms, such as chest pain, syncope, or exertional dyspnea. These include hypertrophic cardiomyopathy, coronary artery disease, other coronary anomalies such as anomalous aortic origin of the left coronary artery (AAOLCA), aortic valve disorders, myocarditis, and arrhythmogenic right ventricular cardiomyopathy. Pulmonary embolism and pericarditis can also present similarly, along with non-cardiac causes such as asthma, mitral valve prolapse, and psychogenic factors such as anxiety or panic attacks. Accurate diagnosis requires thorough clinical evaluation and advanced imaging modalities, including echocardiography, CCTA, or CMR to distinguish AAORCA from these conditions.

Based on the imaging findings and clinical symptoms, a diagnosis of AAORCA was made.

## 3. Discussion

Anomalous aortic origin of a coronary artery is a congenital condition in the pediatric population and is considered the second leading cause of sudden cardiac death in young athletes.[9] Although the embryological development of coronary arteries is not fully understood, developmental disruptions may lead to anomalies in the origin of coronary arteries (arising from the aorta or pulmonary artery) or incomplete vessel formation. These anomalies can manifest as coronary artery fistulae or coronary sinusoids. Among pediatric patients, anomalous origins of the RCA are observed in 50% of cases.[10]

The intramural course and acute angle of takeoff associated with anomalous aortic origin of the coronary artery (AAOCA) can predispose the affected vessel to blood flow obstruction and resulting myocardial ischemia.[11] The most commonly reported clinical presentations of AAOCA include anginal chest pain, exercise-induced syncope, or non-vagally mediated arrhythmias. However, some individuals remain asymptomatic, with sudden cardiac death as their first clinical manifestation, a scenario more frequently observed in young athletes.[12] While the overall incidence of sudden cardiac death in patients with AAOCA is low, it is higher in those with AAOLCA compared to the AAORCA, particularly in asymptomatic individuals.[13] Surgical interventions, such as coronary artery bypass grafting or intracoronary stenting, are more strongly recommended for AAOLCA to mitigate the risk of myocardial ischemia and sudden cardiac death.[14,15]

Our patient presents with an AAORCA, where the RCA arises obliquely and eccentrically from a separate origin on the left coronary cusp. Imaging reveals an approximately 1.7 cm interarterial course between the aorta, main pulmonary artery, and right ventricular outflow tract, with a suspected intramural course. Her primary symptom is occasional dizziness, without any reports of chest pain or syncope during exertion.

Given her diagnosis of AAORCA and the absence of exertional chest pain or syncope, as of the time of writing this paper, our current management plan involves regular monitoring for symptom progression. We will repeat an exercise stress test and maintain routine follow-ups with her cardiologist. Once the patient reaches a weight of 40 kg (anticipated next year), a stress perfusion CMR imaging will be performed to evaluate for any signs of myocardial ischemia. Participation in sports and physical activities is permitted, contingent on the availability of an automated external defibrillator on site. If she develops symptoms such as chest pain, she should immediately discontinue all sports activities and seek urgent medical attention. Overall, the patient showed signs of recovery and will continue to be closely monitored to ensure timely intervention if her condition changes.

## 4. Conclusion

The findings and discussion in this report highlight the complexity and critical nature of diagnosing and managing AAORCA. This rare coronary anomaly, associated with significant clinical risks, demands a high degree of clinical vigilance, particularly in younger, active individuals. Early diagnosis is pivotal for identifying high-risk anatomical features and determining appropriate management strategies. Ongoing advancements in imaging and surgical techniques underscore the need for further research to refine long-term management and risk stratification for AAORCA. With a focus on individualized care and regular monitoring, outcomes can be optimized, significantly reducing the potential for adverse events. The patient is currently stable and will undergo regular monitoring to allow prompt intervention in the event of any clinical deterioration.

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## Conflict of interest

The authors declare no conflicts of interest.

## Author contributions

*Conceptualization:* Zafar Qureshi
*Investigation:* All authors

*Methodology:* All authors
*Writing – original draft:* All authors
*Writing – review & editing:* All authors

## Ethics approval and consent to participate

The patient has given verbal consent to participate in this study.

## Consent for publication

Verbal consent has been obtained from the patient to publish his data.

## Availability of data

Not applicable.

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