A High Take-Off Right Coronary Artery with Intramural Course: A Rare Case Review

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AUTHOR CONTRIBUTIONS

VNN- contributed to writing and editing the manuscript prior to submission and was involved in patient care.
TTH- contributed to writing the manuscript prior to submission.
TTN- contributed to writing and editing the manuscript prior to submission.
ANN- contributed to editing the manuscript prior to submission and was involved in patient care.
VTN- contributed to writing and editing the manuscript prior to submission and was involved in patient care.
All authors read and approved the final manuscript.

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DISCLOSURES

The authors declare that they have no financial or non-financial competing interests to disclose.

CONSENT

Yes, Informed consent was obtained from the patient for the publication of data from her clinical history and the necessary images.

HUMAN AND ANIMAL RIGHTS

The procedures followed were in accordance with the ethical standards of the responsible committee on human experimentation and with the Helsinki Declaration of 1975, as revised in 2000.

ABSTRACT

Background: Anomalous aortic origin of a coronary artery (AAOCA) is a rare congenital condition (prevalence 0.1%–0.33%) frequently associated with sudden cardiac death in young populations. While a "high take-off" (origin >1 cm above the sinotubular junction) is often incidentally found and considered benign, its clinical significance increases when paired with high-risk features like an intramural course or slit-like ostium.

Case Presentation: We present the case of an asymptomatic 60-year-old male whose routine evaluation via Coronary Computed Tomography Angiography (CCTA) revealed a high take-off of the right coronary artery (RCA). The ostium was located 11 mm above the right sino-tubular junction, exhibiting an elliptical, slit-like morphology and an acute take-off angle. Uniquely, the RCA followed a prolonged 14-mm intramural course within the aortic wall, though notably absent of a "malignant" interarterial path between the aorta and pulmonary artery.

Conclusion: The presence of a significant intramural course does not necessitates surgery in all patients. In the absence of an interarterial course and clinical symptoms, particularly in patients over age 35, conservative medical management and rigorous surveillance provide a safe alternative to high-risk surgical correction.

Keywords: AAORCA, High take-off, Intramural course, CCTA, Congenital coronary anomaly.

CASE REPORT

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BACKGROUND

Coronary artery anomalies (CAAs) affect 0.6–1.3% of the population. While most are benign and discovered incidentally, variants involving an intramural course are clinically significant due to their association with myocardial ischemia and sudden cardiac death. High take-off of the right coronary artery (RCA) with an intramural segment is an rare variant that presents unique diagnostic and procedural challenges. This anatomical configuration results in dynamic systolic compression of the vessel within the aortic wall, necessitating multimodality imaging for accurate risk stratification. We report a case of high take-off RCA with a significant intramural course, highlighting the clinical presentation, imaging characterization, and management considerations for this uncommon anomaly.

CASE REPORT

A 60-year-old male presented for routine cardiovascular follow-up. He remained asymptomatic, with no reports of chest pain or dyspnea. Physical examination was unremarkable, with stable vital signs and no significant findings in the cardiac, pulmonary, or neurological systems.

Coronary Computed Tomography Angiography (CCTA) revealed a congenital coronary anomaly: the right coronary artery (RCA) originated abnormally from a site 14.4 mm above the right sino-tubular junction at the wall of the ascending aorta (Figure 1a,1b). The proximal RCA exhibited an elliptical, slit-like ostium (Figure 1c) with an acute take-off angle of approximately 10.1 degrees (Figure 3a). Interluminal space (ILS) measured 0.8 mm at 2 mm from the ostium (Figure 3b). The RCA followed an intramural course of 14.1 mm (Figure 2a,2b), but no interarterial course was observed between the ascending aorta and the pulmonary trunk.

The patient is currently managed conservatively with medical therapy and advised to undergo regular monitoring, with instructions to seek immediate medical attention should any cardiac symptoms arise.

DISCUSSION

Anomalous aortic origin of the right coronary artery (AAORCA) is an uncommon congenital coronary anomaly in which the right coronary artery (RCA) arises from an abnormal site—most often the left sinus of Valsalva rather than the right [1-3]. Prevalence in the general population is roughly 0.1–0.33%, and AAORCA is more common than anomalous aortic origin of the left coronary artery (AAOLCA) [4-6].

AAORCA arises from abnormal coronary artery formation during fetal development [1,2]. Two major contributors are commonly discussed: Complex embryologic development: Coronary vessel formation depends on multiple embryologic cell sources and tightly regulated molecular signals and hemodynamic cues [1,3-5]. Disruptions in these coordinated processes can prevent normal formation and connection of the coronary "stems" to the aorta, resulting in an abnormal ostial location or connection pattern [1,3,4]. Genetic and familial factors: Although the precise inheritance pattern in humans is not fully defined, familial clustering of anomalous aortic origin of a coronary artery has been reported, including among first-degree relatives [6,7]. This supports a role for inherited factors, and genetic evaluation may be considered in some families [6].

AAORCA is commonly categorized by the *course* the anomalous RCA takes from its ectopic origin (often the left sinus) to its usual myocardial territory. The most clinically significant is the interarterial course, in which the RCA travels between the ascending aorta and the pulmonary artery and is often considered a potentially "malignant" variant due to increased risk of ischemia and sudden cardiac death, frequently including an intramural segment [1-4]. Other anatomic variants are generally benign: the retroaortic course passes posterior to the aortic root and is typically not ischemia-producing; the prepulmonic (anterior) course runs anterior to the pulmonary artery/pulmonary outflow tract and is also usually considered benign [1,2]; the intraseptal (subpulmonic/intraconal) course traverses the interventricular septum or ventricular myocardium [1]; and the rare posterior atrioventricular groove (retrocardiac) course passes behind the heart [1,4]. Beyond an origin in the left coronary sinus, the RCA may also originate anomalously from other aortic sites, including the posterior (non-coronary) sinus [1,6] or the ascending aorta ("high take-off") [1,7].

A high take-off coronary anomaly refers to a coronary ostium arising from the ascending aorta noticeably above its expected position within the sinus of Valsalva. Normally, coronary ostia are located at, just below, or slightly above the sinotubular junction (STJ) within the aortic root [1], but high take-off is commonly defined as an origin more than 1 cm (>1.0 cm) above the STJ [2]. During angiography, if the ostium is not found within the sinuses, examining the proximal ~2 cm of the ascending aorta often identifies the ectopic opening [5]. High take-off is typically grouped among “anomalies of coronary artery connection to the aorta/systemic circulation” [6-9]. It can involve either the RCA or the left main trunk (LCA/LMT), but it is substantially more common in the RCA [10,11]. Aside from the elevated ostial location, the downstream coronary course and myocardial distribution are often otherwise normal [5].

In a large angiographic series of 126,595 patients, high take-off of the RCA occurred in approximately 0.15% of the overall cohort (11.2% of isolated coronary anomalies), while high take-off of the LMT was much rarer at ~0.013% (0.95% of isolated anomalies) [11]. In the EURO-AAOCA registry, high origin accounted for 6.1% of RCA cases and 1.9% of LCA cases [12].

As with other coronary connection anomalies, high take-off is congenital and reflects abnormal embryologic development [13,14]. The proximal aorta and the distal coronary vascular plexus develop separately; disturbances that impair their precise alignment and fusion can lead to a coronary stem connecting higher on the ascending aorta than intended [15,16].

High take-off coronary arteries, when occurring in isolation as an ectopic origin from the ascending aorta, are generally considered benign and not typically associated with ischemia or hemodynamic compromise [5,17]. However, when high take-off is present alongside other high-risk anatomic features—such as an Intramural course—it may be considered a high-risk feature because it can influence ostial geometry and the take-off angle, potentially increasing the risk of exertional ischemia [2,18].

In AAORCA, an intramural course describes a proximal segment of the anomalous coronary artery that runs within the wall (tunica media) of the ascending aorta before exiting to the epicardial surface [1,2].

When the RCA arises from the left sinus and follows an interarterial course, an intramural segment is common [3,4]. Imaging suspicion may be raised when the interluminal space is <0.95 mm at 2 mm from the ostium [5,6]. Compared with left-sided anomalies, the intramural length in AAORCA is often shorter (around 2–3 mm), and the ostium may lie near the commissure between the right and left coronary cusps. Intramural anatomy is one of the principal "high-risk" contributors to ischemia and sudden cardiac death in AAORCA [7]. Proposed mechanisms include lateral compression, in which the intramural lumen often becomes ovoid rather than circular [10]; proximal hypoplasia, where growth constraints within the aortic wall can lead to a narrowed proximal segment [11]; and dynamic worsening with exercise, as increased cardiac output and aortic expansion during exertion can further compress the intramural segment, and in combination with a slit-like orifice may function like a dynamic "valve," reducing perfusion and provoking ischemia [7,10,12]. IVUS can be especially useful because it can directly demonstrate lateral compression and pulsatile narrowing throughout the cardiac cycle [10,13].

Management for asymptomatic AAOCA is individualized and depends on the involved artery (left vs right), patient age, anatomy, and functional ischemia testing. For asymptomatic AAOLCA, surgery is generally favored because interarterial AAOLCA carries higher sudden cardiac death risk [1,2]; the 2017 AATS guidelines provide a Class I recommendation to offer surgery for asymptomatic AAOLCA with an interarterial course [3,4], and AHA/ACC guidance similarly considers surgery "reasonable" (Class IIa) even without symptoms or inducible ischemia [5,6]. In contrast, for asymptomatic AAORCA, observation is often acceptable because AAORCA is generally lower-risk than left-sided disease [7,8]; both surgery and conservative management may be considered (AHA/ACC Class IIb) [9,10], and AATS guidance recommends stress testing for inducible ischemia—if negative, conservative management and (after counseling) sports participation may be allowed [3,4,8].

Age is an important factor in management decisions for AAORCA. In younger patients (<35 years), AAOCA-related sudden cardiac death occurs predominantly in children, young adults, and athletes during exertion, which may lower thresholds for intervention in some cases [1,11,12]. In contrast, older adults (>35 years) have a declining incidence of sudden cardiac death after age ~35, and the survival benefit from surgery for older adults with AAORCA is less certain, supporting conservative management unless ischemia is clearly demonstrated [5,11,12,15].

TEACHING POINTS

Anomalous aortic origin of the right coronary artery (AAORCA) with "high take-off" subtype is characterized by an aberrant origin more than 1 cm above the sinotubular junction in the ascending aorta.

Cardiac computed tomography angiography (CCTA) is essential for identifying an intramural course, which carries the highest risk for ischemia and sudden cardiac events. High-risk markers include: interluminal space (ILS) <0.95 mm measured 2 mm from the ostium (100% sensitivity for intramural course), intramural length ≥14 mm (the portion of artery embedded within the aortic wall without intervening fat), asymmetry index (ellipticity)—the ratio of long axis to short axis indicating ovoid or slit-like luminal compression, absence of peri-coronary fat, slit-like orifice appearing narrow and elongated rather than circular, and acute take-off angle <30° (or <45°) from the aortic wall,

Conservative management may be appropriate for asymptomatic AAORCA patients when: right coronary involvement (lower risk than AAOLCA), negative stress testing (AATS permits sports participation after counseling), age >35 years (declining sudden cardiac death risk, uncertain surgical benefit), normal FFR (favors conservative management despite high-risk anatomy), and no severe dynamic compression on stress IVUS (surgery reserved for significant pressure gradients or severe dynamic compression during stress).s).

QUESTIONS

Question 1: How many types of anomalous origin sites are described for the right coronary artery in AAORCA?

A) 2 sites
B) 3 sites
C) 4 sites
D) 5 sites

Correct Answer: B) 3 sites

Explanation: The discussion describes three main anomalous origin sites for the RCA in AAORCA: the **left sinus of Valsalva**, which is the most common ectopic origin site (mentioned as "most often the left sinus of Valsalva rather than the right"); the **posterior (non-coronary) sinus**, an alternative aortic sinus origin; and the **ascending aorta ("high take-off")**, where the origin is from the ascending aorta above the sinotubular junction.

Question 2: Which anatomic course of the anomalous right coronary artery (RCA) in AAORCA is most risky for sudden cardiac death?

A) Retroaortic course

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B) Prepulmonic (anterior) course
C) Interarterial course
D) Intramural course

Correct Answer: C) Interarterial course

Explanation: The discussion explicitly states that the **interarterial course** is "the most clinically significant" and is "often considered a potentially 'malignant' variant due to increased risk of ischemia and sudden cardiac death."

In this variant, the RCA travels between the ascending aorta and the pulmonary artery, frequently including an intramural segment. The discussion notes that this intramural anatomy is "one of the principal 'high-risk' contributors to ischemia and sudden cardiac death in AAORCA."

The other course variants mentioned (retroaortic, prepulmonic, intraseptal, and posterior atrioventricular groove) are described as "generally benign" or "typically not ischemia-producing."

Question 3: How many diagnostic criteria or features are described for identifying an intramural course of the RCA in AAORCA using CCTA?

A) 3 criteria
B) 5 criteria
C) 6 criteria
D) 7 criteria

Correct Answer: D) 7 criteria

Explaination: **Seven diagnostic criteria help identify an intramural course of the RCA in AAORCA using CCTA.** Quantitative CCTA criteria include: (1) interluminal space (ILS) <0.95 mm measured at 2 mm from the ostium (100% sensitivity), (2) intramural length measuring the portion of artery embedded in the aortic wall without intervening fat (e.g., 14 mm is significant), and (3) asymmetry index/ellipticity representing the ratio of long axis to short axis, indicating an ovoid or slit-like cross-section. Qualitative anatomical markers include: (4) absence of peri-coronary fat—no visible fat or adventitia between the coronary artery and aortic media (the most reliable visual sign on CCTA), (5) slit-like orifice where the ostium appears as a narrow, elongated slit rather than circular, (6) acute take-off angle with entry angle <30° (or <45° in some sources) from the aortic wall, and (7) high take-off with origin >1 cm above the sinotubular junction (STJ).

Question 4: How many criteria support conservative management for asymptomatic AAORCA?

A) 2 criteria
B) 3 criteria
C) 4 criteria
D) 5 criteria

Correct Answer: D) 5 criteria

Explaination: Five criteria support conservative management for asymptomatic AAORCA: (1) the anomaly involves the right coronary artery rather than the left, as AAORCA is generally lower-risk than AAOLCA; (2) stress testing is negative for inducible ischemia, in which case AATS guidance permits conservative management and even sports participation after counseling; (3) patient age is greater than 35 years, since sudden cardiac death incidence declines after this age and the survival benefit from surgery in older adults is uncertain; (4) invasive functional testing (FFR) is normal, which favors conservative management even when high-risk anatomic features are present; and (5) stress IVUS shows no severe dynamic compression, as surgery is typically reserved for cases with functionally significant pressure drops or severe dynamic compression during stress.tress"

Question 5: How many criteria support surgical management for asymptomatic AAORCA?

A) 2 criteria
B) 3 criteria
C) 4 criteria
D) 5 criteria

Correct Answer: C) 4 criteria

Explaination: Four criteria support surgical management for asymptomatic AAORCA: (1) the anomaly involves the left coronary artery (AAOLCA) rather than the right, as AAOLCA carries higher sudden cardiac death risk and receives Class I (AATS) and Class IIa (AHA/ACC) recommendations for surgery even without symptoms; (2) stress testing demonstrates inducible ischemia, which lowers the threshold for intervention; (3) patient age is less than 35 years, particularly in children, young adults, and athletes, as AAOCA-related sudden cardiac death occurs predominantly during exertion in this age group; and (4) invasive assessment shows functionally significant abnormalities, specifically abnormal FFR (indicating hemodynamically significant pressure drops) or severe dynamic compression on IVUS during pharmacologic stress, as surgery is reserved for these cases by many multidisciplinary teams.

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FIGURES

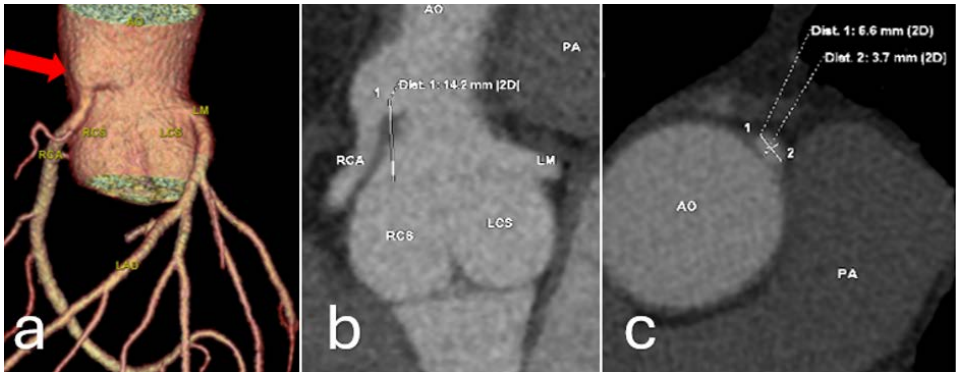


Figure 1: Dual-energy CT (1975 slices, GE Apex Resolution) coronary angiography shows the right coronary artery origin (red arrow) above the sinotubular junction (STJ) (a), measuring approximately 11 mm (b). The asymmetry index/ellipticity—the ratio of long axis (6.6 mm) to short axis (3.7 mm)—indicates an ovoid or slit-like cross-section (c).

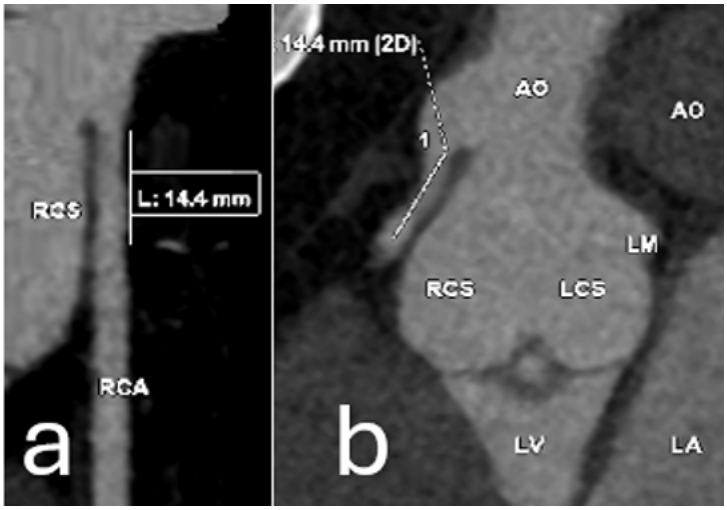


Figure 2: Dual-energy CT (1975 slices, GE Apex Resolution) coronary angiography shows a 14.4 mm portion of the right coronary artery embedded in the aortic wall without intervening fat (intramural course) (a,b).

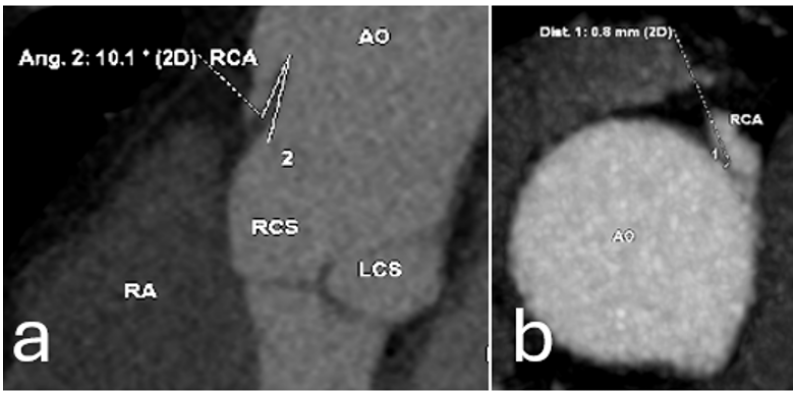


Figure 3: Dual-energy CT (1975 slices, GE Apex Resolution) coronary angiography shows the RCA intramural course has an acute take-off angle with an entry angle of approximately 10.1 degrees from the aortic wall (a). Interluminal space (ILS) measures 0.8 mm at 2 mm from the ostium (b).

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KEYWORDS

AAORCA (Anomalous Aortic Origin of the Right Coronary Artery), AAOLCA (Anomalous Aortic Origin of the Left Coronary Artery), congenital coronary anomaly, interarterial course, intramural segment, high take-off coronary artery, retroaortic course, prepulmonic course, intraseptal course, right coronary artery (RCA), sudden cardiac death, myocardial ischemia, lateral compression, proximal hypoplasia, exercise-induced ischemia, embryologic development, genetic and familial factors, sinotubular junction (STJ), IVUS (Intravascular Ultrasound), surgical management, conservative management, stress testing, AATS guidelines, AHA/ACC guidance

ABBREVIATIONS

AAOCA = Anomalous Aortic Origin Of Coronary Artery
AAOLCA = Anomalous Aortic Origin Of The Left Coronary Artery
AAORCA = Anomalous Aortic Origin Of The Right Coronary Artery
AATS = American Association For Thoracic Surgery
AHA/ACC = American Heart Association/American College OF Cardiology
FFR = Fractional Flow Reserve
IVUS = Intra Vascular Ultra Sound
LCA/LMT = Left Coronary Artery/Left Main Trunk
RCA = Right Coronary Artery
STJ = Sino Tubular Junction

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