Anomalous Origin of Coronary Arteries: A Diagnostic Dilemma

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Abstract

With the increasing use of imaging for screening and diagnostic purposes, particularly coronary CT angiography, the number of adult patients diagnosed with anomalous origin of the coronary arteries (AOCA) has risen significantly. While current guidelines offer a general framework for managing and treating AOCA, patients present with diverse anomalies, symptoms, and clinical presentations, making broad recommendations less universally applicable. Notably, a wide range of treatment options exists, but there is no clear consensus on the best intervention strategy. Presented here are three cases of AOCA emphasizing the anatomical variations, clinical presentations, and the utility of coronary CT angiography in delineating anatomy and identifying high-risk features.

Keywords

Anomalous origin coronary artery, cardiac CT angiography, fractional flow reserve, high-risk features

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Given the significant advances in imaging techniques, particularly coronary CT angiography (CTA), the number of patients diagnosed with anomalous aortic origin of coronary arteries (AAOCA) has increased in recent years. AAOCA encompasses a wide range of anatomical variations and clinical presentations, and current guidelines offer broad diagnostic and management strategies. However, optimal treatment often needs to be individualized based on the patient's clinical status and imaging findings, as the applicability of these recommendations in clinical practice is limited.

We present three cases of AAOCA, emphasizing the anatomical variations, clinical presentations, and utility of CTA in clearly delineating anatomy and identifying high-risk features.

Case 1

A 68-year-old man with a history of tobacco use disorder and a recent diagnosis of lung cancer underwent outpatient coronary CTA due to incidental coronary calcifications noted on chest CT. Coronary CTA revealed an anomalous origin of the left main coronary artery (L-AAOCA) arising from the right coronary sinus, following a pre-pulmonic course (*Figure 1A*). The left main artery bifurcated into branches that supplied the left anterior descending (LAD), ramus, and circumflex (LCX) arteries (*Figure 1B*). Moderate non-obstructive atherosclerotic disease (<50% stenosis) was noted in the left coronary system. The right coronary artery (RCA) originated normally from the right coronary sinus and was hyperdominant (*Figure 1C*), giving rise to a large caliber posterior descending and posterior lateral branches. Additionally, an obtuse marginal (OM) artery arose from the distal RCA supplying the lateral wall of the left ventricle. The RCA had only mild atherosclerotic disease causing <25% stenosis.

Given the benign course of this anomalous coronary and lack of chest pain, the decision was made to monitor the patient.

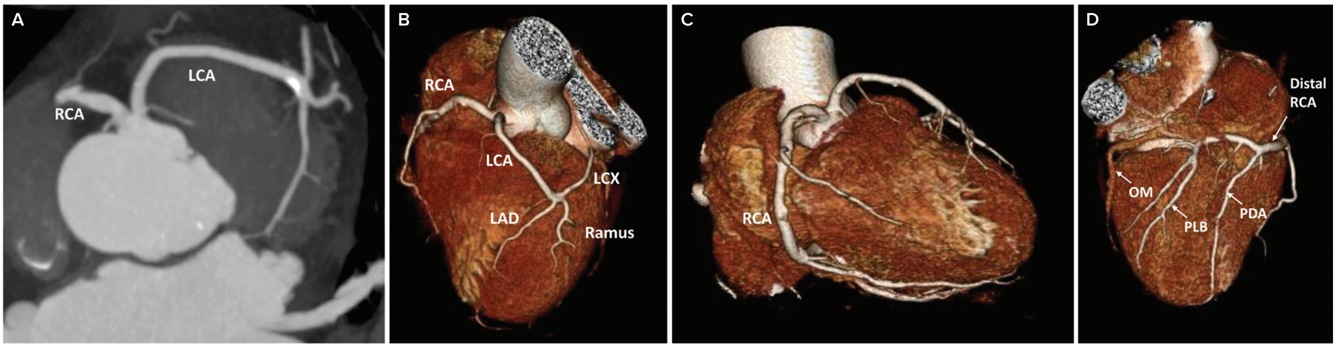
Case 2

A 49-year-old woman was evaluated at the cardiologist's office for complaints of chest pain and intermittent palpitations. An exercise stress echocardiogram achieved 93% of predicted maximum heart rate and demonstrated preserved ejection fraction and no evidence of inducible ischemia. Chest pain was not reproducible with exertion. A 1-week Holter monitor did not reveal any significant arrhythmias. Due to ongoing intermittent chest pain, a coronary CTA was performed.

CTA revealed an anomalous left main coronary artery (L-AAOCA) arising from the RCA, following a retro-aortic course to the left side of the heart (*Figure 2A*). The left main artery was relatively small in caliber and bifurcated into the LAD and LCX arteries (*Figure 2B*). The RCA originated high above the right coronary sinus and gave off a large posterior descending artery that wrapped around the apex to supply the distal left ventricular myocardium (*Figure 2C*). No significant atherosclerotic disease was noted in any coronary segments.

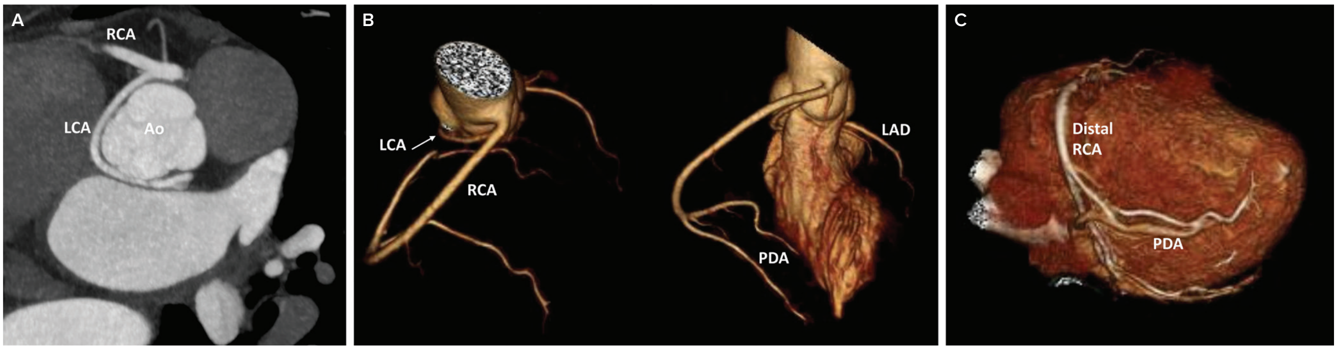
The decision was made to conservatively treat the patient, given the benign course of her left main artery, negative functional assessment and lack of EKG changes, and troponin elevation. The patient continued to

Figure 1: Coronary CT Angiography of Patient 1



A: Anomalous left main coronary artery arising from the right coronary sinus, following a pre-pulmonic course. B: The left coronary artery bifurcates into branches that supply the left anterior descending, ramus, and circumflex arteries. C: Hyperdominant right coronary artery originating normally from the right coronary sinus. D: Right coronary artery giving rise to large caliber posterior descending artery, posterior lateral branches and obtuse marginal artery. LAD = left anterior descending; LCA = left coronary artery; LCX = left circumflex; OM = obtuse marginal; PDA = posterior descending artery; PLB = posterior lateral branches; RCA = right coronary artery.

Figure 2: Coronary CT Angiography of Patient 2



A: Anomalous left main coronary artery arising from the right coronary artery, following a retro-aortic course to the left side of the heart. B: Small caliber left coronary artery bifurcating into the left anterior descending and circumflex arteries. C: Right coronary artery branching into a large posterior descending artery that wraps around the apex to supply the distal left ventricular myocardium. AO = aorta; LAD = left anterior descending; LCA = left coronary artery; LCX = left circumflex; PDA = posterior descending artery; RCA = right coronary artery.

follow up throughout the years with outpatient stress testing that was persistently negative, and she continues to fare well.

Case 3

A 48-year-old woman with a recent history of pulmonary embolism presented to the emergency department with substernal chest pain. Serial high-sensitivity troponin levels were within normal limits. As part of her evaluation, the patient underwent an EKG, which showed no changes suggestive of ischemia, and an echocardiogram, which demonstrated preserved left ventricular ejection fraction and no significant abnormalities. Coronary CTA revealed a dominant RCA arising anomalously from the left coronary sinus (R-AAOCA; *Figure 3A*), with an interarterial course between the aorta and pulmonary artery (*Figure 3B*). The proximal segment of the RCA featured a 12 mm intramural segment, an elliptical orifice, and an acute-angle takeoff from the left sinus – characteristics considered high risk (*Figures 3C and 3D*). The RCA was free of atherosclerotic disease.

Physiological testing with fractional flow reserve (FFRCT) indicated only a moderate reduction in lesion-specific fractional flow reserve at 0.87 and significant hemodynamic decrease in the distal vessel, with an FFR value of 0.71. Stress exercise myocardial perfusion imaging (achieved 94% of predicted maximum heart rate) was negative for ischemia and symptoms were not reproduced during exertion.

Given the non-cardiac nature of her chest pain and resolution of her symptoms, the patient was discharged with close follow-up with cardiology. At 3 months follow-up, she remained active and asymptomatic, and repeat exercise stress myocardial perfusion imaging was negative.

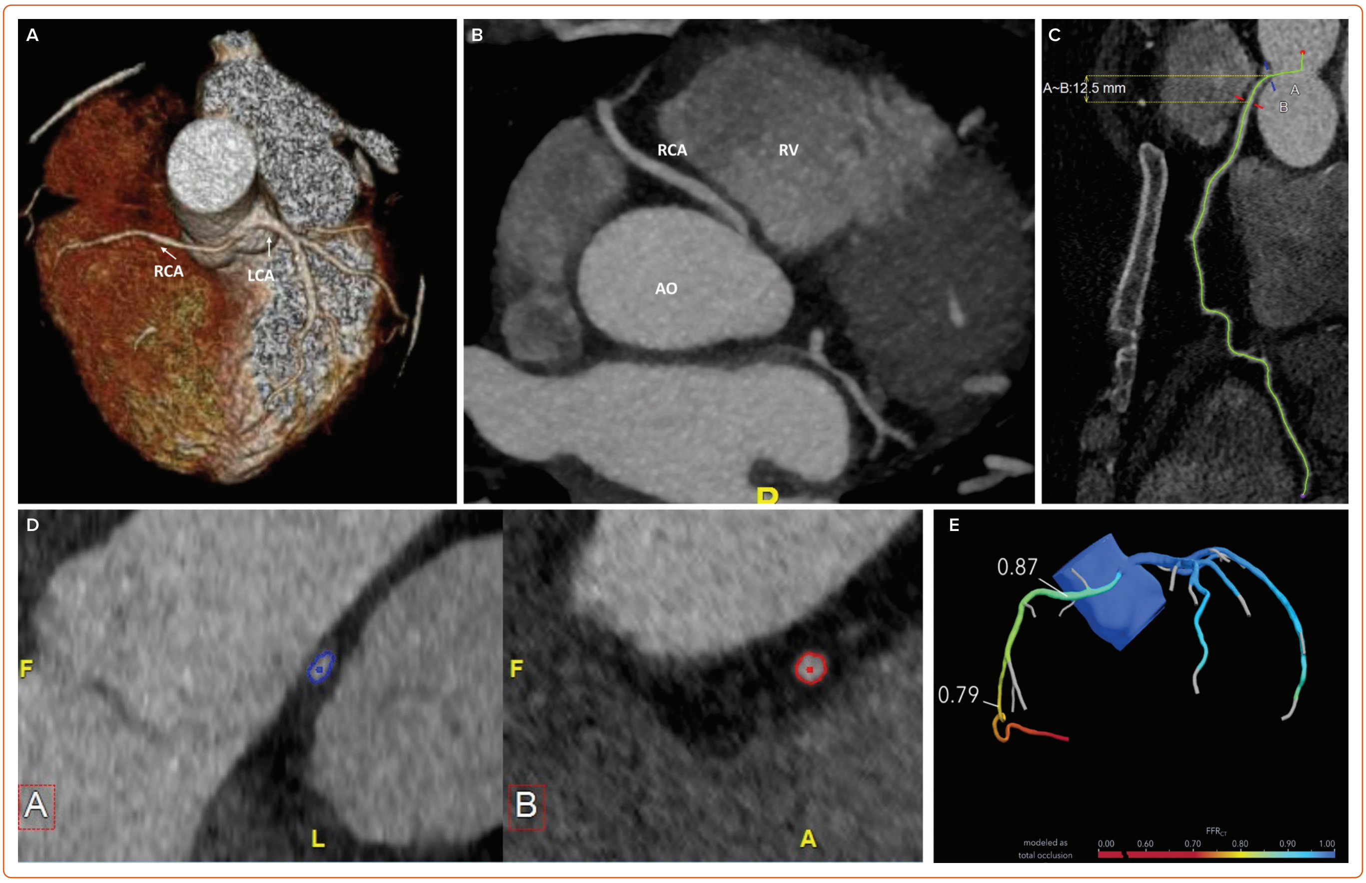
Discussion

As illustrated by the cases presented here, coronary CTA is highly effective for diagnosing AAOCA and precisely delineating anatomical details and high-risk features.

Current guidelines vary in their recommendations for optimal management based on the specific artery involved and its anatomical course. Both US and European guidelines recommend surgical intervention when ischemia is proven, or symptoms are present. However, in the absence of ischemia, the guidelines diverge on management strategies for high-risk features.[1,2] US guidelines advocate for surgical intervention in all patients with L-AAOCA, regardless of ischemia or symptoms. In contrast, European guidelines recommend surgery only if high-risk features are present or if the patient is under 35 years of age.

Guidelines also recommend physiological evaluation for patients with AAOCA, ideally using non-pharmacological functional imaging methods such as echocardiography, nuclear imaging, cardiac magnetic resonance stress testing, or invasive assessment with intracoronary imaging and

Figure 3: Coronary CT Angiography and Fractional Flow Reserve of Patient 3



A: RCA arising anomalously from the left coronary sinus. B: RCA with an interarterial course between the aorta and pulmonary artery. C: Acute takeoff angle of RCA with proximal intramural segment measuring 12 mm. D: Elliptical orifice of RCA ostium (A) compared with round contour of RCA beyond the intramural segment. E: Fractional flow reserve (FFR) CT showing moderate hemodynamic decrease immediately after the narrowed segment (FFR 0.87) but severe decrease in the distal vessel, with an FFR value of 0.71. AO = aorta; LCA = left coronary artery; RCA = right coronary artery; RV = right ventricle.

functional assessment.[3,4] When dynamic compression is suspected, dobutamine or physical stress testing is recommended rather than adenosine or regadenoson, which are less sensitive in detecting both fixed and dynamic components of stenosis present in patients with AAOCA. In addition, single-photon emission CT can be falsely negative due to subendocardial ischemia below its spatial resolution.[5] However, there is no general consensus on the optimal imaging modality. Additionally, there is a lack of prospective studies assessing the prognostic value of ischemia detection or the impact of intervention on reducing clinical event risk.

Although FFRCT has not been validated in patients with AAOCA, recent studies have suggested an emerging role in functional ischemia assessment in these patients to help guide treatment.[6,7] In addition, while moderate hemodynamic reductions measured by FFRCT have not been consistently associated with higher long-term cardiovascular event rates, significantly reduced FFRCT values may have a prognostic impact.[8] However, FFRCT measurement is limited to the evaluation of a fixed component such as coronary artery disease which might limit its sensitivity for dynamic compression and subendocardial ischemia, and therefore limit the applicability of FFRCT in patients with AAOCA.[9]

Future novel computational fluid dynamics models are needed to address the 'dynamic compression' component in patients with an AAOCA. An additional important consideration is that the management approach of AAOCA in young athletes may differ from older patients, given the increased risk of sudden cardiac death with strenuous exercise when high-risk anatomic features are present. Therefore, a lower threshold for consideration of surgical evaluation should be adopted.

In the cases presented here, the first patient had an L-AAOCA arising from the right coronary sinus which was incidentally caught due to screening purposes. Despite the literature describing pre-pulmonic left main as having a benign course, the 2018 American College of Cardiology/American Heart Association guidelines recommend surgical repair (class 2a) for all anomalous origin of the coronary arteries involving the left main.[1] However, given this was an incidental finding and the patient was not experiencing any chest pain, the decision was made to monitor him.

The second case is particularly notable, as the left main coronary artery originated from the RCA rather than the right coronary sinus, which is rarely reported in the literature. Current guidelines do not provide specific recommendations for this type of AAOCA. However, CTA did not demonstrate high-risk anatomical features, based on the retro-aortic course and the absence of slit-like ostium, acute takeoff angle, intramural course, or significant stenosis. Based on the CTA findings, stress testing was not absolutely necessary given that anatomy alone excluded significant ischemic risk. Therefore, the patient was managed conservatively and remains stable.

The third case demonstrates an R-AAOCA with an interarterial course and a long intramural segment, which have been described as high-risk features in literature. Current recommendations are to surgically manage in the presence of chest pain.[3] In this particular patient the chest pain was thought to be secondary to pulmonary embolism rather than having a cardiac etiology. In addition, although terminal vessel FFRCT was significantly reduced, FFRCT more proximally in the vessel revealed only moderate reductions in lesion specific ischemia. Measuring FFRCT just beyond the stenosis has been shown to improve the diagnostic performance of FFRCT and ensures that values are due to lesion-specific ischemia which reduces the rate of unnecessary invasive procedures.[10] This could explain the discrepancy between terminal vessel FFRCT results and stress myocardial perfusion imaging. In addition, it is important to acknowledge that the absence of ischemia and/or chest pain may not effectively prognosticate sudden cardiac death risk in the presence of high-risk anatomic features and it remains unclear whether asymptomatic ischemia in AAOCA warrants surgical intervention to prevent future myocardial ischemia. Ultimately, the care team elected to proceed with conservative management. Follow-up nuclear stress testing was negative, and the patient did not experience recurrent episodes of pain. Long-term follow-up is crucial in this patient population to assess the progression of ischemia or symptoms.

All three cases highlight the heterogeneity in anatomical variations and clinical presentations in patients with AAOCA and emphasize the need to tailor treatment based on individual cases rather than following guidelines which provide general recommendations.

Conclusion

Despite existing recommendations, the management of AAOCA remains tailored to the individual, based on factors such as symptoms, presence of ischemia, high-risk features, patient age, and comorbidities. It is important to emphasize that increased detection of AAOCA due to the broader use of CT should not automatically lead to increased revascularization procedures. In addition, stress testing and FFRCT have limitations in this patient population and the presence of high-risk anatomic features should prompt further evaluation even in the absence of ischemia or symptoms. ❑

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