

A Rare Anatomical Variant: Congenital Absence of Right Coronary Artery with Super-dominant Left Circumflex Artery

Randy Dwirizaldi Hidayat, , Faras Hilmy, , Iyus Maolana Yusup, , Dian Komala Dewi .

Abstract

Background

Congenital absence of the right coronary artery (RCA) is an exceptionally rare Single Coronary Artery variant where right ostium development fails, making global myocardial perfusion single-trunk dependent. This study highlights the critical diagnostic role of Cardiac Computed Tomography Angiography (CCTA) in identifying this high-stakes anomaly.

Case Presentation

A 77-year-old male with hypertension presented with a two-year history of progressive exertional dyspnea. A 256-slice CCTA was performed due to persistent symptoms, revealing a complete absence of the right coronary ostium and an RCA calcium score of 0. The circulation exhibited complete left dominance via a markedly elongated, super-dominant left circumflex (LCx) artery supplying the entire posterior and inferior myocardial territories. A critical, heavily calcified stenosis was identified in the middle segment of the left anterior descending (LAD) artery, which was subsequently managed with a successful confirmatory invasive angiography and percutaneous coronary intervention (PCI).

Conclusion

Congenital RCA absence is a critical anomaly easily misdiagnosed as chronic total occlusion. CCTA serves as an invaluable non-invasive roadmap guiding targeted secondary prevention.

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Introduction

Congenital absence of the right coronary artery (RCA) is an extraordinarily rare cardiovascular malformation within the anatomical spectrum of a Single Coronary Artery (SCA) network [1,2]. While normal myocardial perfusion is sustained by independent dual-source arteries, an isolated left SCA network forces the entire myocardial architecture to rely exclusively on a single coronary ostium to maintain cardiac output^{3,4}. Large-scale retrospective studies estimate the overall prevalence of an isolated SCA to range between 0.024% and 0.066% among patients undergoing angiographic evaluation, with complete territorial agenesis of the RCA trunk standing out as one of the least frequently documented variants [1,2].

The embryological pathogenesis of this malformation stems from either a primary developmental failure during the budding phase of the right coronary cord or a premature involutonal occlusion of the proximal right vascular plexus during the first trimester of intrauterine life [3]. Although many individuals remain asymptomatic for decades, this single-vessel dependent heart presents a severe pathophysiological liability due to the lack of alternative routes for collateral circulation [2,4]. Consequently, if atherosclerotic plaque progression develops within the lone functioning tree, patients become exceptionally vulnerable to extensive transmural ischemia, malignant arrhythmias, or sudden cardiac death [4].

Historically, complex coronary anomalies were discovered incidentally during conventional invasive coronary angiography (ICA), which often encounters high engagement failure rates when managing an absent ostium[2,5]. In the modern era, multi-slice Cardiac Computed Tomography Angiography (CCTA) has emerged as the premier non-invasive tool, providing superb spatial resolution and three-dimensional multiplanar reconstructions to meticulously delineate complex vascular trajectories and plaque morphology[5]. This case report highlights the critical diagnostic role of 256-slice CCTA in non-invasively identifying a congenital absence of the RCA with a super-dominant left circumflex (LCx) artery, providing a precise anatomical roadmap for a subsequent successful confirmatory intervention and percutaneous coronary intervention (PCI).

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Case Presentation

A 77-year-old male with hypertension presented with a two-year history of progressive exertional dyspnea and easy fatigability. He described the dyspnea as a heavy pressing sensation on his chest during physical activity. He denied typical anginal pain, palpitations, or lower extremity edema. His medical history was significant only for well-controlled hypertension treated with amlodipine 5 mg once daily. Physical examination and laboratory profiles, including a lipid panel, were unremarkable. Baseline transthoracic echocardiography (TTE) demonstrated normal chamber sizes with no regional wall motion abnormalities. Given the distinct clinical mismatch between his unremarkable echocardiographic findings and debilitating symptoms, a 256-slice cardiac computed tomography angiography (CCTA) was performed to evaluate the coronary architecture. The coronary artery calcium scoring protocol yielded a total Agatston score of 219 (left circumflex [LCx]: 177, left main artery [LMA]: 38, left anterior descending [LAD]: 4). Notably, the calcium scores for both the right coronary artery (RCA) and posterior descending artery (PDA) were absolute 0.

Contrast-enhanced CCTA revealed a complete congenital absence of the RCA, characterized by the absolute non-visualization of the right coronary ostium and its proximal trunk at the right aortic sinus. Consequently, the coronary circulation exhibited complete left dominance. The LMA originated normally and bifurcated into the LAD and LCx. The LCx presented as a super-dominant, markedly elongated vessel that wrapped continuously around the atrioventricular groove, extending well past the cardiac crux to retrogradely supply the entire posterior and inferior myocardial territories.

Evaluation of the atherosclerotic burden across this single network revealed a critical, heavily calcified plaque in the middle segment of the LAD, resulting in severe luminal stenosis, alongside mild non-obstructive plaques in the LMA and proximal LCx. Extra coronary findings included a localized Grade 2 calcification at the non-coronary cusp (NCC) of the aortic valve with an Agatston score of 102.

Guided by the precise CCTA roadmap, the patient was referred for conventional invasive coronary angiography (ICA). The catheterization confirmed the complete absence of the right coronary origin, verifying the retrograde perfusion from the left network with 100% diagnostic accuracy. Given the critical nature of this single-vessel dependent network, a successful Percutaneous Coronary Intervention (PCI) was immediately performed with a drug-eluting stent deployed across the severe mid-LAD lesion. Post-procedure, the patient's exertional dyspnea completely resolved, and he was discharged on guideline-directed medical therapy.

Discussion

Non-visualization of the RCA on standard imaging presents a significant diagnostic challenge, requiring a

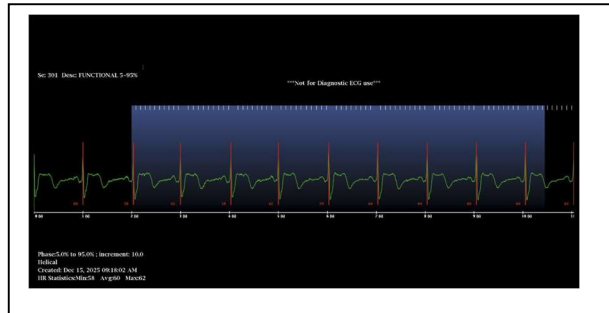


Figure 1. Graphical representation of the prospectively electrocardiographically triggered (ECG-gated) coronary computed tomography angiography (CCTA) protocol. The acquisition window is strategically targeted during the mid-to-end diastole phase to ensure optimal image resolution and eliminate cardiac motion artifacts.

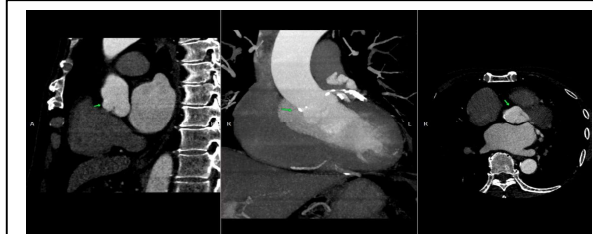


Figure 2. Axial contrast-enhanced cardiac computed tomography angiography (CCTA) reconstruction at the level of the aortic root. The image definitively demonstrates the true congenital absence of the right coronary artery (RCA), characterized by the absolute non-visualization of the right coronary ostium at the right aortic sinus (indicated by the area of interest). Concurrently, the left coronary network originates normally from the contralateral left aortic sinus.

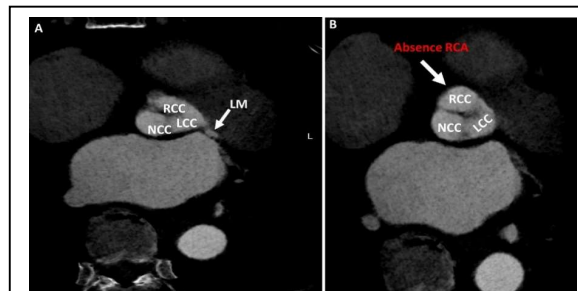


Figure 3. Axial cross-sectional CCTA image focusing on the aortic sinus morphology. The landmark labeled "LM" confirms the normal structural origin of the left main coronary artery arising from the left aortic sinus. Conversely, the contralateral right aortic sinus displays complete structural agenesis, with no identifiable right coronary ostium or proximal arterial trunk, confirming a single left coronary network.

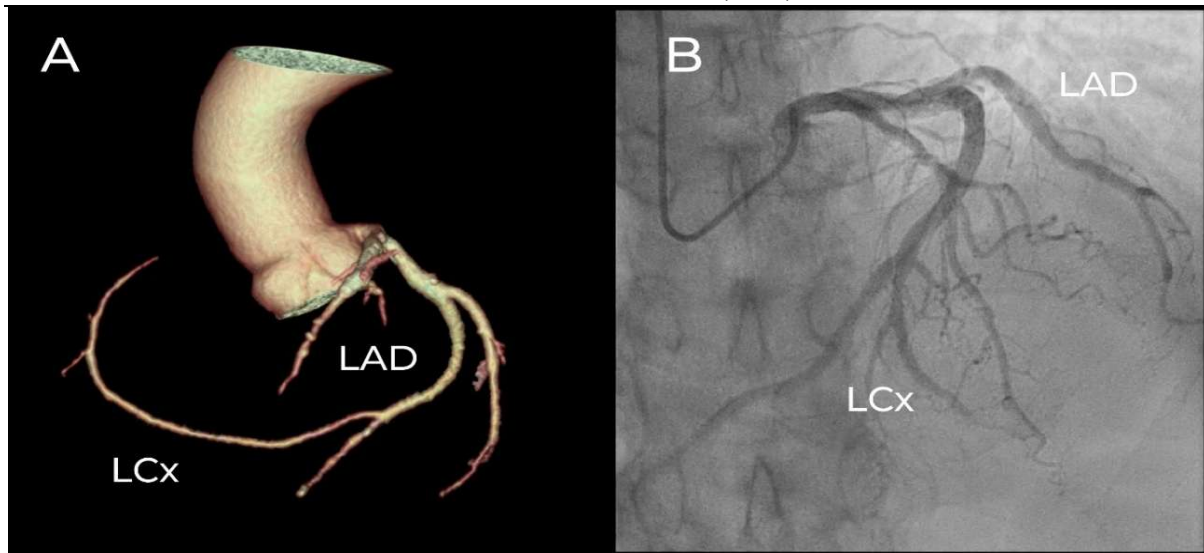


Figure 4. Dual-modality anatomical correlation of the single left coronary artery (SCA). (A) Invasive coronary angiography (ICA) confirms the absence of the right coronary artery (RCA) ostium, with retrograde perfusion supplying the inferior-posterior territories. (B) Corresponding 3D volume-rendered (3D-VR) CCTA reconstruction highlights the hypertrophied, super-dominant left circumflex (LCx) artery wrapping around the atrioventricular groove to sustain global myocardial perfusion.

a clear differentiation between true congenital agenesis, LAD artery, categorized under the CAD-RADS an acquired atherosclerotic Chronic Total Occlusion framework⁵. This finding revealed that the patient's (CTO), and an anomalous coronary origin [5,7]. In our single-vessel dependent heart was in a highly vulnerable patient, a true congenital absence was confirmed by the state, as any progression of the LAD lesion could compromise global anterior myocardial perfusion and a residual blind-ended vascular stump at the right without any safety net.

aortic sinus [6]. This conclusion was reinforced by a dedicated Agatston calcium score of 0 along the expected RCA and PDA tract, effectively ruling out a long-standing, heavily calcified occluded native vessel [5]. Concurrently, multiplanar reconstruction verified the definitive classification of a single left coronary artery network [7].

To compensate for the embryonic developmental failure of the right coronary system, the heart undergoes profound intrauterine hemodynamic remodeling, triggering structural hypertrophy and compensatory elongation of the left circulation [3,8]. In this specific architecture, the LCx artery functions as a "super-dominant" vessel [2,6]. As demonstrated in our patient's CCTA, this super-dominant LCx tracked continuously along the left atrioventricular groove, extended well past the cardiac crux, and retrogradely entered the right atrioventricular sulcus to successfully supply the posterior and inferior myocardial walls—territories normally served exclusively by a native RCA [5,8].

The primary clinical hazard of an isolated single left coronary system is its extreme vulnerability to atherosclerotic progression [2,4]. Because global myocardial perfusion is entirely single-trunk dependent, a hemodynamically significant stenosis cannot be bypassed or mitigated by alternative contralateral collateral channels [4,9]. In this case, the pre-procedural CCTA played a pivotal role by identifying a critical, severe calcified stenosis in the middle segment of the

Guided by the precise CCTA-derived anatomical roadmap, subsequent invasive coronary angiography (ICA) confirmed the absent RCA origin and retrograde left circulation with 100% diagnostic accuracy [10]. This dual-modality correlation provided the definitive diagnostic confirmation required to safely proceed with an immediate, successful Percutaneous Coronary Intervention (PCI). Stent implantation across the severe mid-LAD lesion successfully secured the patient's lone functioning vascular tree and immediately restored optimal anterograde flow.

Conclusion

This case provides an illustrative example of the remarkable boundaries of long-term human myocardial adaptation, demonstrating that a single left coronary network can successfully sustain global cardiac function into late senescence despite the complete congenital absence of the RCA [2]. For radiology residents and practicing cardiovascular specialists, maintaining a high level of vigilance when evaluating asymmetric coronary distributions on cardiac CT is paramount [3,6,10]. Performing an initial CCTA-based evaluation is essential to prevent the erroneous diagnosis of an acquired atherosclerotic occlusion, avoid hazardous or unnecessary invasive procedural attempts to selectively engage a non-existent right ostium, and provide a precise anatomical roadmap that ensures safer, targeted planning for subsequent high-stakes confirmatory invasive angiography and percutaneous therapeutic

interventions^{3,6}.

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