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# A Rare Anatomical Variant: Congenital Absence of the Right Coronary Artery with Left Circumflex Artery Supplying the Right Coronary Artery (RCA) Territory

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Statistical Analysis C  
Data Interpretation D  
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**Conflict of interest:** None declared

**Patient:** Female, 66-year-old  
**Final Diagnosis:** Coronary artery anomalies  
**Symptoms:** Chest discomfort • shortness of breath  
**Medication:** —  
**Clinical Procedure:** Coronary angiography  
**Specialty:** Cardiology

**Objective:** Rare disease

**Background:** While most coronary congenital anomalies are uncommon, comprising less than 2% of the general population, a single coronary artery is an especially rare finding within this broader category. It involves a lone vessel forming a single ostium from the aortic trunk. This vessel is then the source through which cardiac blood supply originates. Such congenital coronary artery anomalies are rare but can have clinically relevant consequences such as sudden death and other cardiac complications. Nonetheless, they are usually incidental findings and can be asymptomatic. This report discusses the case of a female patient found to have an absence of the right coronary artery, with the left circumflex supplying the right coronary artery territory.

**Case Report:** We report a rare case of a 66-year-old woman who presented with substernal chest pain and shortness of breath. Vital signs, laboratory work-up, and resting electrocardiogram did not reveal an underlying etiology. However, an exercise stress test was positive. Left heart catheterization was performed and revealed a dominant left circumflex artery supplying the entire right coronary territory. Further imaging revealed the absence of a separate right coronary artery ostium. The patient was treated with a beta-blocker, high-intensity statin, and a nitrate.

**Conclusions:** Single left coronary artery, especially with the absence of the right coronary artery, is particularly rare, but can have significant clinical implications. Prompt diagnosis is important but challenging considering the variable presentation of this condition. It is important to review diagnostic modalities available and the treatment for patients.

**Keywords:** Cardiac Catheterization • Congenital Abnormalities • Coronary Angiography • Coronary Vessel Anomalies

**Abbreviations:** LV – left ventricle; CT – computed tomography; CTA – computed tomography angiography; RCA – right coronary artery; LCX – left circumflex; LMCA – left main coronary artery; ECG – electrocardiogram; TTE – transthoracic echocardiogram; TEE – transesophageal echocardiogram


Full-text PDF: <https://www.amjcaserep.com/abstract/index/idArt/932248>



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## Background

Congenital coronary anomalies are relatively uncommon and comprise less than 2% of the general population [1]. Single coronary arteries are very rare, with an incidence on autopsy of 0.014-0.066% [2-4]. Single coronary artery (SCA) is an anatomical entity abnormality. It occurs when only 1 ostium is formed from the aortic trunk, which is then the basis of blood flow to the heart [5]. The underlying etiology is still unclear. A number of reports have been published in countries with high rates of consanguineous relationships [6]. While consanguinity has been associated with multiple cardiac conditions, no clear correlation with SCA has been determined [7]. Saudi Arabia, a nation with an estimated 57.7% prevalence of consanguinity, does not appear to have a greater prevalence of SCA. However, considering the low incidence of this condition, further studies are needed before a conclusion can be drawn.

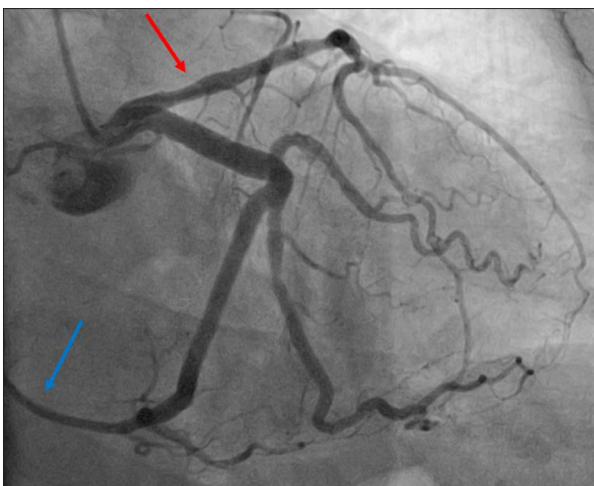
While rare, the complications resulting from this unique pathophysiology are highly significant and can even be as extreme as sudden death [8]. Anatomy resulting in an interarterial course is especially associated with increased mortality. This is often observed with an anomalous vessel originating from the right coronary cusp and traveling between the aorta and the main pulmonary artery to supply the left heart [9]. Furthermore, the process of diagnosis can often be complex, with multiple modern imaging modalities available. The best course of treatment is still not agreed upon, but both medical and surgical management have been discussed. As such, it is important to be

aware of such anomalies when they do arise. Here, we present a clinical case of a patient found to have single coronary syndrome with absent RCA after she presented for chest pain.

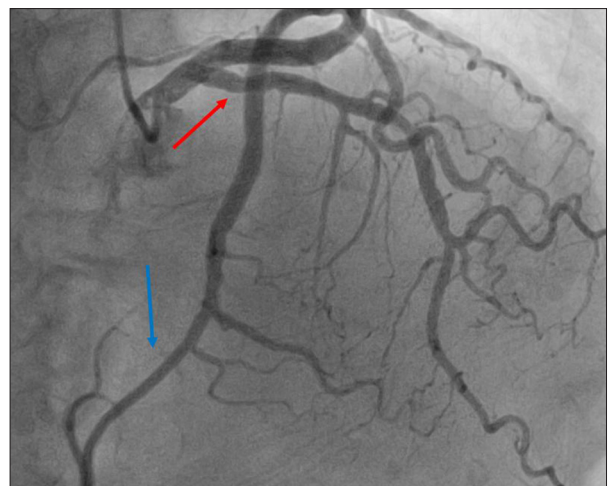
## Case Report

A 66-year-old white woman with a known history of hypertension and hyperlipidemia presented to the outpatient cardiology clinic for progressively worsening, non-radiating, retrosternal discomfort associated with shortness of breath over the past 3 to 4 weeks. Her vital signs were within normal limits and her physical exam was unremarkable. Resting ECG revealed normal sinus rhythm with nonspecific ST-T changes. While admitted, cardiac monitoring did not reveal arrhythmia and there was no correlation with pain symptoms. Cardiac enzymes were normal and BNP was normal. Routine blood work was unremarkable.

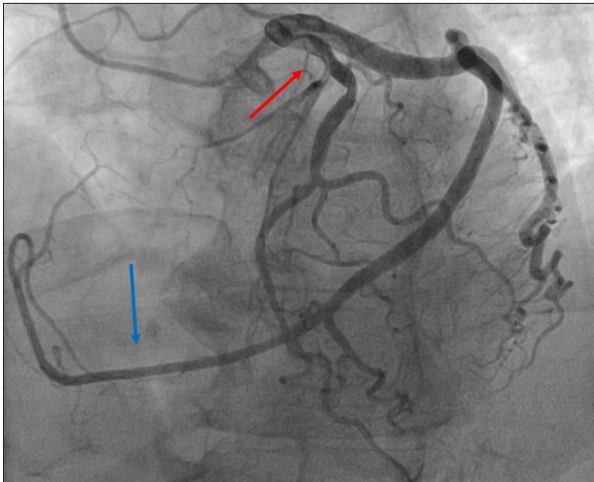
A transthoracic echocardiogram a few months prior to presentation revealed a normal left ventricular systolic function with mild mitral and pulmonary insufficiency. An exercise stress test was performed in the office, which revealed > 1 mm horizontal ST-segment depressions in the inferolateral leads, with interval development of chest pain at 88% predicted maximal heart rate. A left heart catheterization was performed and revealed mild non-obstructive coronary artery disease in the left system, with a dominant left circumflex artery supplying the entire right coronary artery territory (Figures 1-6). Mild disease



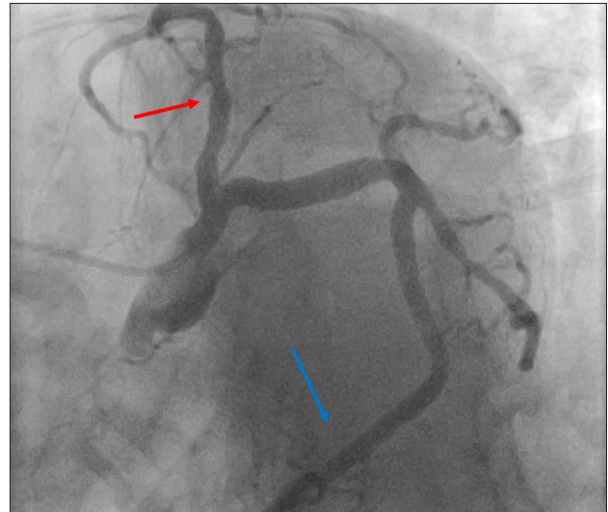
**Figure 1.** Coronary Angiography: Engagement of the left coronary artery via the right radial artery approach using a 6 French Judkins left (JL 3.5) catheter. The red arrow points toward the left anterior descending artery. The blue arrow points toward the left circumflex artery that supplies a territory of myocardium normally supplied by the right coronary artery. 14° right anterior oblique and 33° caudal view. Overall, there is evidence of mild epicardial coronary artery disease.



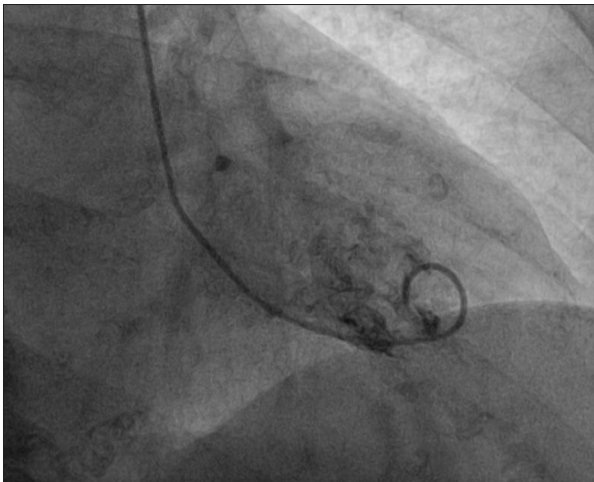
**Figure 2.** Coronary Angiography: Engagement of the left coronary artery via the right radial artery approach using a 6 French Judkins left (JL 3.5) catheter. The red arrow points toward the left anterior descending artery. The blue arrow points toward the left circumflex artery that supplies a territory of myocardium normally supplied by the right coronary artery. 18° right anterior oblique and 38° cranial view. Overall, there is evidence of mild epicardial coronary artery disease.



**Figure 3.** Coronary Angiography: Engagement of the left coronary artery via the right radial artery approach using a 6 French Judkins left (JL 3.5) catheter. The red arrow points toward the left anterior descending artery. The blue arrow points toward the left circumflex artery that supplies a territory of myocardium normally supplied by the right coronary artery. 24° left anterior oblique and 18° cranial view. Overall, there is evidence of mild epicardial coronary artery disease.



**Figure 4.** Coronary Angiography: Engagement of the left coronary artery via the right radial artery approach using a 6 French Judkins left (JL 3.5) catheter. The red arrow points toward the left anterior descending artery. The blue arrow points toward the left circumflex artery that supplies a territory of myocardium normally supplied by the right coronary artery. 41° left anterior oblique and 42° caudal view. Overall, there is evidence of mild epicardial coronary artery disease.



**Figure 5.** Ventriculography: Performed in the right anterior oblique view (25°) using a 6 French Pigtail catheter. Left ventricular systolic function appears preserved (ejection fraction estimated to be in the range of 60-65%).

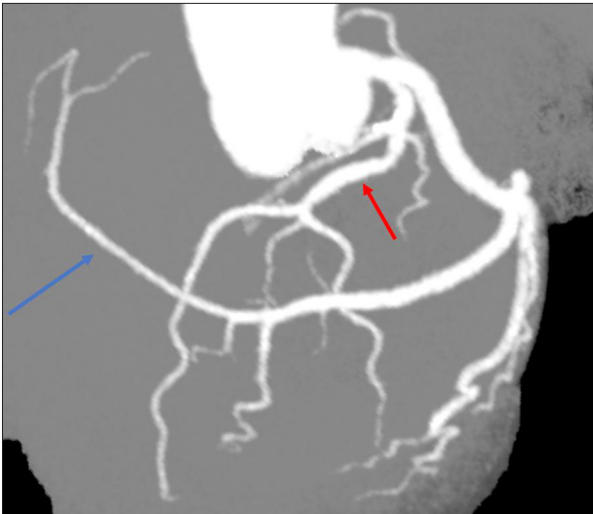


**Figure 6.** Ascending aortogram performed using a 6 French Pigtail catheter in the left anterior oblique view (30°). Aortography revealed absence of opacification of the right coronary artery confirming its absence.

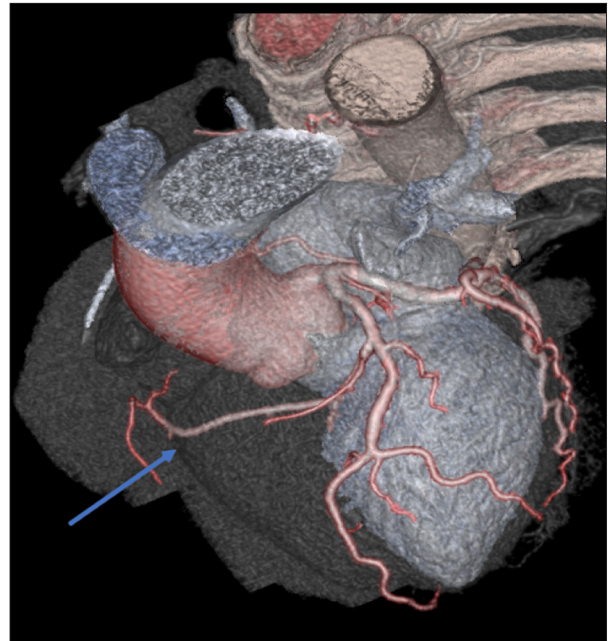
was noted in the OM branch, but was considered unlikely to be related to a patient's symptoms. A subsequent aortogram revealed the absence of a separate right coronary artery ostium from the aorta. Coronary CTA was obtained and confirmed the findings on cardiac catheterization (Figures 7, 8). Furthermore, no significant obstructive disease was noted. There was mild non-calcified plaque noted with luminal narrowing ranging from 0% to 25%. This was considered unlikely

to be contributing to the chest pain. The patient was treated with a beta-blocker, high-intensity statin, and a nitrate. While follow-up visits were made for the patient, she is yet to return to the clinic. Considering there have been reports of symptoms managed with aggressive medical therapy, maximizing





**Figure 7.** Coronary analysis obtained from coronary CTA. 30° left anterior oblique and 30° cranial view showing absence of a right coronary artery with the left circumflex supplying the distribution normally supplied by a native right coronary artery (blue arrow). The red arrow points to the left anterior descending artery.



**Figure 8.** Volume-rendering image obtained from coronary CTA. 30° left anterior oblique and 60° cranial view showing absence of a right coronary artery with the left circumflex supplying the distribution normally supplied by a native right coronary artery (blue arrow).

her medical management will be the first step if symptoms continue. If significant symptoms persist despite maximally tolerated medical management, she will have to be evaluated for further intervention.

## Discussion

Single coronary artery (SCA) anomalies are extremely rare, with an incidence of 0.014-0.066%, and they usually present in conjunction with other embryonic structural abnormalities [2-5]. The absence of a right coronary artery with a dominant left circumflex artery supplying the right coronary artery territory is a rare entity within the SCA anomalies, as its prevalence is low in cardiovascular malformations [1]. Development of coronary anatomy is a complicated subject, but may shed light on the underlying pathophysiology. It was originally thought that the coronary arteries were outgrowths from the aorta itself [10]. However, this was refuted with the demonstration of the major coronary arteries without the presence of aortic orifices. Conversely, once the orifices were visible, so too were the coronary arteries. This has led to the belief that the coronary arteries may grow into the aorta rather than growing out of it. The driving and directing forces of this growth are unclear. Further understanding of coronary artery genesis may reveal the underlying etiology of SCA. Thus far, it has been postulated that failure of a culprit artery to develop or development then occlusion of an artery while in embryo may explain the resulting single ostium and artery [10].

As the patient ages, embryological abnormalities can have clinical consequences. In general, the RCA supplies the SA and AV nodes. If there is a single left coronary artery supplying the SA and AV nodes in conjunction with the rest of the cardiac structures, one can postulate the early degeneration of SA or AV node due to inadequate blood supply [1]. Degeneration of SA or AV nodes can produce arrhythmias and ischemia. Moreover, the overworked single left coronary artery can potentially become dilated, form fistulas, and potentially compress nearby structures or branches and cause ischemia, and incur early endothelial damage, which can cause early atherosclerosis. In 2001, Virmani and associates examined 21 patients with SCA. Of these, 10 were noted to have died from a cardiac related cause. Importantly, 5 experienced sudden death [11]. Thus, it is essential to diagnose patients with this anomaly early, perform active surveillance, and have a low threshold for invasive procedures.

While usually an incidental finding, when symptomatic, patients with SCA present with a myriad of symptoms, which may include palpitations, syncope, heart failure, chest pain, and even sudden death [12-14]. Although there have been cases of symptomatic patients early in life, young patients are usually asymptomatic, while older patients begin to develop symptoms concomitant with the development of atherosclerotic disease [11]. This is likely the case in our patient, who presented in her late 60s. Hypertension and hyperlipidemia may

have contributed to her presentation. She did not smoke, use illicit drugs, or drink alcohol. Patients may present with angina and nonspecific ST-T changes on ECG, or the presentation may be more sudden with little time for evaluation [11]. Thus, such patients are often misdiagnosed as having coronary artery disease, and without multi-modality imaging or invasive procedures, they remain undiagnosed.

The diagnosis of this entity can be challenging, especially when patients are asymptomatic, which is the case in most young patients. Multi-modality imaging with coronary computed tomography angiography (coronary CTA) and cardiac MRI (CMR) are useful tools to diagnose patients prior to consideration of a left heart catheterization and its associated complications [12]. Moreover, CMR has been used to elucidate accurate ejection fraction, early diastolic dysfunction, diagnosis of other structural abnormalities, and degree of fibrosis or damaged myocardium. Nonetheless, considering that an interarterial course of an anomalous vessel is associated with greater risk, it is important to assess the entire pathway of the vessel [9]. However, most patients who are symptomatic will usually have a coronary angiogram in an ischemic work-up prior to further imaging [13].

Once diagnosed, there is unfortunately no criterion standard for treatment. Many anomalies are incidental findings and do not warrant specific treatment in patients with no related symptoms. Treatment may also be driven by managing co-morbidities. Non-invasive treatment modalities include lipid-lowering, blood pressure management as well as anti-platelet therapy. Beta-blockade or medical rhythm control may also be considered. While multiple case reports have documented statin use in management, no specific data support this practice. Statin therapy has demonstrated a reduction in angina in patients with non-obstructive coronary artery disease and thus may have a role [15]. Nonetheless, further studies are needed to

establish long-term benefits. More invasive therapy can also be considered when clinically warranted. This includes PCI, placement of a pacemaker, and CABG [13]. Furthermore, surgical procedures such as creating a new ostium, termed “neo-ostium”, have been performed with success for similar conditions [16]. Surgical intervention is most often considered when high-risk features are present. Most often this involves an interarterial course of an anomalous artery or signs of ischemia. PCI may also be done if there is significant atherosclerotic disease. With this in mind, further study is required to elucidate the best management and balance potential benefits with risks [16].

## Conclusions

Congenital coronary anomalies, although rare, need a high clinical suspicion. Single left coronary artery supply, especially with an absence of the right coronary artery, is an extremely rare congenital anomaly. Early diagnosis and appropriate treatment, according to the patient’s clinical manifestations, are paramount. Left heart catheterization can be used to diagnose this anomaly but can be cumbersome because multiple projections, increased use of contrast, and increased radiation exposure are often encountered. Multi-modality imaging including coronary CTA and CMR are therefore very useful for diagnosis. Treatment, while not always warranted on incidental findings, often involves non-invasive medical management, although it can also require revascularization (PCI, CABG), pacemaker implantation, or surgical procedures.

## Declaration of Figures Authenticity

All figures submitted have been created by the authors who confirm that the images are original with no duplication and have not been previously published in whole or in part.

## References:

1. Yan GW, Bhetuwal A, Yang GQ, et al. Congenital absence of the right coronary artery: A case report and literature review. *Medicine*. 2018;97:e0187
2. Lipton MJ, Barry WH, Obrez I, et al. Isolated single coronary artery: Diagnosis, angiographic classification, and clinical significance. *Radiology*. 1979;130:39-47
3. Namgung J, Kim JA. The prevalence of coronary anomalies in a single center of Korea: origination, course, and termination anomalies of aberrant coronary arteries detected by ECG-gated cardiac MDCT. *BMC Cardiovasc Disord*. 2014;14:48
4. Turkmen S, Yolcu M, Sertcelik A, et al. Single coronary artery in 215140 patients undergoing coronary angiography. *Folia Morphol (Warsz)* 2014;73:469-74
5. Smith JC. Review of single coronary artery with report of 2 cases. *Circulation*. 1950;1:1168-75
6. Abdulshakour B, Saeed M, Taher M. Anomalous coronary artery anatomy with a single coronary ostium arising from the right coronary: A case report and literature review. *Int Med Case Rep J*. 2019;12:135-41
7. Smettei O, Sayed S, Abazid R. The prevalence of coronary artery anomalies in Qassim Province detected by cardiac computed tomography angiography. *J Saudi Heart Assoc*. 2017;29(2):84-89
8. Yamanaka O, Hobbs RE. Coronary artery anomalies in 126,595 patients undergoing coronary arteriography. *Cathet Cardiovasc Diagn*. 1990;21:28-40
9. Said S, Voogt W, Bulut S, et al. Coronary artery disease in congenital single coronary artery in adults: A Dutch case series. *World J Cardiol*. 2014;6(4):196-204
10. Silva Junior GO, Miranda SWS, Mandarim-de-Lacerda, CA. Origin and development of the coronary arteries. *Int J Morphol*. 2009;27(3):891-98
11. Virmani R, Burke A, Farb A. Sudden cardiac death. *Cardiovasc Pathol*. 2001;10:211-18
12. Kervancioglu M, Tokel K, Varan B, Yildirim SV. Frequency, origins and courses of anomalous coronary arteries in 607 Turkish children with tetralogy of Fallot. *Cardiol J*. 2011;18:546-51
13. Angelini P. Coronary artery anomalies – current clinical issues: Definitions, classification, incidence, clinical relevance, and treatment guidelines. *Texas Heart Inst J*. 2002;29:271-78

14. Li S, Ni Q, Wu H, et al. Diagnostic accuracy of 320-slice computed tomography angiography for detection of coronary artery stenosis: Meta-analysis. *Int J Cardiol.* 2013;168:2699-705
15. Manfrini O, Amaduzzie P, Bergami M, Cenki E. Effects of statin treatment on patients with angina and normal or nearly normal angiograms. *Eur Cardiol.* 2019;15:e15
16. Han WS, Park PW, Cho SH. Neo-ostium formation in anomalous origin of the left coronary artery. *Korean J Thorac Cardiovasc Surg.* 2011;44(5):355-57