

# Coronary steal syndrome due to right coronary ostium agenesis: a case report

## *Síndrome de robo coronario por agenesia del ostium derecho: reporte de caso*

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### ABSTRACT

A single coronary artery ostium with anomalous right coronary artery (RCA) origin from collateral circulation of the left anterior descending artery is a rare variant associated with multiterritorial ST elevation myocardial infarction and high surgical complexity. We present a 54-year-old man who suffered cardiac arrest secondary to pneumonia-induced myocarditis. Cardiac Computed Tomography Angiography (CCTA) revealed a 2.6 mm LAD-RCA fistula. Stress testing showed anterior ischemia with steal syndrome, and coronary angiography confirmed the anomaly. The patient underwent successful RCA revascularization.

**Keywords:** coronary vessels, coronary vessel anomaly, single coronary artery, coronary steal syndrome, collateral circulation.

### Abbreviations:

CCTA = Cardiac computed tomography angiography

LVEF = Left ventricular ejection fraction

RCA = Right coronary artery

STEMI = ST-Elevation myocardial infarction

### RESUMEN

Un ostium coronario único con origen anómalo de la arteria coronaria derecha (CD) a partir de la circulación colateral de la arteria descendente anterior izquierda (DA) es una variante rara asociada con infarto del miocardio con elevación del segmento ST multiterritorial y elevada complejidad quirúrgica. Presentamos el caso de un hombre de 54 años que sufrió paro cardíaco secundario a miocarditis inducida por neumonía. La angiotomografía coronaria reveló una fistula DA-CD de 2.6 mm. La prueba de esfuerzo mostró isquemia anterior con síndrome de robo, y la angiografía coronaria confirmó la anomalía. El paciente fue sometido a revascularización exitosa de la arteria CD.

**Palabras clave:** vasos coronarios, anomalía de las arterias coronarias, arteria coronaria única, síndrome de robo coronario, circulación colateral.

A single coronary ostium with an anomalous origin of the Right Coronary Artery (RCA), arising from collateral circulation of the left anterior descending artery represents an uncommon variation of a single coronary artery.<sup>1</sup> Although it generally has a benign prognosis, it can potentially

**How to cite:** Calderón-Abbo M, Gómez-Peña F, Zonana-Schatz E, Carrillo-Cornejo M, Lara-Calvillo AL, Pérez-Bañuelos A. Coronary steal syndrome due to right coronary ostium agenesis: a case report. Cir Card Mex. 2026; 11 (1): 31-34. <https://dx.doi.org/10.35366/122234>

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Received: 27/04/2025. Accepted: 14/10/2025.

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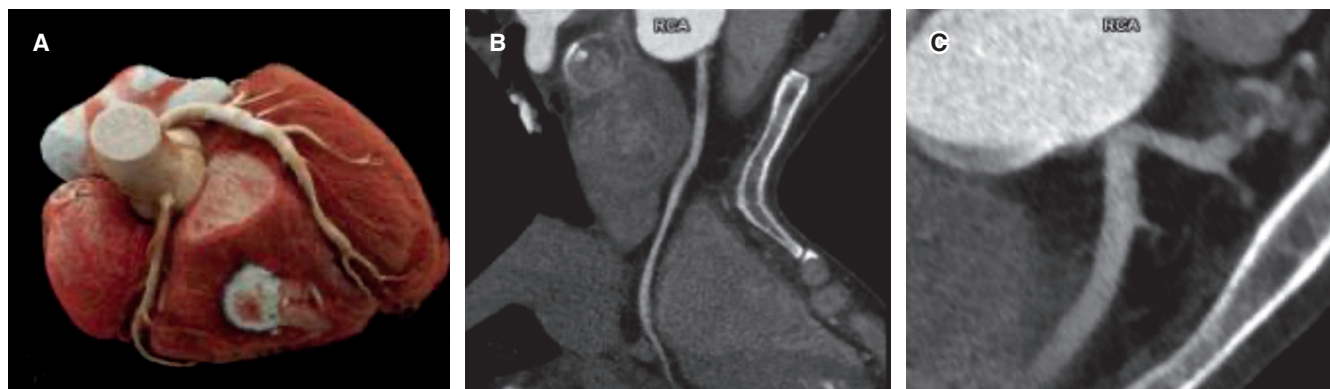
present as an ST-Elevation Myocardial Infarction (STEMI) affecting the anterior, inferior, and right ventricular regions simultaneously. The management of STEMI in anomalous coronary arteries can pose frequent surgical challenges, requiring interventional cardiologists and thoracic surgeons to be familiar with these unusual coronary patterns, as they may necessitate innovative and adaptable technical approaches.<sup>2</sup>

We present the case of a 54-year-old man with a two-week clinical course characterized by cough, orthopnea, progressive exertional dyspnea, and lower limb edema, complicated by septic shock secondary to community-acquired pneumonia, which led to septic myocarditis and a post-cardiac arrest state. After the patient was stabilized, a coronary CT angiography was performed, revealing a 2.6 mm fistula originating from the left anterior descending artery, located between its mid and proximal segments, with apparent communication toward the right coronary artery. An exercise stress test demonstrated exercise-induced angina and a steal phenomenon in the anterior wall. Subsequently, a diagnostic coronary angiography was performed to more precisely delineate the coronary anatomy and confirm collateral circulation from the left anterior descending artery.

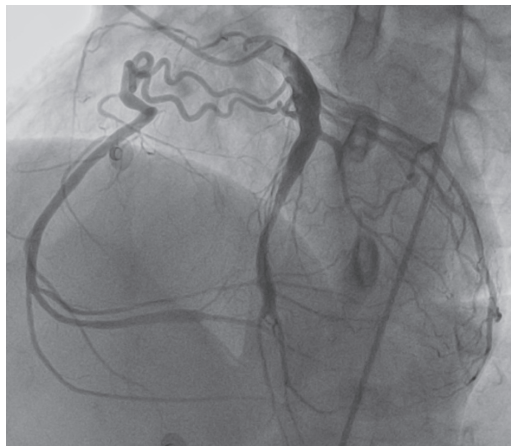
## CASE REPORT

A 54-year-old man with no significant past medical history developed a rapidly evolving cardiopulmonary syndrome. Initial complaints included a two week-history of cough, orthopnea, progressively worsening dyspnea on moderate exertion, and lower extremity edema. This presentation was complicated by septic shock secondary to community-acquired pneumonia, which precipitated septic myocarditis, a subsequent post-cardiac arrest state, and acute kidney injury classified as Kidney Disease: Improving Global Outcomes (KDIGO) stage IIIB. Initial laboratory

workup revealed atrial fibrillation and an elevated brain natriuretic peptide level of 1160 pg/ml. Following prompt resuscitation with vasopressors and broad-spectrum antibiotic therapy. A transthoracic echocardiogram revealed generalized biventricular hypokinesia with a markedly reduced Left Ventricular Ejection Fraction (LVEF) of 20%. Initial intensive management included vasopressors for hemodynamic stabilization, dobutamine to enhance cardiac output, and furosemide for decongestion. Atrial fibrillation was controlled with amiodarone, digoxin, and anticoagulation. The resulting clinical improvement reflected a modest improvement in systolic function with an LVEF rising to 30%. However, there remained evidence of a dilated left ventricle with eccentric hypertrophy, hypokinesia in the inferoseptal, basal, and mid segments, and biatrial enlargement. A stress test was performed, which demonstrated exercise-induced angina and a steal phenomenon in the anterior wall. In parallel with these echocardiographic findings, a Cardiac Computed Tomography Angiography (CCTA) was performed as part of the heart failure workup. The CCTA revealed a 2.6 mm fistula originating from the left anterior descending artery, located between the mid and proximal thirds, with apparent communication to the right coronary artery, in addition to cardiac chamber dilation and LVEF of 30% (*Fig. 1*). Subsequently, a diagnostic coronary angiography was performed to delineate the coronary anatomy further and confirm the collateral circulation from the left anterior descending artery (*Fig. 2*). The patient underwent cardiac surgery, including right coronary artery revascularization, closure of anomalous collateral vessels, mitral valve repair with a full semi-rigid ring, and left atrial appendage exclusion (*Fig. 3*). Given the complexity of the procedure, an intra-aortic balloon pump was placed intraoperatively. Postoperatively, the patient showed clinical improvement



**Figure 1:** **A)** Overview of coronary vessels course. **B)** Anatomical course of the anomalous right coronary artery. **C)** CCTA showing absent right coronary artery ostium and the proximal segment of the right coronary artery.



**Figure 2:** Coronary angiography delineating the coronary anatomy and confirm the collateral circulation from the left anterior descending artery.

in the intensive care unit and was discharged home in hemodynamically stable condition.

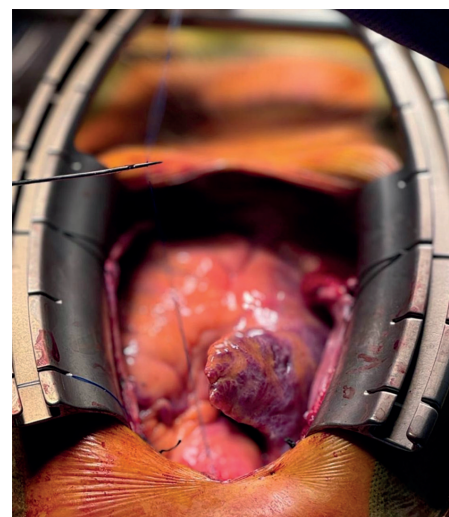
## COMMENTS

Anomalous origin of the right coronary artery is a rare congenital anomaly first described in 1948 by White and Edwards,<sup>3</sup> its prevalence is between 0.026 and 0.25% of the general population. An anomalous origin of the right coronary artery can lead to angina pectoris, myocardial infarction, or sudden death, in the absence of atherosclerosis.<sup>4</sup> The interarterial course of the anomalous RCA between the aorta and pulmonary artery predisposes it to mechanical compression, kinking, and stretching, particularly during exertion when diastolic expansion of the great vessels occurs, ischemia is primarily attributed to the intramural course rather than the ostial location.<sup>4</sup> The diagnosis of coronary artery anomalies relies on multiple imaging modalities, including echocardiography, cardiac magnetic resonance imaging, and CCTA. Among these, CCTA stands out as an excellent complementary technique for evaluating congenital coronary anomalies due to its superior spatial and temporal resolution. With a temporal resolution of just 66 ms, CCTA enables high-quality imaging even at elevated heart rates, surpassing some of the limitations of the cardiac magnetic resonance imaging. This modality provides high-resolution visualization of distal coronary vessels, allowing for detailed 3D reconstruction of the coronary anatomy and its relationship with extracardiac structures and the sternum. Additionally, it facilitates the detection of calcified or stenotic coronary segments while enabling the simultaneous assessment of ventricular function and associated cardiovascular abnormalities using a single imaging dataset.<sup>5,6</sup> Despite advancements in non-invasive imaging techniques, cardiac catheterization

remains a critical diagnostic modality. Its advantages lie in the ability to comprehensively evaluate the entire course and distribution of the coronary arteries. Selective angiography is particularly valuable in detecting small, discrete occlusions or interruptions of the coronary artery, where collateral circulation reconstitutes the vessel. Additionally, direct catheter-based imaging provides superior assessment of distal flow distribution and vessel patency, overcoming limitations associated with non-invasive methods. Intravascular ultrasound allows for direct visualization of coronary artery wall architecture, while fractional flow reserve enables quantitative evaluation of flow alterations across coronary stenoses.<sup>6</sup> Various surgical techniques have demonstrated favorable outcomes, with reimplantation emerging as an option that provides anatomic correction while avoiding long-term complications associated with coronary artery bypass grafting. Although unroofing is widely performed, studies have reported cases of persistent ischemia, suggesting the need for an individualized approach based on the patient's anatomical characteristics. Despite technical challenges, reimplantation has shown excellent mid-term outcomes by eliminating the intramural segment and associated stenotic disease, achieving anatomic correction while circumventing the late complications of conduit failure associated with bypass grafting, positioning itself as an effective strategy in the management of this condition.<sup>7</sup>

## CONCLUSIONS

This case highlights the varied presentation of coronary anomalies and emphasizes the importance of prompt multimodal imaging and personalized surgical management, including revascularization of the RCA and mitral valve repair, to optimize outcomes in complex anatomical settings.



**Figure 3:**

Coronary artery revascularization.

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**Funding:** none.

**Disclosure:** the authors have no conflict of interest to disclose.