

# Anomalous Origin of the Right Coronary Artery From the Left Anterior Descending Artery With Anomalous Course Between the Great Vessels: A Case for Conservative Management With Review of the Literature

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A single left coronary artery is a rare coronary anomaly. In such situations, the right coronary artery arises from the left anterior descending artery and traverses an unusual proximal course between the aorta and pulmonary trunk. There are only 10 such reported cases in the medical literature to date. After a detailed risk-to-benefit consideration, the decision was made for conservative management. In this report, we describe this rare case with a detailed review of the literature.

[Rev Cardiovasc Med. 2015;16(3):221-224 doi: 10.3909/ricm0774]

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## KEY WORDS

Anomalous coronary artery • Atherosclerosis • Coronary artery disease • Angina • Angiogram  
• Cardiac catheterization

**A** 55-year-old man presented with complaints of chest pain of 2 years' duration. The pain was brought on by exertion and relieved at rest. He had risk factors for coronary artery disease, including hypertension and hypercholesterolemia. He consumed approximately 2 to 3 glasses of vodka every night. He was given a stress test at another institution that was not indicative of inducible ischemia

and revealed normal chronotropic competence. His home medications included amlodipine and aspirin. Results of physical examination and laboratory investigation were unremarkable. His electrocardiogram revealed normal sinus rhythm at a rate of 80 beats/min, left axis deviation, left atrial abnormality, and T-wave inversion in lead V<sub>6</sub>. Due to exertional chest pain suggestive of angina, he underwent



Figure 1. Anomalous right coronary artery (RCA) originating from the left anterior descending artery and passing between the aorta and the pulmonary trunk. Note the narrowed segment of the RCA as it courses between the two major vessels.

cardiac catheterization, which revealed moderate left ventricular dysfunction with an ejection fraction of 30% to 35% and global hypokinesis. This was thought to be secondary to chronic alcoholism. Surprisingly, he also had an anomalous right coronary artery (RCA) originating from the left anterior

artery (Figure 1). The caliber of the vessel in this region appeared to be small but not significantly stenosed in comparison with a more normal distal caliber. Cardiothoracic surgery consultation was sought. Although single-vessel coronary artery bypass grafting was initially considered, the decision was later

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descending artery (LAD) with hemodynamically insignificant coronary artery disease. Cardiac computed tomography angiography was performed to identify the course and distribution of the RCA, which revealed a right dominant circulation with an anomalous RCA arising from the LAD, coursing between the aorta and pulmonary

made to manage the patient conservatively. Alcohol cessation was strongly advised. Standard heart failure medications, including  $\beta$ -blockers and angiotensin-converting enzyme inhibitors, were prescribed. There was no improvement in left ventricular ejection fraction on the follow-up echocardiogram. Implantable

cardioverter defibrillator placement was recommended but the patient refused. The patient has been stable without any cardiac events on follow-up visits.

## Discussion

Anomalous coronary arteries have varied clinical outcomes, depending on the anomaly found. The symptoms range from a totally asymptomatic status to sudden cardiac death (SCD). This has been a subject of multiple clinical and autopsy studies to date. International studies show a relatively consistent incidence of anomalous coronaries of less than 1.5%<sup>1-6</sup>; these studies cite the right coronary anomaly as the most common, with the exception of one study<sup>2</sup> in which the circumflex artery was noted to be the most common anomalous vessel. The incidence of an anomalous RCA is variable in different populations. The highest reported incidence has been in Indians (0.46%) and the lowest has been in Germans (0.04%).<sup>4-16</sup> Origins of anomalous RCA are variable and may arise from the contralateral sinus, descending thoracic aorta, left main coronary, left circumflex artery, above or from the left sinus of Valsalva, the pulmonary arteries, or even below the aortic valve.<sup>1,3,4,15-21</sup>

RCA anomalies can involve origin and/or course of the artery. When the RCA arises anomalously from the left coronary sinus,<sup>1-6</sup> it often follows a course between two great vessels and is particularly prone to compression. A single left coronary artery arising from the left coronary sinus is a rare anomaly, with the RCA arising from either the left main stem or the branches downstream from the main stem. Coming off the left main stem, the anomalously arising RCA can course anteriorly, posteriorly, or between the great

arteries. The last course suggests a malignant type of anomaly due to the risk of extrinsic compression. An RCA arising from the LAD in the absence of a normally situated right coronary ostium is considered a variant of a single left coronary

pulmonary artery and the aorta have received much attention because of their association with SCD.<sup>25-28</sup> Many patients with that type of abnormality are asymptomatic before the fatal event. Thus, when these patients are identified,

younger patients may also limit the available revascularization options, should they require a repeat bypass at a future date.

Noticeable differences are seen in the presentation and clinical course among patients with anomalous left coronary artery (ALCA) and anomalous right coronary artery (ARCA) abnormalities. The majority of deaths have occurred in ALCA patients, whether previously diagnosed or not.<sup>32</sup> This may justify surgical repair upon diagnosis in all ALCA patients. However, operative decisions regarding asymptomatic patients with ARCA must be evaluated subject to a risk-benefit analysis comparing the rate of occurrence of SCD and the number of asymptomatic patients who have the anomaly.<sup>33</sup> Considerations for operative risks related to ARCA surgery, as well as surgical outcomes, are important in the decision-making process.

The clinical outcome of ARCA is often benign. The incidence of SCD in asymptomatic patients with ARCA is extremely low. Over the past 25 years, clinical reports describe 10 patients with an RCA arising from the left coronary artery sinus who died suddenly. Eckart and colleagues<sup>34</sup> screened 6.3 million military recruits for SCD over a 25-year period for the Department of Defense Medical Mortality Registry. Basso and colleagues<sup>27</sup> reviewed two large registries in the United States and Italy,

*... anomalous coronary arteries originating from the contralateral aortic sinus coursing between the pulmonary artery and the aorta have received much attention because of their association with SCD.*

artery. Only 10 such cases have been described in the literature to date.<sup>7-13</sup> In most of these cases, the RCA arises from the proximal or mid LAD and courses to the right toward the right atrioventricular groove or the margin of the right ventricle, with resulting ischemic symptoms primarily due to stenotic disease in the LAD or, rarely, in the RCA.<sup>7-11</sup>

Most anomalies are asymptomatic and are seen as incidental findings on coronary angiography. Clinically symptomatic presentations include exertional angina, myocardial infarction, arrhythmias, and even SCD. The exact pathophysiologic basis of ischemia is unclear in cases of a single coronary artery without obstructive lesions arising from either the left or right sinus of Valsalva. It could be related to mechanical compression of an anomalous coronary artery between the aorta and pulmonary artery, particularly during exercise.<sup>1,21,22</sup> Another proposed mechanism is the presence of valve-like ridges and the acute angulation of the artery as it traverses from left to right.<sup>23</sup> Also, slit-like coronary orifice, acute angle of take-off of the anomalous artery from the aorta, and the presence and length of intramural segments of coronary vessel with proximal intussusception of the ectopic artery within the aortic root<sup>24</sup> are other possible causes. However, anomalous coronary arteries originating from the contralateral aortic sinus coursing between the

aggressive surgical management is often recommended. Nonetheless, decisions regarding the management of asymptomatic patients with these coronary anomalies are less well defined. Amid significant controversy regarding the treatment strategy, mainly due to a paucity of data, some authors have suggested surgical management for younger patients with anomalous coronaries arising in the opposite sinus, whether they are symptomatic or not.<sup>29,30</sup> The unpredictable nature of the clinical course, leading to ischemia and SCD among younger individuals with this kind of anomaly, is the primary reason for this recommendation. Various surgical options are available, including coronary bypass (with or without ligation of the anomalous vessel using an internal mammary [IM] or saphenous vein graft), coronary reimplantation, or unroofing procedures.<sup>31</sup>

Because the anomalous vessel may have a dynamic limitation of flow during exercise and decent

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flow at rest, there is a risk of graft closure due to competitive flow through the native vessel if it is bypassed. Although survival after bypass may be better when an IM graft is used, it is prone to atrophy due to competitive flow within the native vessel. Using an IM graft in

reporting SCD in young competitive athletes over a 10- to 20-year period; 27 deaths were recorded in patients who had no history of symptoms, 14 patients had an LCA arising from the right coronary sinus, and only 1 patient had an RCA arising from the left coronary sinus.

Brothers and associates<sup>35</sup> reported significant subclinical changes indicative of myocardial ischemia in postoperative patients after surgical unroofing procedures. In their report, 9 of the 24 patients met criteria for ischemia at a mean of 15 months after surgical repair. These included 8 of 16 patients with ARCA and only 1 of 10 with ALCA. Thus, based on this postsurgical report, a more conservative approach is probably ideal in asymptomatic patients with an ARCA. ■

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## MAIN POINTS

- A single left coronary artery is a rare coronary anomaly. In such situations, the right coronary artery (RCA) arises from the left anterior descending artery (LAD) and traverses an unusual proximal course between the aorta and pulmonary trunk. Most anomalies are asymptomatic and are seen as incidental findings on coronary angiography.
- Anomalous coronary arteries originating from the contralateral aortic sinus coursing between the pulmonary artery and the aorta have received much attention because of their association with sudden cardiac death. Many patients with that type of abnormality are asymptomatic before the fatal event. Thus, when these patients are identified, aggressive surgical management is often recommended.
- Various surgical options are available, including coronary bypass (with or without ligation of the anomalous vessel using an internal mammary or saphenous vein graft), coronary reimplantation, or unroofing procedures.
- Noticeable differences are seen in the presentation and clinical course among patients with anomalous left coronary artery (ALCA) and anomalous right coronary artery abnormalities; the majority of deaths have occurred in ALCA patients.