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CASE REPORT

INTERMEDIATE

CLINICAL CASE: CORONARIES

Dual Left Main Coronary Arteries With Ischemia and Coronary Steal

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ABSTRACT

A 59-year-old man had angina and an abnormal perfusion scan. Work-up revealed 2 left main coronary arteries: the anomalous artery originated from the right coronary cusp and took an aberrant interventricular septal course; the other artery was atretic. He underwent surgical unroofing, with resolution of symptoms. (**Level of Difficulty: Intermediate.**) (J Am Coll Cardiol Case Rep 2022;4:764-769) © 2022 The Authors. Published by Elsevier on behalf of the American College of Cardiology Foundation. This is an open access article under the CC BY-NC-ND license (http://creativecommons.org/licenses/by-nc-nd/4.0/).

HISTORY OF PRESENTATION

A 59-year-old White man was evaluated for exertional angina and dyspnea for 2 years and an abnormal exercise myocardial perfusion imaging (MPI) with single-photon emission computed tomography (SPECT). His vital signs and physical examination, including cardiovascular examination, were unremarkable. A resting electrocardiogram showed sinus rhythm. He was taking no medications.

LEARNING OBJECTIVES

- To understand the role of coronary anomalies as a cause of ischemic symptoms.
- To differentiate different types of coronary anomalies and their clinical implications.
- To recognize the importance and value of coronary CTA vs invasive angiography or stress testing in managing patients with coronary anomalies.

PAST MEDICAL HISTORY

He had hyperlipidemia and 40-pack-year history of cigarette smoking. He had no history of diabetes or hypertension.

DIFFERENTIAL DIAGNOSIS

The main differential diagnosis was obstructive coronary artery disease. Other less likely possibilities included microvascular angina, valvular heart disease, and hypertrophic cardiomyopathy. Infiltrative cardiomyopathy, pericardial disease, and cardiac masses were considered unlikely because of the lack of relevant history and signs.

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INVESTIGATIONS

Exercise MPI with SPECT revealed a medium-sized area of moderate-severity anterolateral and inferior wall ischemia and global moderate left ventricular hypokinesis with an ejection fraction (EF) of 36%.

An echocardiogram showed no valvular disease. On cardiac catheterization, the left main (LM) artery seemed to arise from the right sinus of Valsalva (SOV) (**Figures 1 and 2,** Videos 1 and 2). There was a small atretic second LM artery also that arose from the left SOV (**Figure 3,** Video 3). There was chronic total occlusion of the distal right coronary artery (RCA) with a patent posterior descending artery (PDA) and a posterolateral artery that filled from collateral vessels (left to right and right to right) (**Figure 4,** Video 4), which led to coronary steal confirmed by anterolateral ischemia on MPI.

Coronary computed tomography angiography (CTA) confirmed both an atretic LM artery with origin from the left coronary cusp and an anomalous LM artery supplying most of the left ventricular myocardium (**Figure 5,** Video 5). This anomalous LM artery was seen originating from the right SOV, first traversing the right ventricular outflow tract (RVOT) myocardium and then the interventricular septum (**Figure 6**), and it gave rise to a patent left anterior descending (LAD) coronary artery, 2 high diagonal arteries, and the left circumflex (LCx) coronary artery (**Figure 7**).

MANAGEMENT

Surgery was offered to the patient to relieve angina and potentially reduce the risk of sudden cardiac death resulting from the anomalous intraseptal course, and he agreed. At surgery, the anomalous LM artery and the RCA were both seen to arise from the right SOV but from separate ostia. The atretic LM artery that arose from the left SOV was left alone. A transconal unroofing of the anomalous LM artery was accomplished by following and exposing its course in the RVOT wall and interventricular septum. Crossing muscle fibers and adhesions were dissected off along this LM artery's entire course until it was fully freed up laterally to its bifurcation into the LAD and LCx arteries. A 1-vessel coronary artery bypass graft with a single saphenous vein graft (SVG) to the right PDA branch was then performed.

DISCUSSION

Coronary artery anomalies occur in 0.03% to 0.28% of the population.[1] The diagnosis of coronary artery anomalies can be challenging because many patients do not exhibit symptoms. Conversely, symptomatic patients may experience exertional chest pain, dizziness, shortness of breath, exercise intolerance, or even syncope.[1] Coronary anomalies may occasionally be incidental findings in patients undergoing work-up for ischemia, such as our patient.

The clinical significance of an anomaly depends on its origin, functional and anatomical features at the point of origin, course, and termination.[2] For example, 1 classification scheme designates the type of coronary artery anomaly on the basis of where the anomalous artery originates, such as the following: the origin of the LM artery from the right SOV; the origin of the RCA from the left SOV; the origin of the LAD artery from the right SOV or RCA; the origin

ABBREVIATIONS AND ACRONYMS

AAOC = anomalous aortic origin of a coronary artery

CMR = cardiac magnetic resonance

CTA = computed tomography angiography

EF = ejection fraction

LAD = left anterior descending

LCx = left circumflex

LM = left main

MPI = myocardial perfusion imaging

PDA = posterior descending artery

RCA = right coronary artery

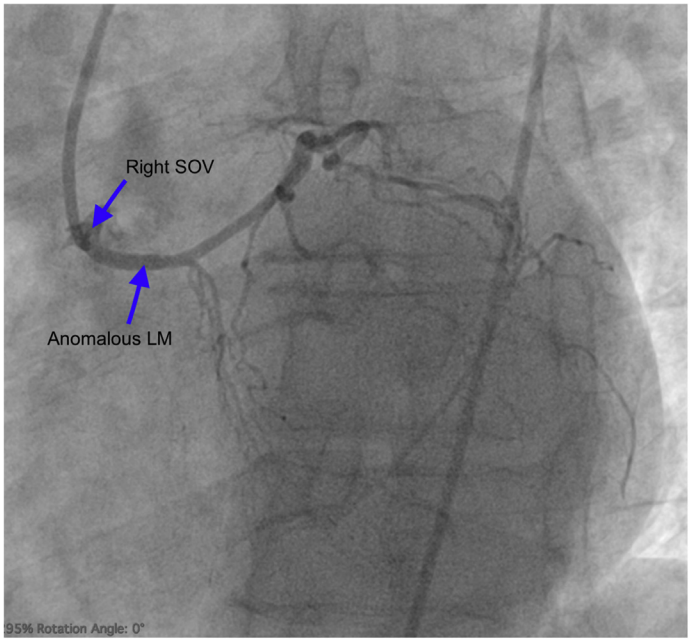
RVOT = right ventricular outflow tract

SOV = sinus of Valsalva

SPECT = single-photon emission computed tomography

SVG = saphenous vein graft

FIGURE 1 Invasive Angiogram of Left Coronary Artery (Left Anterior Oblique 37° and Caudal 5°)



LM = left main (artery); SOV = sinus of Valsalva.

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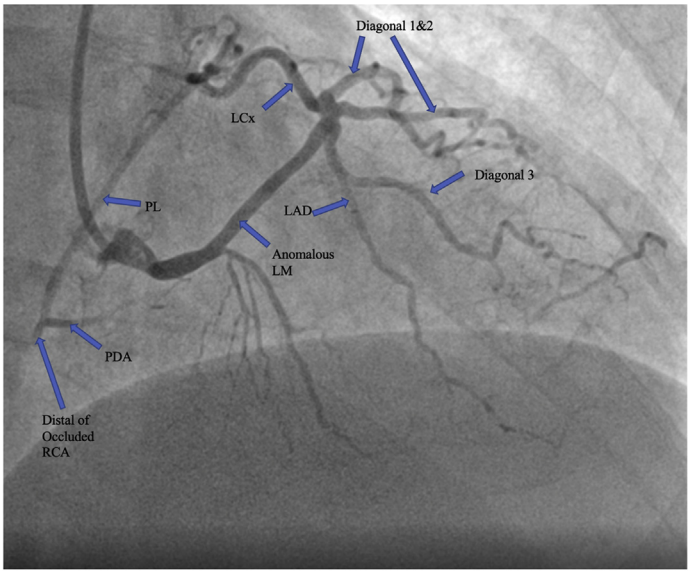
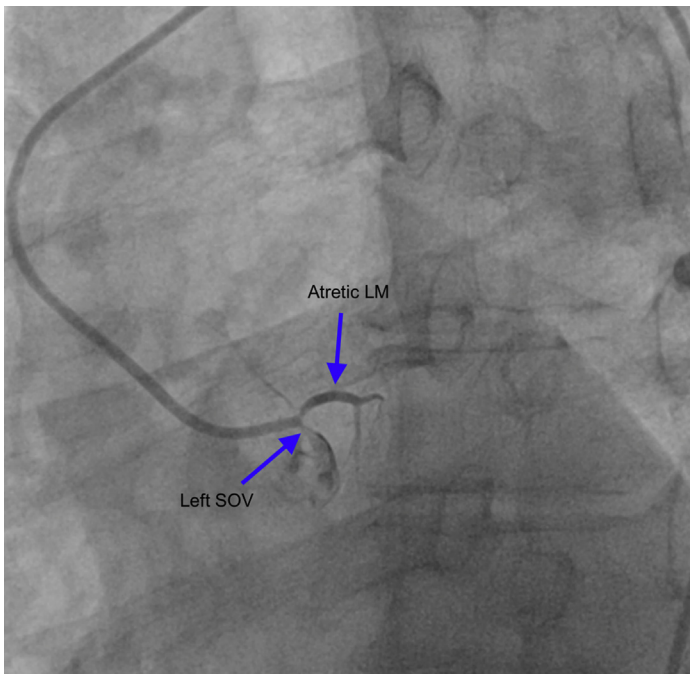
FIGURE 2 Invasive Angiogram of Left Coronary Artery (Right Anterior Oblique 22° and Cranial 25°)



LAD = left anterior descending (artery); LCx = left circumflex (artery); LM = left main (artery); PL = posterolateral (artery); PDA = posterior descending artery; RCA = right coronary artery.

FIGURE 3 Invasive Angiogram of Atretic LM Artery (Left Anterior Oblique 37° and Caudal 5°)



Abbreviations as in Figure 1.

of the LCx artery from the right SOV or RCA; a single coronary artery; and finally (very rarely), inverted coronary arteries, wherein both the left coronary artery and the RCA arise from the opposite SOV.[2]

Patients with anomalies where either the left coronary artery or the RCA originates from the opposite SOV have a higher risk of major adverse cardiac events and sudden cardiac death. Other anatomical features can also increase the likelihood of sudden death, including an intramural, interarterial course, a slitlike ostium, and acute angle take-off. Hence, surgical correction of an anomalous aortic origin of a coronary artery (AAOC) (from the opposite SOV) is a Class I recommendation when a patient has evidence of ischemia.[3]

The risk of sudden cardiac death in patients with an anomalous LM artery increases with exercise, especially in younger adults and with a "malignant" course (if the anomalous artery travels between the aorta and pulmonary artery,[1] possibly secondary to a scissor-like effect on the proximal course, or with a slitlike orifice that is seen in a majority of these cases, or an acute angle of take-off, or an ostial ridge that acts as a valvelike obstructive flap[4]). If these high-risk anatomical features are absent, the decision for surgical or medical management depends on a nuanced and detailed patient-physician discussion, especially given that stress testing does not reliably predict the risk of sudden death, and even patients without ischemia (by stress testing or symptoms) may experience adverse events.

Multiple surgical options exist. Our patient underwent unroofing of the anomalous coronary and an SVG (provided a new conduit for blood flow to the distal RCA), ameliorating a left-to-right steal. Other surgical options include ostial splitting, sphincteroplasty, reimplantation, and patch augmentation. Unroofing is preferred because it is associated with lower morbidity and mortality (**Central Illustration**).[1]

In our patient, the anomalous LM artery took a septal course. This was previously believed to have minimal risk of sudden cardiac death compared with the more "malignant" interarterial course.[5] However, newer evidence has challenged this idea: a higher rate of adverse cardiac events including sudden death has been reported in these patients.[5,6] According to Torres et al,[7] trying to identify subtypes with conventional coronary angiography is "too simplistic and does not capture the entire anatomical spectrum detected by CT [computed tomography]."[8] Coronary CTA can detect which sinus the anomaly originates from, the exact location on the aortic wall, the take-off angle, the initial or distal vessel diameter, any

fibrous hyperplasia or stenosis, plaque or outward remodeling, abnormal collateral connections, the exact course (whether in the epicardial fat or intramural in the pulmonary artery or RVOT or the aorta), and the potential for compression as it traverses in the aortopulmonary window of fat. Coronary CTA has the advantage of rapid scan time and excellent spatial resolution but exposes the patient to radiation and iodinated contrast material, a limitation that is particularly relevant in younger patients or those with renal insufficiency. Cardiac magnetic resonance (CMR) (including a noncontrast version of coronary CMR angiography) may be an alternative if radiation and contrast exposure are concerns and also gives additional information about scar, ischemia, and cardiac function. Its limitations include suboptimal visualization of the distal coronary arteries and lower spatial resolution (1.5 mm vs 0.4 mm for coronary CTA), longer scan times, higher costs, and less availability.[1] The guidelines give both coronary CTA and coronary CMR angiography a Fremont, I indication to image AAOC.[1]

FOLLOW-UP

Our patient was discharged 4 days postoperatively. Echocardiography at 6 weeks showed an improved EF of 50%. Two months into follow-up, he was walking briskly without any symptoms.

CONCLUSIONS

The diagnosis of anomalous coronary arteries is usually suspected on cardiac catheterization and can be confirmed by coronary CTA. Whether a coronary anomaly is benign depends on its course and other anatomical features that are better delineated by coronary CTA.

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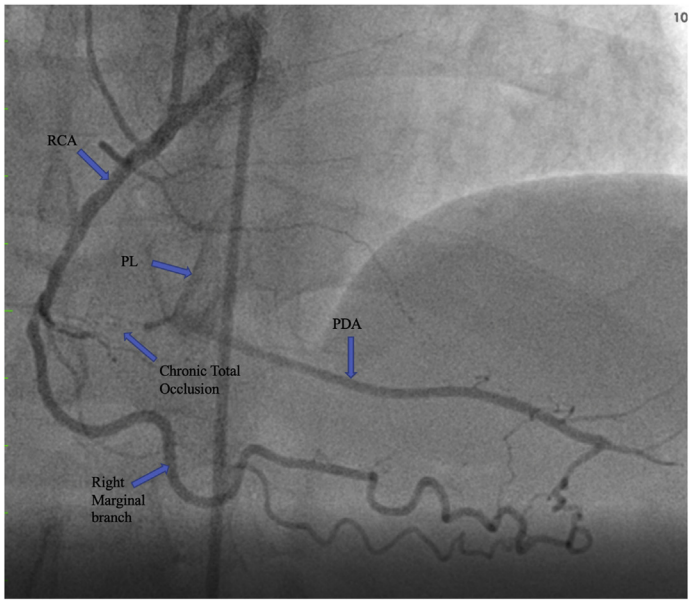
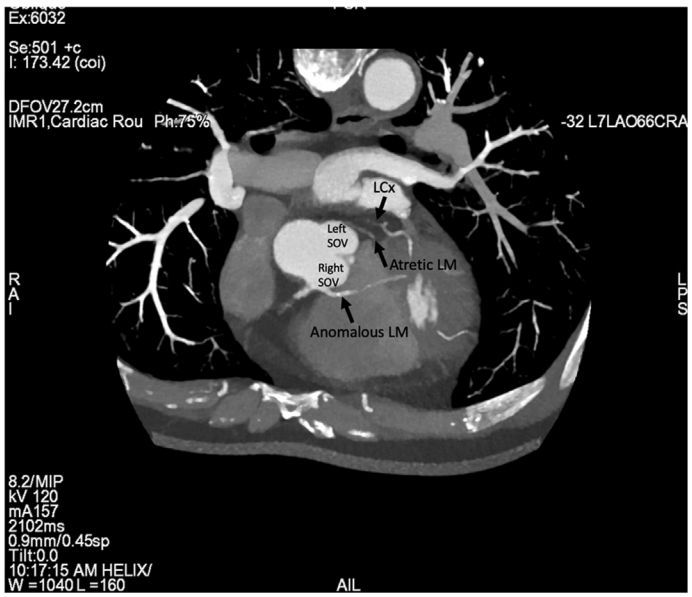
FIGURE 4 Invasive Angiogram of RCA (Right Anterior Oblique 1°/Cranial 30°)



Abbreviations as in Figure 2.

FIGURE 5 Coronary Computed Tomography Angiography Showing Both LM Arteries



Abbreviations as in Figures 1 and 2.

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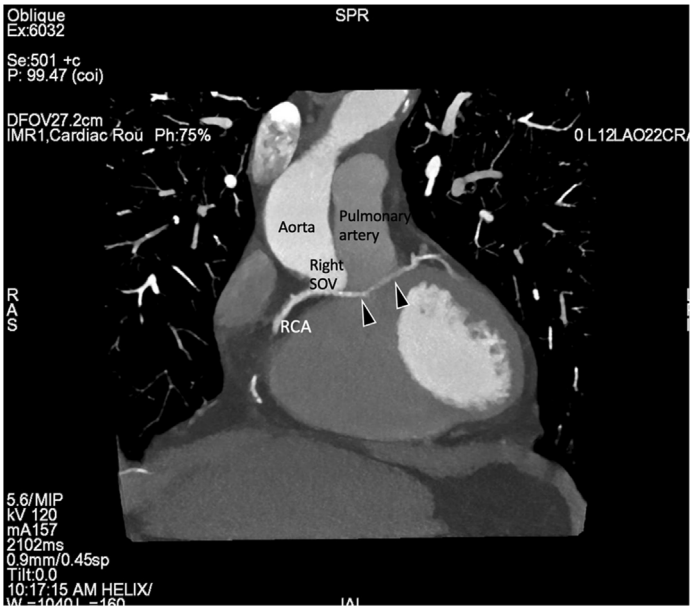
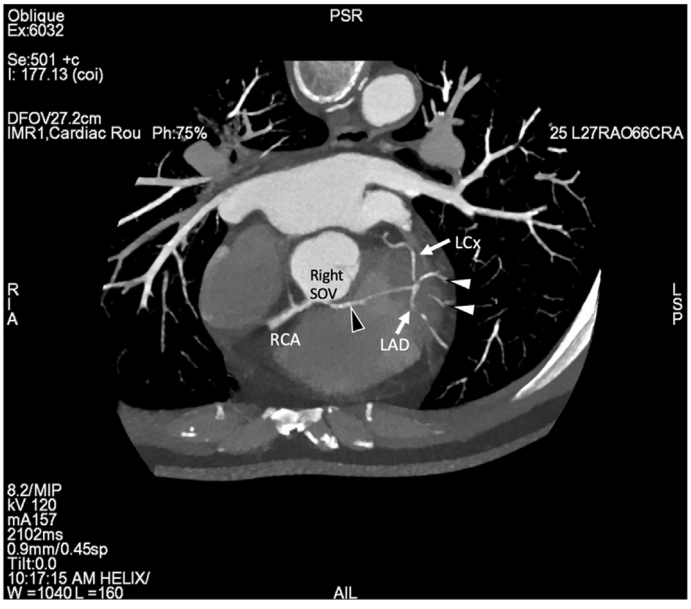
FIGURE 6 Coronary Computed Tomography Angiography of Anomalous LM Artery



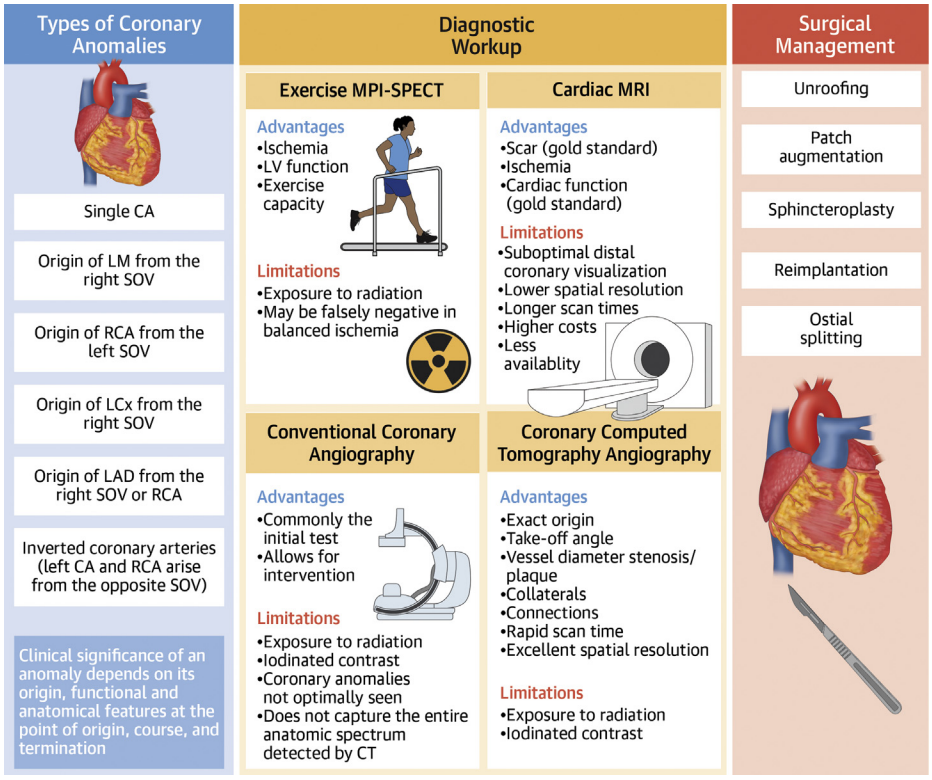
The image shows the anomalous left main (LM) artery **(arrowheads)** with an interventricular or intraseptal course. Abbreviations as in Figures 1, 2, and 5.

FIGURE 7 Coronary Computed Tomography Angiography of RCA and Anomalous LM Artery



Computed tomography angiography of the RCA and an anomalous LM artery **(black arrowhead)** connected to the LAD artery, 2 diagonal arteries **(white arrowheads),** and the LCx artery. SOV = sinus of Valsalva; other abbreviations as in Figure 2.

CENTRAL ILLUSTRATION **Approach to a Patient With an Anomalous Coronary Artery**



Zheng S, et al. J Am Coll Cardiol Case Rep. 2022;4(13):764–769.

CA = coronary anomaly; CT = computed tomography; LAD = left anterior descending; LCx = left circumflex; LM = left main; LV = left ventricle; MRI = magnetic resonance imaging; RCA = right coronary artery; SOV = sinus of Valsalva.

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KEY WORDS computed tomography, coronary angiography, coronary vessel anomaly, myocardial ischemia

APPENDIX For supplemental videos, please see the online version of this article.