INTERVENTIONAL CARDIOLOGY PERSPECTIVES

Inter Cardio Per

DOI: 10.4274/intercardiopers.2025.2025-10-1

CASE REPORT

Acute Coronary Syndromes in Patients with Coronary Artery Anomalies: Clinical Challenges and Management Strategies

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ABSTRACT

With the increasing use of coronary angiography, the recognition of congenital coronary artery anomalies (CAAs) has risen, although their overall prevalence remains approximately 1%. Acute coronary syndrome caused by an anomalous coronary artery arising from the opposite sinus and identified as the culprit vessel is extremely rare. Such anatomical variations may contribute to delays in coronary reperfusion in certain presentations. Here, we describe two patients with comparable CAAs who presented with acute myocardial infarction and were successfully managed using different treatment approaches. These cases highlight the importance of individualized evaluation and tailoring therapeutic strategies to each patient's specific anatomical features.

Keywords: Acute coronary syndrome, coronary angiography, coronary artery anomaly

INTRODUCTION

Coronary artery anomalies (CAAs) are congenital abnormalities, most of which remain asymptomatic throughout life. With the increasing use of coronary angiography and multi-slice cardiac computed tomography, CAAs are now more frequently identified as incidental findings. Large invasive angiographic studies report a prevalence ranging from 0.78% to 1.3%.[1,2] The most common anomaly is the widely separated origin of the left anterior descending (LAD) and left circumflex (LCx) arteries from the left sinus of Valsalva. Acute myocardial infarction caused by an anomalously originating coronary artery identified as the culprit vessel is extremely rare. In patients presenting with acute coronary syndrome (ACS), certain anomalous coronary anatomies may delay coronary reperfusion and complicate management. Here, we describe two patients with similar CAAs who presented with ACS and were successfully treated using different therapeutic strategies.

CASE REPORT

A 76-year-old man presented to the emergency department with chest pain. Physical examination revealed bilateral crackles on lung auscultation and hypotension (blood pressure, 80/50 mmHg). Resting electrocardiography showed sinus rhythm with negative T waves in the inferior leads. Transthoracic echocardiography (TTE) demonstrated regional hypokinesia of the posterior left ventricular wall. High-sensitivity troponin I was markedly elevated (>50,000 pg/mL; normal <36 pg/mL), supporting the need for an early invasive strategy. The patient was classified as Killip Class II–III.

A urinary catheter was inserted, and inotropic support with norepinephrine was initiated. Intravenous furosemide (20 mg) was administered to relieve pulmonary edema. The patient also received 300 mg of chewable acetylsalicylic acid and 5,000 units of intravenous heparin before coronary angiography. After partial symptomatic improvement, he was transferred immediately to the catheterization laboratory.

Coronary angiography revealed atherosclerotic plaques in a type 1 dual LAD artery, while the LCx artery was not visualized within the left coronary system (Figure 1A). The right coronary artery (RCA) was diminutive, but a critical proximal LCx lesion was identified; the LCx originated from a separate ostium adjacent to the right aortic sinus (Figure 1B). As expected, the anomalous LCx coursed posterior to the aorta before reaching its usual distribution.

Given the patient's hemodynamic instability and the presence of a single critical stenosis in the anomalous LCx, percutaneous coronary intervention (PCI) was performed. The LCx was selectively cannulated using a 6 Fr Amplatz Right (AR-2) guiding catheter. A PT2 hydrophilic moderate-support guidewire successfully crossed the lesion, followed by balloon angioplasty with a 2.0×20 mm semi-compliant balloon. A

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Cite as: Demir M, Özcan AV. Acute coronary syndromes in patients with coronary artery anomalies: clinical challenges and management strategies. *Inter Cardio Pers*. [Epub Ahead of Print]

Received: 06.10.2025
Accepted: 10.12.2025
Epub: 16.12.2025

3.0×24 mm drug-eluting stent was then deployed in the proximal LCx (Figure 1C). The procedure was completed without complications.

The patient remained stable on medical therapy and was discharged without adverse cardiac events. Informed consent was obtained.

A 71-year-old man presented to the emergency clinic with chest discomfort. His medical history revealed type 2 diabetes mellitus diagnosed 10 years earlier. Physical examination showed bilateral crackles on lung auscultation, and his blood pressure was 140/85 mmHg. Electrocardiography demonstrated sinus rhythm with ST-segment depression in leads V4–V6. TTE revealed regional hypokinesia involving the apical and posterior segments of the left ventricle. High-sensitivity troponin I was markedly elevated (>439 pg/mL).

Coronary angiography showed severe stenosis in the mid-LAD artery, the proximal first diagonal branch, and a significant bifurcation lesion at the LAD–diagonal branch junction (Medina 0,1,1) (Figure 2A). The LCx artery was not visualized in the left coronary system. Right coronary angiography revealed critical stenosis distal to the right ventricular branch of the RCA. Notably, the LCx originated anomalously from the proximal RCA and exhibited severe ostial stenosis (Figure 2B). The prevalence of this coronary anomaly is approximately 0.37% in published reports.[1,2]

After reviewing the angiographic findings, the patient's SYNTAX I and SYNTAX II scores were calculated as 20 and 34, respectively. Considering the complexity of the LAD–diagonal bifurcation lesion, the presence of a CAA with significant ostial and proximal LCx disease, three-vessel coronary artery disease, and comorbid type 2 diabetes mellitus, the multidisciplinary heart team concluded that coronary artery bypass grafting (CABG) would provide the most durable and appropriate treatment strategy. The patient underwent successful CABG surgery and was discharged uneventfully one week later. Informed consent was obtained.

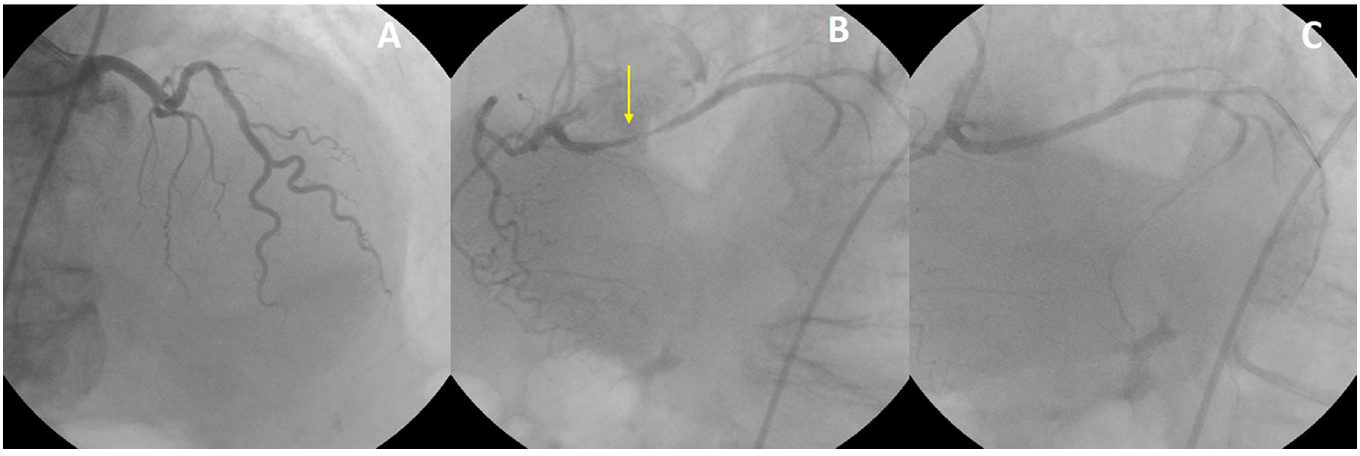

Figure 1. (A) Atherosclerotic plaques on the mid left anterior descending segment anteroposterior cranial view. (B) Significant lesions on the proximal left circumflex (LCx) (yellow arrow) that arise from a separate ostium in the left cranial view. (C) The imaging of LCx after successful revascularization

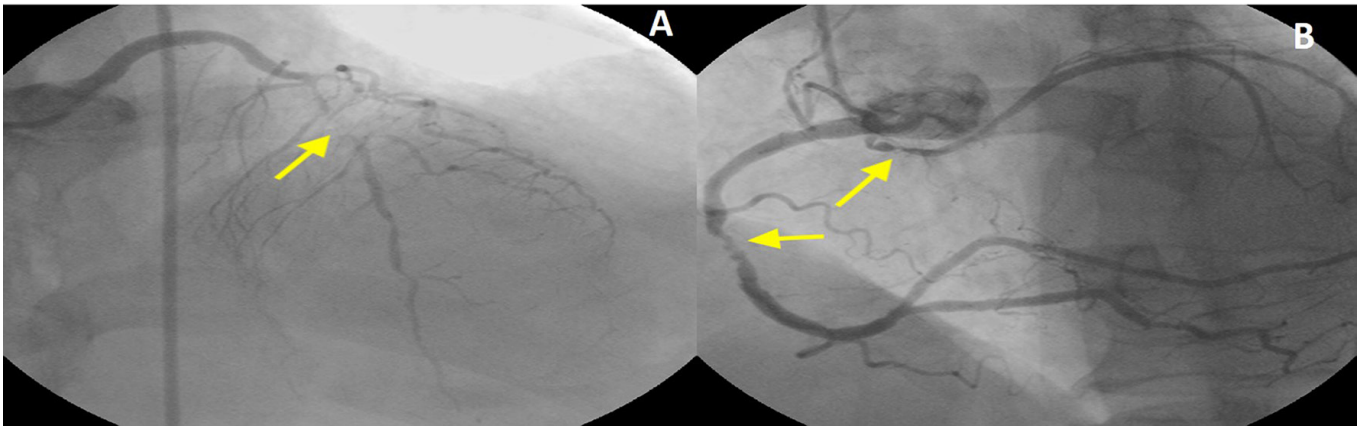

Figure 2. (A) Coronary angiography showed severe stenosis in the middle segment of the left anterior descending (LAD) (yellow arrow) and the proximal segment of the first diagonal branch, as well as severe stenosis at the LAD-diagonal bifurcation (Medina 0,1,1) lesion on the anteroposterior cranial view. (B) Right coronary angiography revealed that the left circumflex (LCx) arises from the proximal part of the right coronary artery (RCA) and shows critical stenosis at the LCx ostium, additionally severe stenosis mid-segment of the RCA (yellow arrows)

DISCUSSION

CAAs represent a heterogeneous group of congenital abnormalities with diverse clinical presentations. Although most CAAs are asymptomatic and detected incidentally, certain forms may lead to myocardial ischemia, malignant arrhythmias, or sudden cardiac death. The anomalous origin of a coronary artery from the opposite sinus is rare and occurs in approximately 1% of the population.[1,2] CAAs are clinically significant because they constitute the second most common cardiovascular cause of sudden death in young individuals.[3]

Under normal circumstances, the LCx travels through the left atrioventricular groove, supplying the lateral wall of the left ventricle. The most frequent coronary anomaly involving this vessel is the separate origin of the LAD artery and LCx from the left sinus of Valsalva. The anomalous origin of the LCx from the opposite sinus was first described by Antopol and Kugel in 1933.[4] The LCx and RCA may arise from a shared ostium or from separate ostia. In most cases, the anomalous LCx courses posterior to the aorta before reaching its typical distribution, with a reported prevalence of 0.32–0.67%.[5]

The relationship between coronary anomalies and the development of atherosclerosis remains controversial. Although the prevailing view is that anomalous arterial segments are not inherently more vulnerable to atherosclerotic disease than normally positioned segments, some studies suggest the opposite. In particular, a higher incidence of stenosis has been reported in LCx arteries arising from the opposite sinus compared with normally originating LCx arteries.[6,7] The variant in which the LCx arises anomalously from the right coronary sinus and courses retroaortically is considered the most common benign coronary anomaly. Although generally not hemodynamically significant, it may complicate aortic valve replacement surgery, posing a risk of iatrogenic LCx occlusion due to inadvertent suture placement. With the advent of transcatheter aortic valve implantation in the last decade, there is also a potential risk of compressing the anomalous LCx with the bioprosthetic valve.

The optimal management of CAAs remains controversial. Current guidelines recommend surgical intervention for patients with an anomalous aortic origin of a coronary artery who exhibit symptoms of myocardial ischemia or demonstrate inducible ischemia on exercise testing.[8,9] Children with an interarterial ("malignant") course are often referred for surgical correction even if asymptomatic, due to the markedly increased risk of sudden cardiac death, particularly during exertion. In contrast, the management of asymptomatic patients with an anomalous RCA is less well-defined, as provocative testing is frequently negative, and treatment decisions must be individualized.[8,9]

Symptomatic patients with CAAs have three main therapeutic options: medical therapy (including beta-blockers and exercise restriction), PCI, or surgical correction. Among surgical approaches, the "unroofing" procedure is often considered the most straightforward both technically and conceptually.

PCI for atherosclerotic lesions in anomalous coronary arteries has been rarely reported in the literature, and long-term outcomes remain limited.[10] The atypical location and non-circular orifice of these vessels make selective catheterization and PCI technically challenging, often due to insufficient guiding catheter support. Guidelines and practical recommendations, such as "Catheter Selection and Angiographic Views for Anomalous Coronary Arteries: A Practical Guide,"[11] suggest that an anomalous LCx arising from the right cusp near the RCA ostium, or toward the left cusp, is best engaged using an Amplatz left or right catheter.

In our first case, this approach was successfully applied using an AR-2 guiding catheter, achieving revascularization without complications. In the second case, an AR-1 catheter was used in the right view to visualize the LCx arising from the proximal RCA with critical ostial stenosis. Considering the patient's multivessel disease, SYNTAX scores, and type 2 diabetes mellitus, CABG was deemed the more appropriate treatment.

CONCLUSION

CAAs pose significant technical challenges in the cardiac catheterization laboratory. Difficulty in locating and engaging the coronary ostium may necessitate higher contrast volumes and result in prolonged radiation exposure. In patients with ACS, these anatomical variations can delay timely intervention if the culprit anomalous artery cannot be quickly engaged. Interventional cardiologists should be familiar with CAAs and their management to minimize procedural time and radiation exposure. Likewise, cardiac surgeons must exercise caution to avoid inadvertent cross-clamping or transection of anomalous arteries during surgery.

Informed Consent: Informed consent was obtained.

Authorship Contributions: Concept: M.D., Design: M.D., Data Collection or Processing: A.V.Ö., Literature Search: M.D., Writing: M.D., A.V.Ö.

Conflict of Interest: No conflict of interest was declared by the authors.

Financial Disclosure: The authors declared that this study received no financial support.

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