



# A unique case of non-ST-elevation myocardial infarction with abnormal origin of left coronary system from the right coronary cusp

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## Abstract

A unique case of non-ST-elevation myocardial infarction is discussed, in which the left main coronary artery and anomalous coronary artery from the opposite sinus of Valsalva were absent. In this case, the left coronary cusp was blunted, and all three coronary arteries trifurcated from a single ostium in the right coronary cusp. The proximal part of the left anterior descending coronary artery had a trans-septal (intermuscular) course, while the left circumflex coronary artery had a retro-aortic course and severe thrombotic stenosis before the terminal portion. Due to the patient's refusal of coronary artery bypass graft, percutaneous coronary intervention was performed.

## Keywords

Anomalous coronary artery from the opposite sinus of Valsalva, non-ST-elevation myocardial infarction, coronary angiography, coronary computed tomography angiography

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## Introduction

The prevalence of anomalous coronary artery from the opposite sinus of Valsalva (ACAOS) in the general population is low; however, the absolute number of detected cases is increasing due to the more frequent use of invasive and non-invasive imaging techniques to rule out coronary artery disease (CAD). It has been suggested that people diagnosed with ACAOS and an interarterial course (IAC) may be at risk of myocardial ischemia and subsequent arrhythmia, even in the absence of atherosclerotic lesions, as the anomalous vessel is prone to dynamic compression during physical exercise.<sup>1</sup> ACAOS is characterized by an anomalous course of a native coronary artery, which does not originate from its own sinus of Valsalva, but instead from the opposite one. It can be classified into malignant or benign variant. Autopsy studies have shown that malignant variants of ACAOS can be an underlying cause of sudden cardiac death (SCD) in young athletes. ACAOS with an IAC has been identified as a potential

underlying cause of SCD<sup>2</sup>; however, older people diagnosed with ACAOS seem to be less susceptible to adverse cardiac events. Malignant ACAOS has an IAC of the anomalous vessel between the pulmonary artery and the aorta, whereas benign variants represent all other courses, such as in front of the pulmonary artery (prepulmonic) or a retro-aortic course.<sup>3</sup>

A study conducted by the University of Bern in Switzerland evaluated middle-aged and older patients with suspected CAD who underwent coronary computed tomography angiography (CCTA). Of the 5634 patients evaluated, 66 were

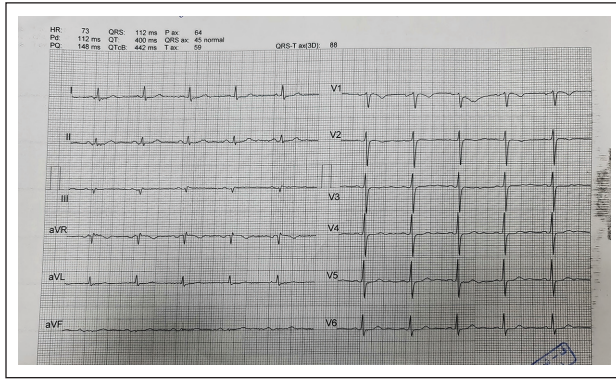
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**Figure 1.** The patient's ECG on admission showed flattening of the T-wave.  
ECG: electrocardiogram.

diagnosed with ACAOS, of which 36 had an IAC, resulting in a prevalence of 0.64%.<sup>3</sup> Other investigations have also shown a prevalence of less than 1%.<sup>4,5</sup> Studies in the US and Europe have shown that young athletes engaging in vigorous physical exercise may be at an increased risk of SCD if they have been diagnosed with ACAOS with an IAC.<sup>6,7</sup>

## Case presentation

A 67-year-old non-smoker man with no past medical history presented to our hospital with anginal chest pain, diaphoresis, nausea, vomiting, and dyspnea on exertion 2 h prior to admission. The patient denied taking any medications. On physical examination, his blood pressure was 130/80 mmHg, heart rate 80 beats per min, respiratory rate 16 breaths per min, oxygen saturation 98%, and temperature 36.5°C. His jugular veins were flat and lungs were clear. Heart examination revealed S1 S2 and S3 S4 gallop, as well as a systolic murmur of 2/6 heard at the apex of the heart. Abdominal examination revealed no tenderness or organomegaly, and pulses in the extremities were symmetrical with no edema.

An electrocardiogram (ECG) showed a normal sinus rhythm and normal axis, with flattening of the T-wave in precordial leads (Figure 1). Except for an elevated cardiac troponin I, all other laboratory data including CBC, BUN, Cr, Na, K, FBS, and lipid profile were normal. Transthoracic echocardiography (TTE) showed normal left ventricular systolic function, with a left ventricular ejection fraction (LVEF) of 55%, and no regional wall motion abnormality (RWMA). Additionally, normal right ventricular size and systolic function as well as mild mitral regurgitation were seen. The aorta had normal dimensions with no evidence of aortic dissection, and there was no significant pericardial effusion.

Due to the positive results of high-sensitive cardiac troponin I (35 ng/ml at the first hour and 40 ng/ml at the fourth hour, with a cut-off of >0.9 ng/ml), the patient was indicated for coronary angiography (CAG) with the impression of

non-ST-elevation myocardial infarction (NSTEMI). CAG was done on the same day (day 0 after referral) and showed that the left anterior descending (LAD) and left circumflex (LCX) originated from the right cusp, and LCX had a severe thrombotic lesion in the distal part before the terminal portion with an acceptable runoff, which was determined to be the culprit lesion (Figure 2).

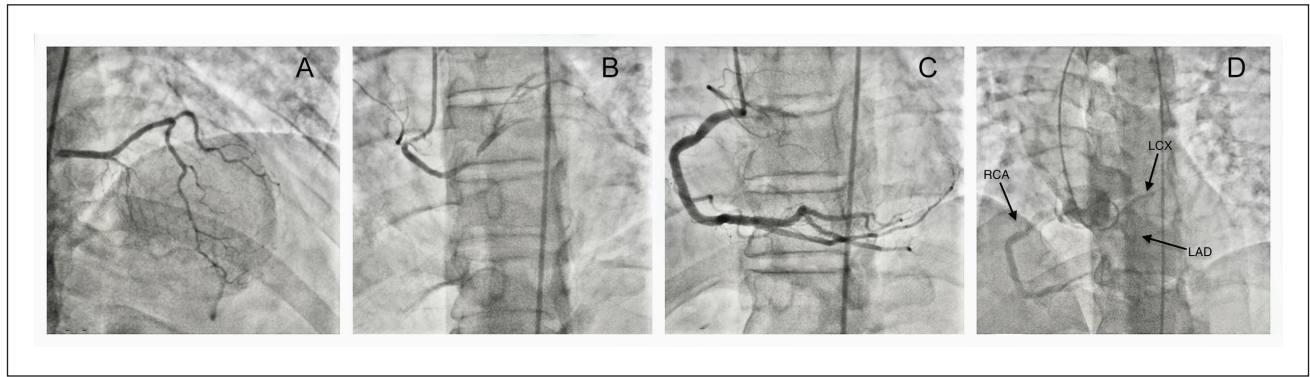
On the day 1 after referral, a CCTA was performed, which revealed a single coronary ostium from the right coronary cusp trifurcating into the LAD, right coronary artery, and LCX. Additionally, the proximal part of the LAD had a transseptal (intermuscular) course and the LCX had a retro-aortic course (Figure 3).

To determine the best revascularization option (coronary artery bypass graft (CABG) or percutaneous coronary intervention (PCI)) for this patient, the Heart Team (the joint team of cardiologists and cardiac surgeons) considered the composite of findings and decided that CABG was the most feasible option according to current guidelines. However, the patient refused CABG, so the Heart Team opted for PCI on LCX as a secondary choice. The PCI was performed on day 3 using a Judkins right 6 mm × 4 mm guiding catheter. Sion Blue wiring was done, followed by predilation with a SC balloon 2 mm × 20 mm and stent Xience 2.75 mm × 23 mm, and postdilation with a NC balloon 3 mm × 15 mm. The patient was followed up every 3 months for a year, with history, physical examination, ECG, and TTE, with no signs or symptoms. Follow up TTEs showed that the patient's left ventricle (LV) systolic function remained normal with an LVEF of 55%, without diastolic dysfunction or RWMA.

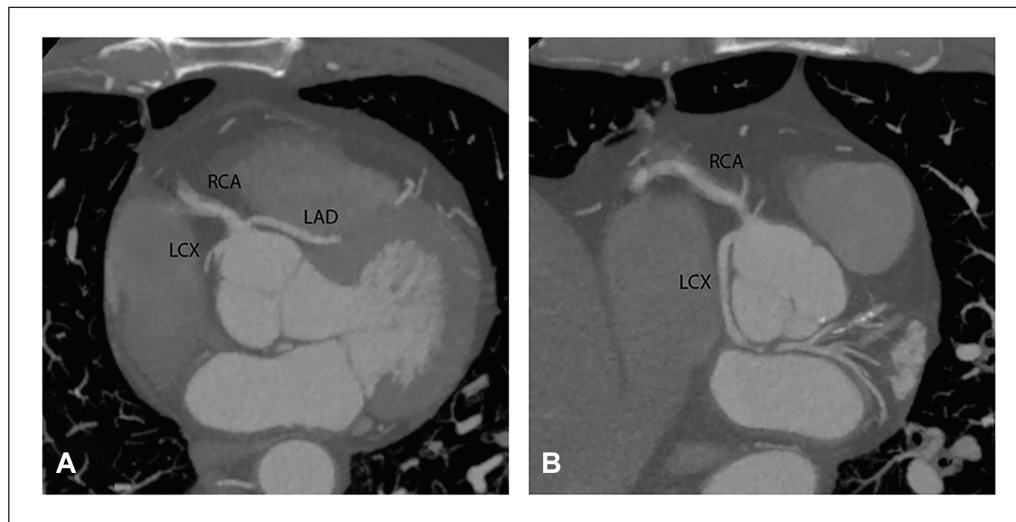
## Discussion

Coronary artery anomalies (CAAs) are rare congenital defects, which can present with a variety of conditions, ranging from asymptomatic to sudden cardiac arrest. Anomalous origin of coronary arteries is a type of CAA,<sup>8</sup> in which one or more of the main branches of the coronary arteries arise from an ectopic origin. These coronary arteries can arise from the wrong sinus of Valsalva (e.g., the LCX artery arising from the right coronary sinus) or from a different structure, such as the pulmonary artery, a branch of another coronary artery, or even a ventricular chamber. The left system that arises from the right aortic sinus typically follows one of four courses: prepulmonic, retroaortic, intraarterial, or transseptal. The IAC of an anomalous left coronary system from the right sinus is associated with SCD during or shortly after exercise in young individuals. Once this anomaly is diagnosed, CABG is recommended, although a stenting strategy has also been reported.<sup>9</sup>

Various imaging techniques can be used to diagnose CAAs, including CCTA, cardiac magnetic resonance, CAG, and echocardiography. As CCTA can provide a better three-dimensional image without the need for an invasive procedure, it is recommended as the gold standard for diagnosis



**Figure 2.** Multiple coronary angiographic views of the patient. (A) LAD from the right coronary cusp (RCC), (B) LCX from the right coronary cusp had a severe thrombotic stenosis in the mid portion, (C) right coronary artery (RCA) from the right coronary cusp, (D) an aortogram showed all three coronary arteries originating from the RCC. LAD: left anterior descending; LCX: left circumflex.



**Figure 3.** CCTA of the coronary arteries, which confirmed the diagnosis of ACAOS. (A) A multiplanar reconstruction of CCTA showed a single coronary ostium from the RCC trifurcating into the LAD, RCA, and LCX, with the intramuscular (trans-septal) course of the LAD and the retro-aortic course of the LCX. (B) A multiplanar reconstruction of CCTA also showed a short axis plane of the aortic root and the retro-aortic positioning of the LCX.

ACAOS: anomalous coronary artery from the opposite sinus of Valsalva; CCTA: coronary computed tomography angiography; LAD: left anterior descending; LCX: left circumflex; RCA: right coronary artery; RCC: right coronary cusp.

over CAG. Nowadays, more patients with ACAOS are detected incidentally due to the increasing use of these imaging techniques.<sup>8,10,11</sup> The 2018 American Heart Association/American College of Cardiology guideline and the 2020 European Society of Cardiology guideline both recommend that in symptomatic patients with ACAOS, exercise should be restricted and surgery should be considered as the treatment of choice. However, in this case, the patient refused surgery and revascularization using a catheter was done instead. Although further evaluation with more data is necessary, this case may help physicians in the management of patients with similar characteristics, especially those over 65, using PCI.<sup>10,11</sup>

It is believed that CAAs of abnormal origin may play an important role in sudden death in young athlete patients; however, it is still unclear if this anomaly could be dangerous in non-athlete elders. According to Opolski et al.,<sup>12</sup> there was no difference in the prevalence of significant CAD between patients with and without ACAOS as detected by CCTA (29% vs 34%,  $p=0.385$ ). In another study conducted by Gräni et al.,<sup>13</sup> 66 middle-aged patients with ACAOS diagnosed by CCTA were compared to a control group. Based on this study, mid-term outcomes for major adverse cardiac events were not statistically different (hazard ratio=0.94,  $p=0.89$ ). These findings raise the question of whether it is necessary to repair ACAOS by surgery in non-athlete elderly patients.



To summarize, although improvements in imaging have made the diagnosis of ACAOS easier, there is still a big question for which we have no specific answer: what is the treatment of choice for different age groups?

## Conclusion

In this report, a case of NSTEMI and its management in a patient diagnosed with ACAOS were discussed. ACAOS is a rare anatomical condition of coronary arteries, which can be easily missed without performing CAG or CCTA; therefore, accurate evaluation is crucial in order to manage affected patients, determining their sport activity, invasive treatment versus surgical correction, and medical treatment. According to the current guidelines, the best approach to treat severe coronary artery stenosis in conjunction with ACAOS would be CABG. Nevertheless, due to the patient's rejection of the treatment of choice, and single CAD of the LCX, a joint team decided that PCI would be an option, which was performed successfully. Consequently, more studies on PCI in ACAOS cases are needed to substantially determine the outcome of this procedure.

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## Ethics approval

Our institution does not require ethical approval for reporting individual anonymized case report or series.

## Patient permission/consent statement

Written Informed Consent was obtained from the patient to include case details or other personal information in this case report.

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## Supplemental material

Supplemental material for this article is available online.

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