# Percutaneous Coronary Intervention For Treatment of Unusual Origin of the Left Main Artery: A Case Report

Authors:	<ul> <li>*Tamer Elkhayat,<sup>1</sup> Yassir Birema,<sup>1</sup> Adel Elhoseiny<sup>1,2</sup></li> <li>1. Cardiac Services Department, King Salman Armed Forces Hospital Northwestern Region, Saudi Arabia</li> <li>2. East Surrey Hospital, Redhill, UK</li> <li>*Correspondence to dr.tamer.el.khayat@gmail.com</li> </ul>
Disclosure:	The authors declare no conflicts of interest.
Received:	17.05.23
Accepted:	19.10.23
Keywords:	Cardiac CT angiography (CCTA), chest pain, coronary artery disease, congenital coronary anomalies, percutaneous coronary intervention (PCI).
Citation:	EMJ Int Cardiol. 2023; DOI/10.33590/emjintcardiol/10301047. https://doi.org/10.33590/emjintcardiol/10301047.

#### Abstract

The left main coronary artery originating from the right sinus of Valsalva is a rare congenital anomaly. A 62-year-old male came in with recurring exertional chest discomfort, as observed by the authors. He was referred to the catheterisation laboratory for coronary angiography, which revealed the presence of a left main coronary artery coming from the right coronary sinus. In the proximal portion of the left anterior ascending coronary artery, a critical stenotic lesion was identified. The cardiac CT angiography demonstrated a benign retroaortic course. The lesion was effectively treated utilising three drug-eluting stents.

#### **Key Points**

1. A 62-year-old male presented with recurring exertional chest discomfort. Coronary angiography confirmed the existence of a left main coronary artery originating from the right coronary sinus.

2. The report details the patient's clinical presentation, diagnostic workup, treatment strategy, and follow-up. It also emphasises the significance of early detection and therapy of this condition to avoid negative results.

3. Congenital anomalies of the origin and/or course of the coronary arteries are a key differential diagnosis for exertion angina. Multimodal coronary imaging is essential for determining the best course of action.

# BACKGROUND

Multiple studies indicate that the incidence of congenital coronary artery anomalies ranges between 0.3–5.6%.<sup>1,2</sup> Due to the wide availability of cardiac CT angiography (CCTA), however, a new study estimates the incidence to be 7.9%.<sup>3</sup> The most prevalent categorisation scheme is Angelini's 1999 proposal, which separates anomalous coronary arteries into three primary categories: anomalies of origin and course, anomalies of termination, and anomalies of intrinsic coronary artery architecture.<sup>4</sup>

Anomalies of origin and course are differences in the location of origin and course of the coronary arteries. They are further categorised into two subtypes: anomalies of opposite sinus of Valsalva (OSV) origin, and anomalies of interarterial course.<sup>5</sup>

Anomalies of OSV origin are coronary arteries that emerge from the opposite aortic sinus and run between the aorta and the pulmonary artery. This subtype comprises an anomalous LM from the right sinus, an anomalous right coronary artery from the left sinus, and an anomalous left circumflex artery from the right sinus.<sup>6</sup>

Anomalies of interarterial course are coronary arteries that run between the aorta and the pulmonary artery, regardless of their origin. This subtype consists of anomalous left main coronary artery (LM) from the right sinus, an anomalous right coronary artery from the left sinus, and the aberrant origin of both coronary arteries from the same aortic sinus.<sup>6</sup>

The abnormality may be classified into five clusters based on the anatomical link between the abnormal vessel, the aorta, and the pulmonary trunk. In a preaortic (interarterial) course, the aberrant vessel travels between the aorta and pulmonary artery. A prepulmonic (anterior) anomalous vessel follows the outflow of the right ventricle anteriorly, a subpulmonic (septal) anomalous vessel takes an intramyocardial path before reappearing in the proximal part of the interventricular groove, and a retroaortic (posterior) anomalous vessel travels posteriorly around the aortic root. In a retrocardiac course, an abnormal vessel flows behind the heart, as opposed to in front of it. The first aberrant pattern is regarded as the most

threatening, since it raises the risk of sudden cardiac death (SCD) in those individuals. $^{5-10}$ 

Anomalies of termination relate to variations in the distribution and termination of the coronary arteries. They are further categorised into ALCAPA and coronary artery fistulas.<sup>11</sup> ALCAPA refers to an aberrant left coronary artery that terminates in the pulmonary artery, as opposed to the aorta. This condition, also known as Bland-White-Garland syndrome, has the potential to produce left ventricular dysfunction and heart failure.<sup>12</sup>

Fistulas of the coronary arteries are aberrant connections between the coronary arteries and other heart chambers or vessels. These connections may result in ischaemia, myocardial infarction, or heart failure by diverting blood away from the myocardium.<sup>13</sup>

Abnormalities of intrinsic coronary artery anatomy include size, shape, and distribution variations in the coronary arteries. They can be further divided into two subgroups: myocardial bridging and aneurysms in the coronary arteries.<sup>11</sup> Myocardial bridging refers to a part of a coronary artery that runs through the myocardium, as opposed to above it. This condition can cause ischaemia during exercise or stress.<sup>14-16</sup> Coronary artery aneurysms refer to localised dilations of the coronary arteries that can be congenital or acquired. They are commonly seen in patients with Kawasaki disease.<sup>17</sup>

Based on the patient's clinical history, symptoms, and risk factors, the diagnosis of aberrant coronary arteries needs a high level of suspicion. In the majority of instances, imaging techniques like echocardiography, CCTA, cardiac MRI (CMRI), and coronary angiography are used to confirm the diagnosis.<sup>18,19</sup>

Echocardiography is a non-invasive imaging modality that can be used to detect anomalous coronary arteries, especially when combined with colour Doppler imaging. However, its sensitivity and specificity are limited, and it may not be able to detect all types of anomalous coronary arteries.<sup>20</sup>

CCTA is a highly sensitive imaging modality that can produce detailed images of the coronary

arteries and their course. It is particularly useful for detecting anomalous origins of the coronary arteries from the OSV, or interarterial course, as well as coronary artery fistulas. CCTA can also be used to assess the severity of stenosis or obstruction of the coronary arteries.

CCTA is a very sensitive imaging technique that can map the coronary arteries in great detail. Coronary artery fistulas and abnormal origins of the coronary arteries from the OSV can be detected using this technique, as well as assessment of coronary artery stenosis or occlusion.<sup>21</sup>

CMRI is a non-invasive imaging technique that creates pictures of the heart's internal structures using magnetic fields and radio waves. It can be used to detect anomalous coronary arteries, especially when combined with contrastenhanced angiography. CMRI can also provide information about myocardial perfusion and function.<sup>22</sup>

Invasive coronary angiography is the diagnostic imaging modality of choice for coronary artery anomalies because of the wealth of information it provides regarding the coronary arteries' anatomy, course, and distribution. Nonetheless, there is a small possibility of repercussions.<sup>23</sup>

Coronary artery anomalies can range from being completely incidental to being potentially fatal. This depends on the severity of the anomaly, and how it affects the arteries' origin, course, and distribution. It is important to take the patient's clinical condition and symptoms into account when deciding how to treat aberrant coronary arteries. In many cases, no treatment is necessary for asymptomatic people who have benign abnormalities, such as myocardial bridging or minor coronary artery aneurysms.<sup>24</sup>

A surgical or interventional procedure may be necessary for individuals who are experiencing symptoms, or who have high-risk abnormalities such as ALCAPA, or an interarterial course of the coronary arteries. Improving myocardial perfusion and avoiding ischemia, myocardial infarction, and SCD are therapy goals.<sup>23,24</sup>

Reimplantation of the anomalous coronary artery into the proper aortic sinus or bypass grafting of the aberrant section may constitute surgical therapy. This method can give a permanent cure and enhance long-term outcomes, but it has a greater risk of complications and a longer recovery time.<sup>25-27</sup>

Interventional therapy may entail percutaneous coronary intervention (PCI) or stent implantation; this procedure is less invasive than surgery, and can provide rapid symptom alleviation. When contemplating PCI, it is crucial to evaluate the course of these anomalies, and rule out malevolent ones. It may not be appropriate for all types of aberrant coronary arteries, and is linked to an increased risk of restenosis or thrombosis.<sup>28,29</sup>

The prognosis of aberrant coronary arteries depends on the type and severity of the anomaly, as well as the promptness and adequacy of treatment. Benign anomalies, such as myocardial bridging or small coronary artery aneurysms, have an excellent prognosis, and may not require any intervention.<sup>4,24</sup>

High-risk anomalies, such as ALCAPA or the interarterial course of the coronary arteries, have a poorer prognosis, and may require prompt intervention to prevent ischaemia, myocardial infarction, or SCD. In general, the long-term results of surgical or interventional therapy are favourable, although they are dependent on the patient's clinical condition and the efficacy of the surgery.<sup>30,31</sup>

The authors describe a patient with an abnormal LM originating from the right coronary cusp, who suffered from recurrent chest pain, and his treatment.

## **CASE PRESENTATION**

A 62-year-old male, with a past medical history of sensorineural deafness, hypertension, and Type 2 diabetes on oral hypoglycaemic medications, had reported to the cardiology clinic after several months of chest pain during exertion, which was deemed to be ischaemic in origin. A clinical examination, ECG, and echocardiography were done, showing normal findings.

Despite beginning treatment with antiischaemic medication, the patient continued to experience symptoms consistent with Canadian Cardiovascular Society (CCS) Grade II angina pectoris; as a result, the decision was made to perform coronary angiography after obtaining informed consent for the procedure. The right radial approach was used, using a 5F Judkins right coronary catheter.

After engaging the right coronary sinus and contrast injection (Figure 1), a small nondominant right coronary artery (RCA) was revealed, and the LM was found to arise from the right coronary ostium, which subsequently divided into the left circumflex artery and the diseased left anterior descending artery (LAD).

## MANAGEMENT

Since coronary angiography revealed a significant proximal lesion in the LAD, there was a need to delineate the course of the coronary arteries, for fear of an intra-arterial course with a high risk of SCD. If present, such a malignant course would require a different management strategy. This led to the decision to opt for CCTA. After receiving the patient's informed consent, a CCTA was performed, which revealed an anomalous LM originating from the right sinus of Valsalva. This aberrant LM followed a retroaortic course, and was the source of both the LAD and the left circumflex artery. The RCA was shown to originate from the more proximal part of the LM (Figure 2).

The patient underwent PCI to LAD through right radial using a 6F MPA1 guide catheter (Medtronic, Watford, Hertfordshire, UK) and BMW wire (Asahi Intecc, Nagoya, Japan), predilatation with non-compliant balloons of 2.25×15 mm and 2.5×20 mm, followed by deployment of three overlapping drug-eluting stents: XIENCE Skypoint<sup>™</sup> Stent (Abbott Vascular, Abbott Park, Illinois, USA) 3.0×28 mm; SYNERGY<sup>™</sup> (Boston Scientific, Marlborough, Massachusetts, USA) 2.75×16 mm, and XIENCE Skypoint<sup>™</sup> Stent (Abbott Vascular) 2.75×12 mm, followed by postdilation with stent balloon up to 26 ATM with excellent results achieving TIMI III flow (Figure 3).

The patient went home on the same day of the procedure on dual antiplatelet,  $\beta$ -blocker, and

Figure 1: Coronary angiography showing the right and left coronary arteries arising from the right sinus of Valsalva demonstrating significant proximal left anterior descending coronary artery lesion.





Figure 2: Cardiac CT angiography 3D volume-rendered image demonstrating the retroaortic course of the left main coronary artery from the right sinus of Valsalva after removing the right atrium and left atrium.

Figure 3: Left anterior descending coronary artery post-stenting with TIMI III flow.



statins as part of anti-ischaemic treatment, as well as angiotensin receptor blockers for control of hypertension, and empagliflozin with linagliptin for diabetes management.

## DISCUSSION

It is challenging to determine the burden of the aberrant origin of the left coronary artery from the right sinus of Valsalva since this abnormality is uncommon, and data from observational studies are very variable.<sup>32</sup> However, it accounts for 0.3–1.0% of cases in the dataset of Angelini,<sup>1</sup> with a retroaortic course being the most prevalent subtype (prevalence: 0.28%; 95% confidence interval: 0.21–0.35%).<sup>7</sup>

Anomalous coronary arteries have been reported to be susceptible to atherosclerosis, even at younger ages.<sup>33,34</sup> Coronary blood flow would be hampered if aberrant coronary arteries emerged from the other side of the coronary sinus, which is positioned between the pulmonary trunk and the ascending aorta, especially if highrisk criteria are present in the form of a slit-like opening in the proximal vessel morphology, acute angle takeoff, and interarterial course, which was not present in the authors' patient.<sup>35-37</sup>

By the time they are 20 years old, many individuals with aberrant LM coming from the opposite right coronary artery and the previously indicated high risk criteria have already passed away, usually during, or shortly after, strenuous exercise, which is characteristic of young, 'healthy' athletes.<sup>38,39</sup> However, the authors' patient presented with significant atherosclerotic changes in the LAD originating from the anomalous LM. The retroaortic route of the aberrant coronary artery suggested that the atherosclerotic changes in the coronary arteries were the true culprit in this patient's case of chest pains, rather than the abnormal artery itself.<sup>40-44</sup>

Despite advances in interventional technology and procedural improvements, coronary artery anomalies remain a challenge for interventional cardiologists. The use of PCI as a treatment for aberrant coronary arteries appears promising; nevertheless, proper topographical identification of the anomalous vessel's origin and proximal course is essential.<sup>38,45,46</sup> Particularly in the setting of coronary anomalies, the long-term care of patients who have had PCI often entails a variety of interventions, including pharmaceutical therapies, lifestyle changes, and routine follow-up visits.<sup>47</sup>

Changes in lifestyle, such as exercise, good nutrition, smoking cessation, and a healthy weight, contribute to heart health. Exercise improves quality of life, cardiovascular health, and the risk of future cardiac events. A healthy diet reduces cholesterol, blood pressure, and heart disease. Stopping smoking improves long-term PCI outcomes, since heart disease is a major risk factor. Maintaining a healthy weight improves overall health and reduces the chance of heart issues.<sup>48</sup> However, the authors' patient was not overweight, or a smoker.

Pharmacological treatments to avoid stent thrombosis include PCI followed by dual antiplatelet therapy (DAPT) with aspirin and a P2Y12 inhibitor. The patient's health, stent type, and bleeding risk will determine the DAPT length. Statins,  $\beta$ -blockers, and angiotensin–converting enzyme inhibitors may be administered as well. Statins,  $\beta$ -blockers, and angiotensin–converting enzyme inhibitors can decrease cholesterol, blood pressure, and heart rate, respectively.<sup>49</sup> The authors' patient was kept on all these medications and DAPT for 12 months.

Regular follow-up meetings with the healthcare practitioner are crucial for monitoring the patient's condition, reviewing the treatment plan's efficacy, and managing problems. Follow-up appointments are usually planned at regular intervals for the first year following the treatment and less often after that. At follow-up sessions, the doctor will examine the patient, check their prescriptions, and prescribe any required tests or imaging investigations. The authors' patient had his first clinic appointment after 1 month, where he was asymptomatic, and in a compensated state. The authors planned for a 3-month follow-up interval for the first year if he remained asymptomatic.<sup>47</sup>

Stress testing can measure symptoms, and stratify people who may be at higher risk for cardiac events. Echocardiography may assess heart function, particularly the ejection fraction. CCTA or coronary angiography can be utilised to visualise any possible anomalies in the coronary anatomy and evaluate the patency of any stents or grafts inserted during PCI. These imaging scans can be repeated to evaluate the patient's status and therapy efficacy.<sup>49</sup> However, because the authors' patient was asymptomatic, no additional evaluation was required.

#### References

- Angelini P. Coronary artery anomalies: an entity in search of an identity. Circulation. 2007;115(10):1296-305.
- Garg N et al. Primary congenital anomalies of the coronary arteries: a coronary arteriographic study. Int J Cardiol. 2000;74(1):39-46.
- Gentile F et al. Coronary artery anomalies. Circulation. 2021;144(12):983-96.
- Villa AD et al. Coronary artery anomalies overview: the normal and the abnormal. World J Radiol. 2016;8(6):537-55.
- Lanjewar CP et al. Anomalous origin of coronary artery from the opposite aortic sinus of valsalva-a single center experience with a therapeutic conundrum. Indian Heart J. 2021;73(3):289-94.
- Peñalver JM et al. Anomalous aortic origin of coronary arteries from the opposite sinus: a critical appraisal of risk. BMC Cardiovasc Disord. 2012;12:83.
- Cheezum MK et al. Anomalous aortic origin of a coronary artery from the inappropriate sinus of valsalva. J Am Coll Cardiol. 2017;69(12):1592-608.
- Krasuski RA et al. Long-term outcome and impact of surgery on adults with coronary arteries originating from the opposite coronary cusp. Circulation. 2011;123(2):154-62.
- Moore KL et al. "Thorax," Clinically Oriented Anatomy (2018) 8th edition, Philadelphia: Wolters Kluwer, pp.78-126.
- Kejriwal NK et al. Retroaortic course of the anomalous left main coronary artery: is it a benign anomaly? A case report and review of literature. Heart Lung Circ. 2004;13(1):97-100.
- Kastellanos S et al. Overview of coronary artery variants, aberrations and anomalies. World J Cardiol. 2018;10(10):127-40.
- Yau JM et al. Anomalous origin of the left coronary artery from the pulmonary artery in adults: a comprehensive review of 151 adult cases and a new diagnosis in a 53-year-old woman. Clin Cardiol. 2011;34(4):204-10.

**EMJ** 

- Sunkara A et al. Coronary artery fistula. Methodist DeBakey Cardiovasc J. 2017;13(2):78-80.
- Roberts W et al. Myocardial bridges: a meta-analysis. Clin Anat. 2021;34(5):685–709.
- Loukas M et al. Myocardial bridges: a review. Clin Anat. 2011;24(6):675-83.
- Corban MT et al. Myocardial bridging: contemporary understanding of pathophysiology with implications for diagnostic and therapeutic strategies. J Am Coll Cardiol. 2014;63(22):2346-55.
- Sheikh AS et al. Coronary artery aneurysm: evaluation, prognosis, and proposed treatment strategies. Heart Views. 2019;20(3):101-8.
- Angelini P et al. Coronary anomalies: incidence, pathophysiology, and clinical relevance. Circulation. 2002;105(20):2449-54.
- Kim SY et al. Coronary artery anomalies: classification and ECG-gated multi-detector row CT findings with angiographic correlation. Radiographics. 2006;26(2):317-33.
- 20. Warnes CA et al. ACC/AHA 2008 guidelines for the management of adults with congenital heart disease: a report of the American College of Cardiology/American Heart Association Task Force on Practice Guidelines (Writing Committee to Develop Guidelines on the Management of Adults With Congenital Heart Disease). Developed in Collaboration With the American Society of Echocardiography, Heart Rhythm Society, International Society for Adult Congenital Heart Disease, Society for Cardiovascular Angiography and Interventions, and Society of Thoracic Surgeons. J Am Coll Cardiol. 2008;52(23):e143-263.
- Oikonomou E et al. Current concepts and future applications of non-invasive functional and anatomical evaluation of coronary artery disease. Life (Basel). 2022;12(11):1803.
- Elmadi J et al. Cardiovascular magnetic resonance imaging: a prospective modality in the diagnosis and prognostication

of heart failure. Cureus. 2022;14(4):e23840.

- Brothers JA et al. Expert consensus guidelines: anomalous aortic origin of a coronary artery. J Thorac Cardiovasc Surg. 2017;153(6):1440-57.
- 24. Grau JB et al. Reimplantation for anomalous right coronary artery. JTCVS Tech. 2021 Jun;7:226-8.
- Jegatheeswaran A et al. Outcomes after anomalous aortic origin of a coronary artery repair: a Congenital Heart Surgeons' Society Study. J Thorac Cardiovasc Surg. 2020;160(3):757-71.e5.
- Lorber R et al.; AAOCA Working Group of the Congenital Heart Surgeons Society. Anomalous aortic origin of coronary arteries in the young: echocardiographic evaluation with surgical correlation. JACC Cardiovasc Imaging. 2015;8(11):1239-49.
- 27. Molossi I et al. Anomalous coronary arteries: a state-ofthe-art approach. Cardiol Clin. 2023;41(1):51-69.
- 28. Hauser M. Congenital anomalies of the coronary arteries. Heart. 2005;91(9):1240-5.
- 29. Al Umairi RS et al. Anomalous origin of the left coronary artery from the pulmonary artery: the role of multislice computed tomography (MSCT). Oman Med J. 2016;31(5):387-9.
- Leong SW et al. Anomalous left coronary artery from the pulmonary artery: case report and review of the literature. Int J Cardiol. 2009;133(1):132-4.
- Khan MS et al. Anomalous origin of left main coronary artery from the right sinus of valsalva: a case series-based review. Cureus. 2020;12(4):e7777.
- 32. Kapoor A et al. An unusual coronary trinity: single left coronary artery with the right coronary artery originating from the left main coronary artery and following a retro-aortic course. J Invasive Cardiol. 2011;23(6):E156-7.
- 33. Park E et al. Atherosclerotic coronary artery disease in a

younger adult with transseptal anomalous left coronary artery. JACC Case Rep. 2022;4(16):1060-4.

- Lee BY. Anomalous right coronary artery from the left coronary sinus with an interarterial course: is it really dangerous? Korean Circ J. 2009;39(5):175-9.
- 35. Park JH et al. Prevalence of congenital coronary artery anomalies of Korean men detected by coronary computed tomography. Korean Circ J. 2013;43(1):7-12.
- Rizzo S et al. Sudden death and coronary artery anomalies. Front Cardiovasc Med. 2021;8:636589.
- 37. Finocchiaro G et al. Anomalous coronary artery origin and sudden cardiac death: clinical and pathological insights from a national pathology registry. JACC Clin Electrophysiol. 2019;5(4):516-22.
- Niwa K. Coronary artery anomaly and sudden death–especially focus on anomalous left coronary artery arising from the right sinus. J Cardiol Cases. 2013;7(3):e86-8.
- Nogic J et al. Anomalous coronary arteries on computer tomography angiography: a pictorial review. Curr Cardiovasc Imaging Rep. 2017;10:35.
- 40. Sousa E, Casanova J. Coronary

artery abnormalities: Current clinical issues. Rev Port Cardiol (Engl Ed). 2018;37(3):227-35.

- Halabchi F et al. Sudden cardiac death in young athletes: a literature review and special considerations in Asia. Asian J Sports Med. 2011;2(1):1-15.
- 42. Graidis C et al. Percutaneous coronary intervention and stenting in a single coronary artery originating from the right sinus of valsalva. Hellenic J Cardiol. 2013;54(5):401-7.
- 43. Constantinides SS et al. Transradial primary percutaneous intervention in a rare case of anomalous origination of the left coronary artery system from the right aortic sinus. JACC Cardiovasc Interv. 2014;7(11):e179-81.
- 44. Koza Y et al. Percutaneous coronary intervention in a rare case of single coronary ostium presented with ST elevation myocardial infarction. Eurasian J Med. 2019;51(3):307-9.
- Han J et al. Sudden cardiac death in athletes: facts and fallacies. J Cardiovasc Dev Dis. 2023;10(2):68.
- Neumann FJ et al.; ESC Scientific Document Group. 2018 ESC/ EACTS guidelines on myocardial revascularization. Eur Heart J. 2019;40(2):87-165.
- 47. Eckel RH et al.; American College of Cardiology/American Heart

Association Task Force on Practice Guidelines. 2013 AHA/ACC guideline on lifestyle management to reduce cardiovascular risk: a report of the American College of Cardiology/American Heart Association Task Force on Practice Guidelines. Circulation. 2014;129(25 Suppl 2):S76-99.

- 48. Levine GN et al. 2016 ACC/ AHA guideline focused update on duration of dual antiplatelet therapy in patients with coronary artery disease: a report of the American College of Cardiology/ American Heart Association Task Force on Clinical Practice Guidelines. J Am Coll Cardiol. 2016;68(10):1082-115.
- 49. Patel MR et al. ACC/AATS/AHA/ ASE/ASNC/SCAI/SCCT/STS 2017 appropriate use criteria for coronary revascularization in patients with stable ischemic heart disease: a report of the American College of Cardiology Appropriate Use Criteria Task Force, American Association for Thoracic Surgery, American Heart Association, American Society of Echocardiography, American Society of Nuclear Cardiology, Society for Cardiovascular Angiography and Interventions, Society of Cardiovascular Computed Tomography, and Society of Thoracic Surgeons. J Am Coll Cardiol. 2017;69(17): 2212-41.