

# Anomalous coronary artery anatomy with a single coronary ostium arising from the right coronary: a case report and literature review

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Bothaina Mohammed  
Abdulshakour<sup>1</sup>  
Muhammad Saeed<sup>2</sup>  
Mohiuddin M Taher<sup>3,4</sup>

<sup>1</sup>Department of Radiology, Al-Noor Specialist Hospital, Makkah, Saudi Arabia;

<sup>2</sup>Faculty of Medicine, Department of Radiology, Umm-Al-Qura University, Makkah, Saudi Arabia; <sup>3</sup>Department of Medical Genetics, Umm-Al-Qura University, Makkah, Saudi Arabia;

<sup>4</sup>Science and Technology Unit, Umm-Al-Qura University, Makkah, Saudi Arabia

**Purpose:** This article reports the case of a single coronary artery with origin from a single ostium in the right sinus of Valsalva (RSOV) with an anomalous course of the left coronary artery anterior to the pulmonary trunk.

**Results:** We present the case of an anomalous single coronary artery detected incidentally during routine coronary computed tomography (CT) angiography performed by a multislice CT method, using a 64-detector-row scanner. The CT examination revealed that the patient had a single large right coronary artery arising from the right sinus of Valsalva, dividing into the right coronary artery, left main coronary artery (LMCA), and left circumflex artery (LCX). The LMCA turned anteriorly in front of the pulmonary outflow tract, reaching the atrioventricular groove. The LCX artery passed posterior to the aorta (retro-aortic) and then between the left atrium and left ventricular outflow tract to reach its normal course on the left side.

**Conclusion:** We present a rare case of single coronary artery congenital anomaly from the Makkah region of Saudi Arabia. This kind of coronary artery anomaly in the absence of stenosis could remain silent and asymptomatic.

**Keywords:** anomalous single-coronary artery, multislice CT (MSCT), solitary coronary ostium, cardiac computed tomography (CCT), coronary angiography, coronary-computed tomography (CT)-angiography

## Introduction

Coronary anomalies are a rare heart condition that affect a small percentage of the general population. Coronary anomalies involve the origin or distribution of the artery and rarely are these anomalies clinically significant. Frequently, coronary artery deformities are related to other cardiac malformations and the diagnosis is made after birth.<sup>1</sup> In rare cases, coronary artery anomalies are an isolated and asymptomatic cardiac malformation.<sup>2,3</sup> Very rarely, all three coronary arteries originate from a single coronary ostium; a single coronary artery (SCA) arising from the left aortic sinus is extremely more uncommon than one arising from the right sinus.<sup>3,44</sup> A SCA in the absence of other major congenital anomalies is very rare with an incidence of 0.024–0.06%.<sup>5,6</sup> In rare cases, it is not possible to perform catheterization of both coronary arteries because of the absence of one of the arteries arising from the ostium of the coronary sinus. The practice of cardiac computed tomography (CT) allows imaging of the coronary anatomy 3-dimensionally and establishes an additional significance to coronary angiography. Visualization of the origin and course of the

Correspondence: Mohiuddin M Taher  
College of Medicine, Al-Abidiya Campus,  
Umm-Al-Qura University, Taif Road,  
21955 Makkah, Saudi Arabia  
Email TMMohiuddin@uqu.edu.sa

Bothaina Mohammed Abdulshakour  
Department of Radiology, Al-Noor  
Specialist Hospital, 3rd Ring Road, 24242  
Makkah, Saudi Arabia  
Email bshakour@gmail.com

left and right coronary arteries can be achieved by a multislice CT method.<sup>7,8</sup> In SCA anomaly, there is only one coronary artery ostium from which both coronary arteries arise with essentially normal peripheral distribution.<sup>2,4</sup> We present a rare case of SCA (Lipton's type RIIIA) congenital anomaly from the Western region of Saudi Arabia.

## Case report

This study was approved by the Institutional Review Board (IRB) for bioethics of Umm al Qura University, Makkah, Saudi Arabia (IRB number HAPO-02-2019-03-310), and was performed in accordance with the principles of the Declaration of Helsinki. Informed consent was obtained from the patient for publishing the images and the case details. A 49-year-old female patient was referred to our hospital in Makkah (Saudi Arabia) for evaluation of atypical chest pain and shortening of breathing. Her vital signs and physical examination were unremarkable. Blood work showed negative cardiac troponin. Echocardiography revealed well-preserved left ventricle systolic function with mild tricuspid valve regurgitation. The electrocardiogram did not show any ischemic changes. The patient was then referred to the radiology department for cardiac CT angiography.

The CT angiogram demonstrated a SCA arising from the single ostium from the right sinus of Valsalva and dividing into the right coronary artery (RCA), left anterior descending (LAD) artery, and left circumflex artery (LCX). Figure 1(A–C) shows the multislice CT volume-rendered reformatted images. The SCA (Figure 1A, B) arises from the single ostium from the RSOV (Figure 1B) and divides into the RCA (Figure 1A, B), LAD artery, and LCX (Figure 1A–C). The LAD artery turns anteriorly in front of the right ventricular outflow tract (Figure 1C), and the LCX shows a retro-aortic course (Figure 1C). The cross-sectional axial view displaying the pre-pulmonic course of the anomalous left coronary artery, pulmonary artery, and aorta (AO) is shown in Figure 2A. Figure 2(B) shows the cross-sectional axial oblique view of the retro-aortic course of the LCX, aortic sinuses, and left ventricle. Cross-sectional axial oblique views showing a single coronary ostium arising from the RSOV and trifurcated into the normally coursed dominant RCA, anomalous pre-pulmonic LAD artery, retro-aortic LCX, and left ventricle are presented in Figure 3(A, B). Cross-sectional longitudinal oblique views in Figure 4(A–C) show a single coronary ostium (Figure 4A) arising from the RSOV and the

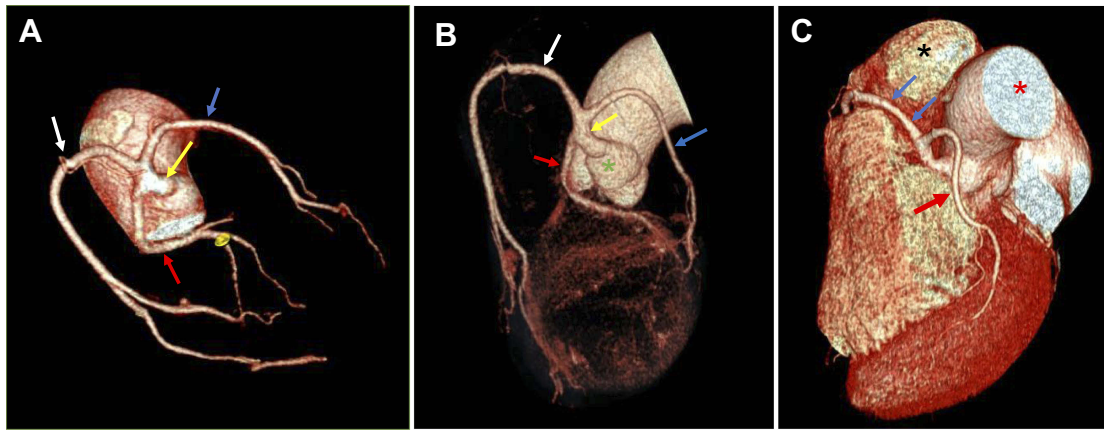
pre-pulmonic course of the anomalous left coronary artery (Figure 4A–C), left ventricle (Figure 4A, B), aorta (Figure 4A–C), and right ventricle (Figure 4A, B); the left atrium (Figure 4A, B), retro-aortic LCX (Figure 4C), and dominant RCA (Figure 4C) are also clear.

The patient tolerated the CT procedure well with no immediate complications and left the department in a stable condition. The patient was referred back to the cardiology department for further management. The patient was advised by the cardiologist with regard to the benign nature of this incidental findings and she was advised for regular follow-up.

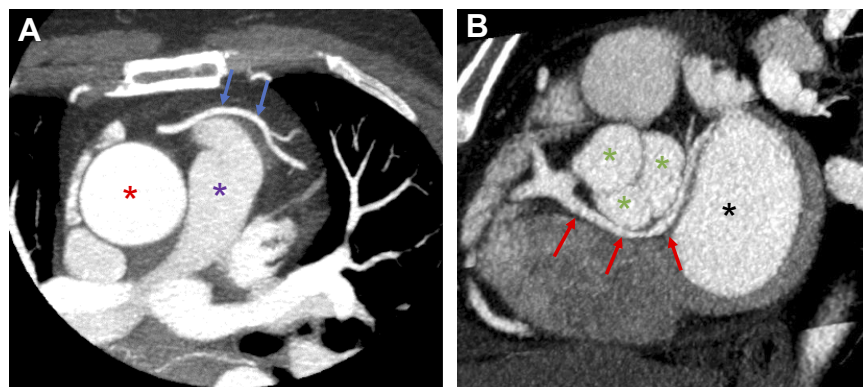
## Discussion

In Saudi Arabia, radiology units in many centers are using regularly the sensitive cardiac-imaging technologies such as coronary CT angiography to screen and detect these coronary abnormalities, and several reports appeared recently from many parts of Saudi Arabia.<sup>1,9–11</sup> Consanguineous marriages are common in many Middle Eastern countries including Saudi Arabia, and with around 57.7% prevalence of consanguinity with first-cousin marriages being the most frequent, we expect higher prevalence of congenital heart diseases in Saudi Arabia.<sup>12,13</sup> However, a recent retrospective analysis of 6 years of coronary CT angiography scans showed the coronary artery anomalies were similar to literature data.<sup>9</sup> However, this abnormality is not very common; few reported cases of a SCA with origin from a single ostium in the right sinus of Valsalva with an anomalous left coronary artery were reported by radiologists. We have searched in the PubMed database for these reported cases and summarize some of the reported cases of anomalous left coronary artery in Table 1.

The most common congenital anomaly is separate origin of the LAD artery and LCX from the left coronary sinus. The second most common anomaly is the origin of the circumflex artery from the right coronary sinus. However, there are several reported cases of anomalous single right artery also reported in other investigations.<sup>14</sup> Also, several cases of RCA arising from the left aortic sinus have also been reported by coronary angiogram and are benign most of the time, and patients are asymptomatic.<sup>3,4,15</sup> A solitary, unique origin of the coronary vessels may be linked with sudden cardiac arrest, particularly in cases with an inter-arterial course, and usually these patients complain initially about angina.<sup>16</sup> The risk factors for myocardial ischemia in anomalous



**Figure 1** (A, B, and C) Multislice computed tomography volume-rendered reformatting images showing a single coronary artery (yellow arrow, A and B) arising from the single ostium from the right sinus of Valsalva (green asterisk, B) and dividing into the RCA (white arrow, A and B), LAD artery (blue arrow) and LCX (red arrow, A–C). The LAD artery turns anteriorly (blue arrows, C) in front of the right ventricular outflow tract (black asterisk, C) and LCX (red arrow, C) showing a retro-aortic course (red asterisk, C).  
**Abbreviations:** LAD, left anterior descending; LCX, left circumflex artery; RCA, right coronary artery.

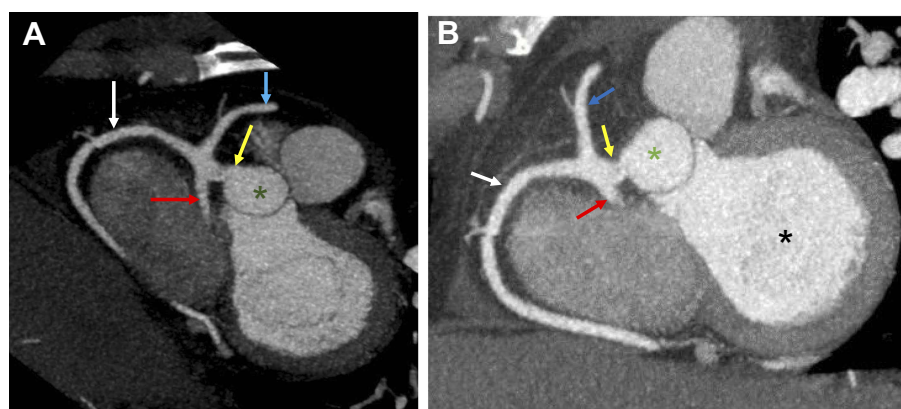


**Figure 2** (A) Cross-sectional axial view showing the pre-pulmonic course of the anomalous left coronary artery (blue arrows), pulmonary artery (purple asterisk), and aorta (red asterisk). (B) Cross-sectional axial oblique view showing retro-aortic course of the left circumflex artery (red arrows), aortic sinuses (green asterisks), and left ventricle (black asterisk).

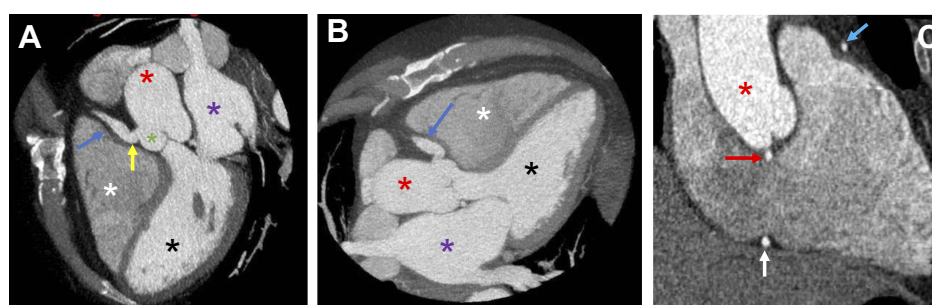
artery patients can be smoking, hypercholesterolemia, a past history of diabetes mellitus, dyslipidemia, obesity, retrosternal chest pain, and a positive family history of premature coronary artery disease.<sup>2–4</sup> In some cases, the initial troponin test can be negative, and an electrocardiogram will not show any ischemic changes.<sup>17</sup>

The case of a SCA with origin from a single ostium in the right sinus of Valsalva with an anomalous course of the left coronary artery anterior to the pulmonary trunk is presented here. The definition of a heart with a SCA is one in which the entire myocardium is nourished by an artery, regardless of distribution, that arises by one ostium from an arterial trunk.<sup>18</sup> The current classification system was introduced by Lipton et al,<sup>19</sup> combining with two previously published systems defined by Smith<sup>18</sup> and by Ogden and Goodyer.<sup>2020</sup> In this classification, the origin of the SCA

from the right sinus of Valsalva is defined as “R” and from the left sinus of Valsalva as “L”. The normal anatomical course of the SCA is defined as “I”. The type “II” SCA provides the contralateral coronary artery and crosses the base of the heart to assume its inherent normal position. In the type “III” SCA, after leaving the right coronary sinus of Valsalva, the LAD artery and LCX arise separately from the proximal part of the artery. A further subdivision can be made according to the anatomical course. In type “A” the main left or right coronary passes anterior to the pulmonary artery, in type “B” it passes between the aorta and pulmonary artery, and in type “P” it is posterior to the aorta. The SCA is an extremely exceptionally rare coronary abnormality, and it is usually diagnosed accidentally during conventional coronary artery angiography or on postmortem evaluation. The etiology of SCA is uncertain, however; in



**Figure 3 (A and B)** Cross-sectional axial oblique view showing a single coronary ostium (yellow arrow) arising from the right sinus of Valsalva (green asterisk) and trifurcated into the normally coursed dominant right coronary artery (white arrow), anomalous pre-pulmonic left anterior descending artery (blue arrow), retro-aortic left circumflex artery (red arrows), and left ventricle (black asterisk).



**Figure 4 (A, B, and C)** Cross-sectional longitudinal oblique views showing a single coronary ostium (yellow arrow, **A**) arising from the right sinus of Valsalva (green asterisk) and the pre-pulmonic course of the anomalous left coronary artery (blue arrow, **A–C**), left ventricle (black asterisk, **A** and **B**), aorta (red asterisk, **A–C**), and right ventricle (white asterisk, **A** and **B**), the left atrium (purple asterisk, **A** and **B**) and right ventricle (white asterisk, **A** and **B**), and the retro-aortic left circumflex artery (red arrow, **C**) and dominant right coronary artery (white arrow, **C**).

some cases, SCA may lead to life-threatening symptoms, including angina pectoris, myocardial infarction, syncope, cardiac arrhythmias, and congestive heart failure, or this anomaly has been associated with sudden cardiac death in younger patients.<sup>19,21</sup> Conventional coronary artery angiography is the gold standard for the evaluation of coronary artery disease; however, non-invasive imaging methods, with better spatial resolution, such as multislice CT, can provide anatomical information that may have prognostic value.<sup>22,23</sup>

Left coronary artery arising from RSOV is a serious abnormality, and patients with the intra-arterial type of this anomaly are usually referred to surgery.<sup>24</sup> Those with the other types are thought to be at low risk and do not require surgery. The patient in this present case complained about chest pain and shortness of breath, maybe because of her mild tricuspid valve regurgitation seen initially by echocardiography or anxiety. There was no compression of the anomalous left coronary artery between the AO and pulmonary artery, and we believe

that this discomfort was less likely caused by the pressure of the aorta on the LAD artery. In the majority of patients with a SCA or anomalous origin of a coronary artery, the native coronary artery provides adequate flow during most situations, because it is a benign lesion. This patient's initial presenting symptoms could be anxiety related or due to mild tricuspid valve regurgitation. For the present case, no grafting is recommended to deliver a stable flow to the left coronary system. In the majority of patients with a SCA or anomalous origin of a coronary artery, the native coronary artery provides adequate flow during most situations, because it is a benign lesion. Surgical correction is generally recommended for patients who have symptoms, or when this anomaly is identified in young age.<sup>24</sup>

## Conclusions

We have diagnosed this patient with a single RCA by dual-source CT angiography with type R, IIIA anomaly. This anomaly is “benign” in the absence of the LAD artery



**Table I** Previously published cases with anomalous left coronary artery arising from the right coronary artery

Clinical Symptoms	Sex	Age (years)	Country of Origin	Author	Reference No
Lateral wall MI	Male	74	Kosovo	Prifti et al	25
Angina pectoris	Female	46	South Africa	Smedema et al	26
Shortness of breath, fever, and night sweats	Male	48	United Kingdom	Neil et al	21
Unstable angina pectoris	Male	56	Canada	Gleeson et al	17
Sudden onset of retrosternal pain	Male	69	The Netherlands	Mihl et al	27
Angina MI	Female	63	Romania	Hailu et al	28
Multiple atypical chest pain	Female	59	USA	Summers et al	29
Chest pain	Female	49	India	Sinha et al	30
Unstable angina pectoris	Female	62	Croatia	Vincelj et al	31
Exertional dyspnea	Female	41	Republic of Korea	Kang et al	32
Hypertension, angina	Female	75	Spain	Tejada et al	33
Chest discomfort on exertion	Male	45	Turkey	Dogan et al	34
Previous MI	Female	47	Italy	Fiorilli et al	35
Exercise-induced spells/neurocardiogenic syncope	Female	16	USA	Johnson et al	36
Atypical chest pain	N/A	N/A	USA	Anuwatworn et al	37
Exertional angina	Male	43	Omani	Panduranga and Riyami	38
Chest pain on exertion	Female	47	Turkey	Karabay et al	39
Non-ST-elevated MI	Male	55	The Netherlands	Liesting et al	23
Atypical chest pain	Male	68	Turkey	Cakar et al	40
Inferior wall MI	Male	68	India	Subban et al	41
Chest pain and dyspnea on exertion	Female	50	Turkey	Okuyan and Dinckal	42
Posterolateral MI	Male	73	Iran	Kheirikhah et al	43
MI during exercise	Male	14	Japan	Okamoto et al	44
Dizziness and syncope on mild exertion, vague left-sided chest discomfort	Female	46	Saudi Arabia	Hassan et al	10
Smoking history, with acute onset typical chest pain	Male	38	Saudi Arabia	Al-Bugami et al	11
Anterior-lateral ST-elevated MI	Male	71	Italy	Marchesini et al	45
Cardiac arrest	Female	10	Italy	De Rosa et al	46
Atrial septal defect	Female	4	Saudi Arabia	Al-Muhaya et al	1

**Abbreviations:** MI, myocardial infarction; N/A, not available.

between the aorta and pulmonary artery. A SCA is a rare congenital anomaly. Coronary CT angiography with better spatial resolution provides accurate angiographic information on the origin, course, and termination of coronary anomalies non-invasively. A SCA, in the absence of coronary stenosis and an anatomical malignant course, could remain silent and asymptomatic.

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## Disclosure

All authors agreed with the contents of this manuscript and all authors declare no conflicts of interest in this work.

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