

## An entire coronary system arising from right coronary cusp: a rare anomaly

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### ABSTRACT

The prevalence of coronary artery anomalies is approximately 0.6% in individuals undergoing angiography. Most of the anomalies are benign, but some can lead to myocardial infarction, cardiomyopathy, and sudden cardiac death. It is very rare to have an entire coronary circulation that arises from the right coronary cusp. We present a case of a 57-year-old male who presented with complaints of chest pain and dyspnea on exertion. An invasive angiogram revealed all the three coronary arteries originating from the right coronary cusp. It is crucial to define coronary anatomy as anomalies dictate which cardiac intervention should be attempted in cases of ischemia.

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## 1. Introduction

Coronary artery anomalies (CAA) are usually an incidental finding in 0.3% to 1% of the healthy individuals [1]. CAA are a cause of sudden cardiac death especially in young patients, with a prevalence of approximately 1% among the general population [2]. An accurate determination of the course of coronary arteries is essential for interventions during invasive angiogram (IA) and cardiac surgery [3].

An origin of all three coronary arteries arising from right coronary cusp is a very rare anomaly. Patel et al. [4] described a 0.6% prevalence of CAA in individuals undergoing angiography; however, the prevalence of this particular anomaly was 3.1% among those 0.6% reported. We present a case of a male who underwent a left heart catheterization for chest pain; however, on IA, it was determined that the entire coronary circulation emerged from the right coronary cusp.

## 2. Case presentation

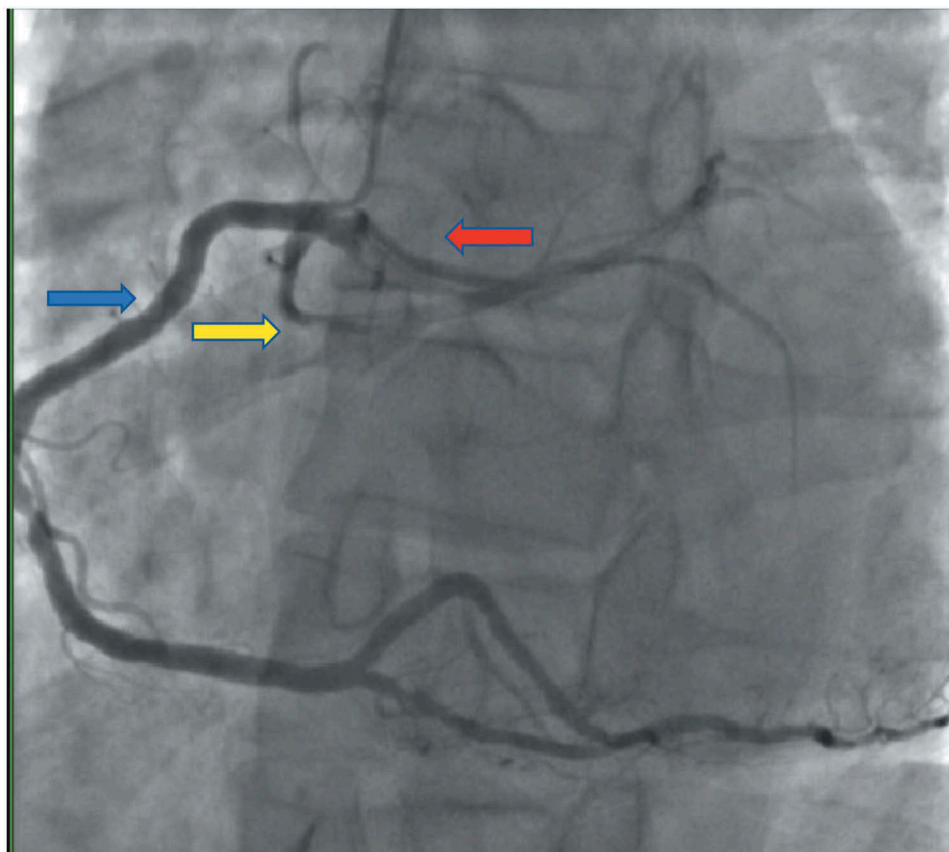
A 57-year-old Caucasian male presented to the cardiologist's office with complaints of chest pain and dyspnea on exertion. The patient had been having chest pain on exertion with relieving at rest along with shortness of breath which was limiting his functional status for the past few weeks. He did have a history of chronic obstructive pulmonary disease, hypertension, and back pain. The patient initially had an echocardiogram (ECHO) that measured ejection fraction to be 60%–65%. A nuclear-gated chemical stress test was

performed that identified ST-depression on EKG on stress images. Therefore, an IA was planned in order to evaluate the coronary circulation. IA identified all three coronary arteries arising from the right coronary cusp (Figure 1). The suspicion to investigate further an anomalous origin of left circulation emerged as there was absent left coronary circulation when contrast was injected from the left coronary cusp. The IA did not identify any obstructive coronary artery disease. The patient was managed medically by advising to avoid strenuous activities and placed on beta blockers.

## 3. Discussion

The prevalence of congenital coronary artery anomalies in patients undergoing coronary angiography is 0.3–5.6% and approximately 1% are identified on routine autopsy [2]. Although generally, coronary artery anomalies are benign in nature, some CAA can be potentially lethal leading to arrhythmias, congestive heart failure, myocardial infarction, syncope and sudden cardiac death (SCD) [5]. CAA are involved with approximately 12% of sports-related sudden cardiac death as compared to 1.2% of non-sports related deaths [6].

Yamanaka O et al. [7] proposed a classification of CAA that projected the origination of left anterior descending (LAD) and left circumflex (LCx) from the opposite sinus to be serious as these can lead to fatal consequences such as sudden cardiac death and myocardial infarction. Anomalous LAD from right cusp has been related to SCD in 59% of patients and post-exercise in 81% of patients [8].



**Figure 1.** Invasive angiogram showing right coronary artery (Blue Arrow), left circumflex artery (Yellow Arrow), left anterior descending artery (Red Arrow).

There have been multiple hypotheses regarding the cause of sudden cardiac death in individuals having the particular CAA with the origin of LAD and LCx from the right coronary cusp. 1) The anomalous artery courses between the great vessels (aorta and right ventricular outflow tract) through a tunnel, 2) an acute angle take-off of the artery, 3) stretch of the intramural segment and compression between the commissure of the right and left coronary cusps, and 4) atherosclerotic disease-causing ischemia. The increase in blood flow in the great vessels during exercise leads to their expansion; therefore, it leads to increase in the angulation of the anomalous artery thereby decreasing its inner diameter which ultimately leads to decrease blood flow causing ischemic symptoms during exercise [6,9,10].

There are a few diagnostic modalities that can detect CAA. The transesophageal echocardiogram (TEE) can help in determining the course of the anomalous artery between the great vessels [11]. Non-invasive imaging including multi-slice computed tomography (MSCT) and cardiac magnetic resonance imaging (MRI) are being used more for evaluating coronary circulation, but utility of MRI is limited by its lack of widespread availability [2]. MSCT are better at visualizing coronary circulation and can also determine the course of anomalous arteries [12]. IA is still the diagnostic modality that

is used generally to evaluate coronary arteries and can identify CAA.

Our patient did not have any obstructive coronary artery disease but still was having symptoms on exercise possibly due to the mechanisms described above. There are no general guidelines to manage this particular CAA; however, due to the association of SCD with anomalous artery and exercise, patients are generally advised to avoid strenuous activity. The management of coronary artery disease in the setting of CAA include medical management, percutaneous coronary intervention (PCI) or surgical repair [5]. PCI is generally complex and technically demanding in these anomalous arteries due to its angulation from aorta, ostial configuration, site of atherosclerotic lesion and the course of the artery [10]. Cardiac surgery includes direct repair of origin of the anomalous artery in the aortic root and coronary artery bypass surgery (CABG). CABG is more feasible; however, it also has limitations due to the longevity of the grafts [6].

#### 4. Conclusion

This case describes a potentially fatal and extremely rare CAA. CAA is essential to know before any interventions as these can be damaged during procedures, ultimately leading to fatal outcomes.

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