

Classification dilemma between right single coronary artery and agenesis of left main coronary artery: A rare coronary anomaly

*Tek sağ koroner arter ve sol ana koroner arter agenezi arasında sınıflamada çelişki:
Nadir bir koroner anomali*

Zafer Kök¹, Erdogan Sökmen², Bilge Bingöl³

Department of Cardiology, Kırşehir Ahi Evran Training and Research Hospital, Kırşehir, Türkiye

The term “coronary artery anomaly” encompasses a broad spectrum of congenital abnormalities that affect the origin, path, and structure of the epicardial coronary arteries. With an estimated occurrence rate of 0.024 to 0.066% in the general population, single coronary artery (SCA) is an uncommon congenital anomaly characterized by the presence of a solitary coronary artery originating from a single coronary ostium, providing blood supply to the entire heart.^[1-3] While the affected cases may be asymptomatic, they also are likely to be admitted with a variety of symptomatology, including typical or atypical anginal chest pain, ventricular arrhythmias, syncope, acute coronary syndrome, or sudden cardiac death.^[2] Herein, we presented a unique case of right SCA that does not fit the current classification system.

A 67-year-old male patient was admitted to our cardiology clinic with a complaint of a burning sensation in his chest after exercising. The patient had a history of smoking for over 25 years and had hypertension. Electrocardiogram was unremarkable other than a 1.5-mm ST-segment elevation in augmented voltage right (aVR). Echocardiographic evaluation revealed a mild mitral regurgitation with a global left ventricular ejection fraction of 55%. During the treadmill test, the patient had to stop after 5 min due to dyspnea and high blood pressure. It was then decided to proceed with invasive coronary angiography to evaluate the patient further, which depicted an absent left main stem (LMS) and normal right heart coronary artery (RCA). Of note, the left anterior

descending artery (LAD) was in its inherited course and was observed to be a continuation of RCA in the posterior interventricular groove near the cardiac apex rather than a bifurcation of LMS in the base of the heart (Figures 1, 2; Videos 1, 2). The left



Figure 1. Left anterior oblique view depicting the interarterial communication between the body of LAD and the RCA. Note that there is no filling of the left main coronary artery.

LAD: Left anterior descending; RCA: Right coronary artery.

Corresponding author: Zafer Kök.

E-mail: zfrkok@gmail.com

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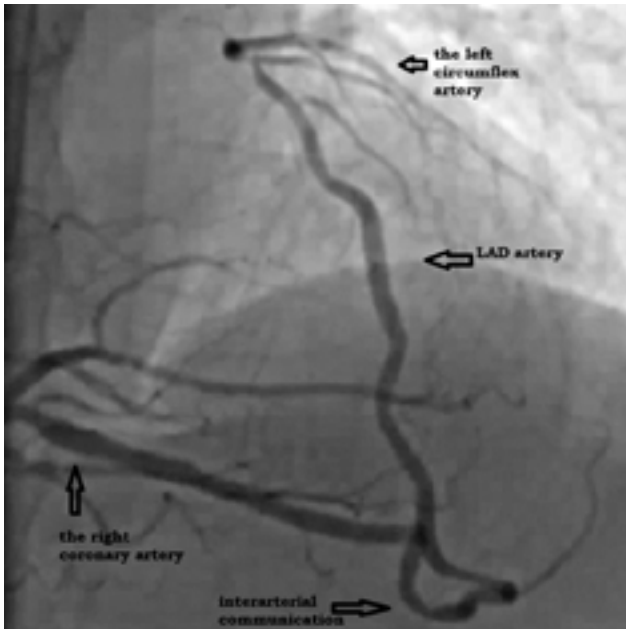


Figure 2. Left anterior oblique view with cranial angulation showing lack of the left main coronary artery.



Video 2. Left anterior oblique view with cranial angulation showing the lack of retrograde filling of the left main coronary artery.



Video 1. Left anterior oblique view showing the interarterial communication between the body of LAD artery and the RCA, retrogradely filling the LAD and the left circumflex coronary arteries.

LAD: Left anterior descending; RCA: Right coronary artery.

circumflex artery (LCX) was observed to originate from the proximal part of the LAD and to traverse its inherent course. A subsequent aortography was unable to localize the LMS (Video 3). At this point,



Video 3. Angiography of the aortic root using a pigtail catheter, which shows the lack of antegrade filling of the left main coronary artery.

a chronic total occlusion was considered in the LMS. Subsequently, a coronary computed tomography scan was planned to delineate the ultimate coronary anatomy, which revealed LMS agenesis and a patent single RCA supplying the whole heart by giving off

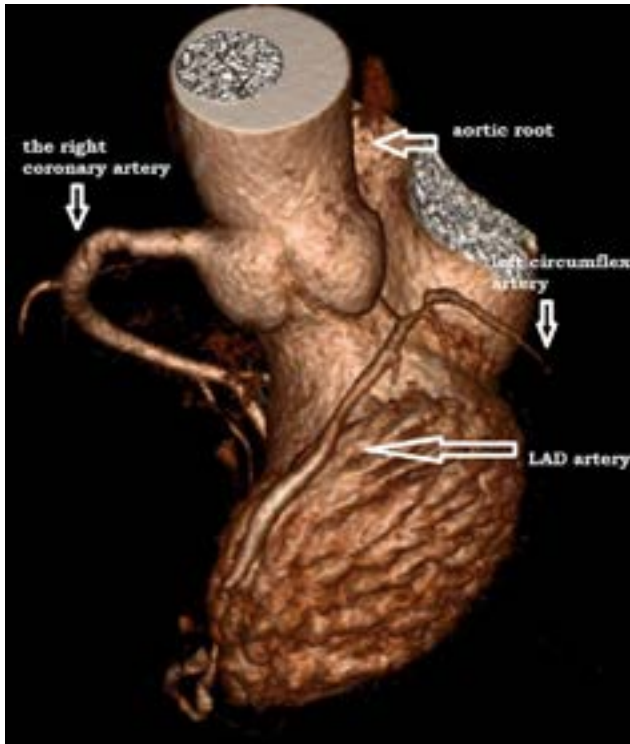


Figure 3. Coronal computed tomography image depicting the agenesis of the left main coronary artery.

the LAD and subsequently the LCX (Figures 3, 4). We decided to follow the patient conservatively with lifestyle modifications and medical therapy as appropriate. A written informed consent was obtained from patient.

Single coronary artery is an extremely rare coronary anomaly, in which only one coronary artery provides blood supply to the entire heart. Lipton et al.^[3] introduced a valuable angiographic classification system for SCA based on the site of origin and anatomical distribution of its branches. This classification divides cases into right-type (R) and left-type (L) based on the origin of the SCA in the right or left sinus of Valsalva. Additionally, each case is further categorized into types 1, 2, or 3 based on the specific anatomical course of the artery. Type 1 denotes a right SCA arising from the right coronary sinus, which then continues in the left atrioventricular groove as LCX before it terminates as LAD or as LMS, branching into LCX and LAD. The LCX then terminates as RCA. In type 2, SCA originates from either the right or left coronary sinus. From the proximal segment of this SCA, an additional anomalous artery arises and crosses the base of the heart before returning to its original path.

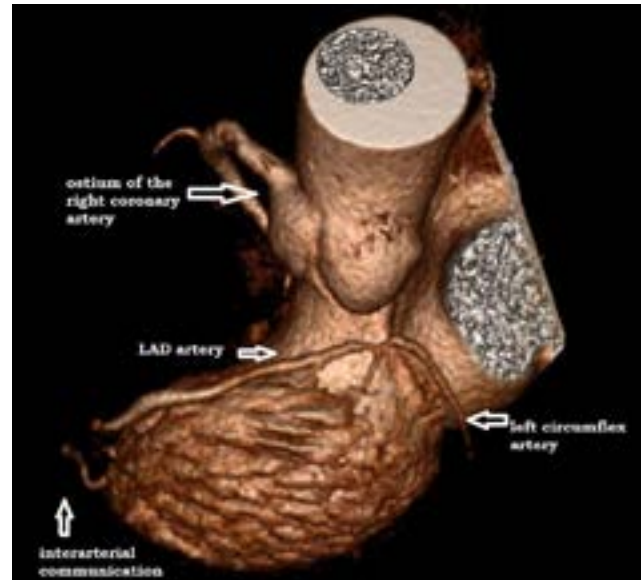


Figure 4. Coronal computed tomography image showing the origination of the RCA from the aortic sinus of Valsalva and agenesis of the left main coronary artery.
RCA: Right coronary artery.

Type 3 corresponds to a scenario where LCX and LAD separately originate from the proximal RCA. Furthermore, Lipton et al.^[3] classified SCA into three categories based on its relationship to the aorta and the main pulmonary artery. Category A refers to the anomalous artery passing in front of the main pulmonary artery. Category B is characterized by the anomalous artery passing between the ascending aorta and the main pulmonary artery. Category P indicates that the anomalous artery passes behind the ascending aorta. In 1990, Yamanaka and Hobbs^[4] further modified this model into a more useful framework for understanding and categorizing different variations of SCA and introduced two additional groups (S and C). Group S denotes a transseptal course of the anomalous artery, while Group C represents a combined course that combines elements from other categories.

The anomaly in our case report does not fit into the modified Lipton classification, and it may be argued that it is not a true SCA example. Therefore, we may propose a unique combination of coronary anomalies where there is a LMS agenesis together with single RCA connected to the LAD artery through an interarterial communication near the apical part of the left ventricle. Interarterial communications arise from fetal coronary circulation patterns and are generally larger than 1 mm in diameter. They possess

straight routes and gentle curvatures, which are their main differentiating factors from the coronary collaterals.^[5] Moreover, most of the coronary interarterial communications occur between RCA and LCX in the posterior interventricular groove and between RCA and LAD in the distal interventricular groove.^[6] Hence, it would also be prudent to regard the connection between LAD and RCA in our case report as a true interarterial communication. To our knowledge, this is the first case report to suggest such a classification dilemma in coronary anomalies.

The optimal treatment approach for SCA is not well-defined. The decision on therapy should be guided by the specific course of the anomalous coronary artery and any associated coronary atherosclerosis. In cases where the anomalous artery courses between the aorta and main pulmonary artery, or when there is significant atherosclerosis, coronary artery bypass surgery may be beneficial. Revascularization strategies, such as percutaneous coronary intervention, have also been reported to be successful in certain cases.^[4] Due to the fact that the entire coronary circulation relies on SCA, any complication would be inadequately tolerated. Therefore, the treatment plan should be tailored to the individual patient's condition and needs. However, it is of paramount importance for cardiologists to be aware of this uncommon condition, particularly before considering any percutaneous coronary intervention for chronic total occlusions. A lack of awareness of the types of SCA anomalies could lead to severe coronary or cardiac perforations. Therefore, coronary computed tomography angiography and magnetic resonance imaging provide accurate depiction of the coronary anatomy for the definitive diagnosis and risk stratification before implementation of a treatment modality.

In conclusion, we presented an extremely rare case of right SCA anomaly. However, failure of our case to comply with the classification criteria proposed in the literature may lead to a classification dilemma. Such coronary anomalies should be kept

in mind before performing percutaneous coronary interventions, and imaging modalities should be used to depict the exact coronary anatomy to any procedural complication.

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Author Contributions: Conceived the presented idea: Z.K., E.S.; Collected and analyzed case images: Z.K., B.B.; Performed the literature review: E.S. All authors contributed to the final manuscript.

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