

# A Rare Case of Single Coronary Artery with Atherosclerotic Lesions Arising from the Right Sinus of Valsalva

Florian Blaschke, Florian Krackhardt, Bherous Kherad, Burkert Pieske, Wilhelm Haverkamp, Matthias Rief<sup>1</sup>

Departments of Cardiology, Campus Virchow Klinikum, and <sup>1</sup>Radiology, Campus Mitte, Charité - University Hospital, Berlin, Germany

## Abstract

**Context:** Congenital coronary anomalies, including anomalous origin, distribution, intercoronary communications, and coronary fistulae occur at a rate of approximately 1% in the general population and are the most incidental findings. **Case Report:** A 49-year-old male patient presented to the emergency department with exercise-induced dyspnea and atypical angina pectoris. Coronary angiography (CAG) and contrast-enhanced 320-slice multidetector cardiac computed tomography with subsequent three-dimensional reconstructions revealed a single coronary artery (SCA) arising from the right sinus of Valsalva with a proximal branch giving rise to the left anterior descending coronary artery. The left anterior descending coronary artery shows severe atherosclerotic lesions and it is occluded afterwards. Adenosine stress perfusion cardiac magnetic resonance imaging (MRI) revealed a stress myocardial ischemia at the anterior wall without signs of fibrosis, scar, or necrosis. **Conclusion:** We present an extremely rare case of a SCA, with the solitary vessel arising from the right sinus of Valsalva. In our patient's case, the atherosclerotic lesions and occlusion in the branch supplying the anterior wall were considered eligible for neither percutaneous intervention nor bypass graft surgery.

**Keywords:** Cardiac magnetic resonance imaging (MRI), congenital coronary anomalies, coronary heart disease, multidetector cardiac computed tomography, single coronary artery (SCA)

**Address for correspondence:** Dr. Florian Blaschke, Department of Cardiology, Campus Virchow-Klinikum, Charité - Universitaetsmedizin Berlin, Augustenburger Platz 1, Berlin - 13353, Germany. E-mail: florian.blaschke@charite.de

## Introduction

Congenital coronary anomalies, including anomalous origin, distribution, intercoronary communications, and coronary fistulae, are relatively common, occurring at a rate of approximately 1% in the general population.<sup>[1]</sup> However, a single coronary artery (SCA), defined as a single aortic orifice, providing blood supply to the entire heart, is an extremely rare congenital anomaly with an incidence of only 0.024-0.066% in the general population.<sup>[1-3]</sup> SCA has been reported in association with and without atherosclerotic lesions or in association

with other congenital anomalies such as coronary arteriovenous fistulas, bicuspid aortic valve, and hypertrophic cardiomyopathy.<sup>[4-6]</sup>

## Case Presentation

A 49-year-old male presented to the emergency department with exercise-induced dyspnea and atypical angina pectoris. He reported a history of reduced exercise

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capacity since 5 months. On physical examination, his blood pressure was 135/85 mmHg and his heart rate was 82 beats per min. A resting 12-lead electrocardiogram (ECG) displayed sinus rhythm without any ST-segment abnormalities and a complete right bundle branch block [Figure 1a]. Transthoracic echocardiography showed normal biventricular function (left ventricular ejection fraction by biplane Simpson's was 60%) without regional wall motion abnormalities. Mitral valve insufficiency (grade I) and mild aortal and tricuspid regurgitation was noted without signs of increased pulmonary artery pressure. Laboratory testing revealed slightly increased C-reactive protein levels (14.7 mg/L). Cardiac enzymes (troponin T and creatin kinase) and other routine biochemistry test values were within the normal range. His cardiovascular risk-factor profile included untreated hypertension, hyperlipidemia, and a history of cigarette smoking (60 pack years). His family history was negative for heart disease.

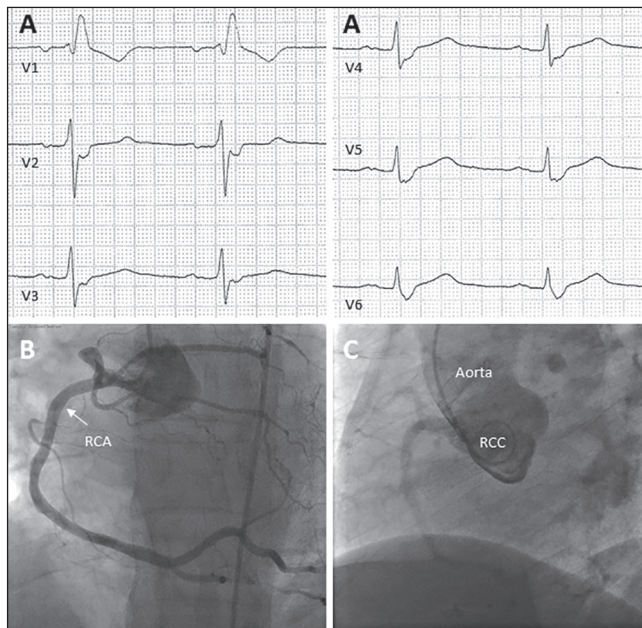
The patient underwent coronary angiography (CAG) that revealed an SCA originating from the right sinus of Valsalva [Figure 1b and c]. An aortic root angiography was performed to rule out the presence of any other coronary ostia [Figure 1c]. Contrast-enhanced 320-row multidetector cardiac computed tomography with subsequent three-dimensional reconstructions confirmed the diagnosis of a solitary vessel arising from the right sinus of Valsalva. The single vessel courses within the

right atrioventricular groove and shows mild luminal irregularities. The left anterior descending (LAD) coronary artery, which emerges as a proximal branch from the solitary vessel, shows significant atherosclerotic lesions and is occluded in the course [Figure 2]. Adenosine stress and rest perfusion cardiac magnetic resonance imaging (MRI) revealed stress-induced perfusion defect at the anterior wall. No significant delayed enhancement as evidence of myocardial fibrosis, scar, or necrosis was observed.

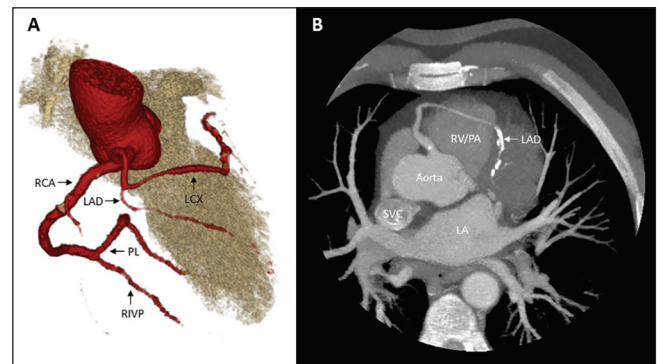
Treatment options such as coronary angioplasty with stenting or coronary artery bypass graft surgery were discussed within the Heart Team. The Heart Team considered the atherosclerotic lesions and occlusion in the branch supplying the anterior wall eligible for neither percutaneous intervention nor bypass graft surgery. Thus, the patient was scheduled for intensified medical therapy to which the patient consented. The patient was treated with acetylsalicylic acid (ASS), angiotensin II type 1 (AT1) receptor blocker, diuretic hydrochlorothiazide, statins, and nitrates and routine cardiac checkups were recommended once in a year.

## Discussion

We present an extremely rare case of an SCA, with the solitary vessel arising from the right sinus of Valsalva.



**Figure 1:** (a) Electrocardiogram (50 mm/s) showing normal sinus rhythm without any ST-segment abnormalities and a complete right bundle branch block. (b) Coronary angiography showing a single coronary artery arising from the right sinus of Valsalva. (c) Aortography in the left anterior oblique view ruled out the presence of any other coronary ostia. RCC: Right coronary cusp. RCA: Right coronary artery



**Figure 2:** (a) Cardiac computed tomography angiography showing the rise of the single coronary artery from the right sinus of Valsalva (\*) with proximal run of the right coronary artery (RCA). The distal parts of the RCA are not shown in this projection. The single coronary artery crosses the right ventricle and the root of the pulmonary artery (RV/PA) anteriorly and gives rise to the heavily calcified left anterior descending (LAD) coronary artery and further the left circumflex coronary artery (LCX). Furthermore, the left atrium (LA) and the superior vena cava (SVC) are shown. (b) Volume rendering reconstruction of contrast-enhanced 320-row multidetector cardiac computed tomography depicting the single coronary artery. The RCA gives rise to a branch that runs anterior to the pulmonary artery and supplies the LAD and LCX. Furthermore, the Ramus interventricularis posterior (RIVP) and a posterolateral branch (PL) originate from the RCA. PV: Pulmonary vein

Lipton *et al.*<sup>[2]</sup> classified the SCA in nine patterns according to the origin, anatomical course, and termination of the anomalous vessel. The case presented here enters into the R-IIA pattern, where R implies that the ostium is located in the right sinus of Valsalva. SCA is commonly associated with other congenital anomalies, such as transposition of the great vessels, coronary arteriovenous fistula, or bicuspid aortic valve. However, in our depicted case no other associated coronary anomalies were found. The spectrum of clinical manifestations of coronary anomalies is very broad from clinically benign to severe symptoms including arrhythmias, syncope, myocardial infarction, or sudden death. Invasive CAG is considered an adequate technique for the diagnosis of coronary anomalies. However, the conventional CAG is technically limited to two-dimensional projections and may not be sufficient to classify the anomaly correctly and delineate the precise course of the anomalous vessel. Thus, noninvasive techniques, such as computed tomography or magnetic resonance CAG, which offer three-dimensional reconstructions are necessary to visualize the coronary anatomy accurately.<sup>[7,8]</sup>

The greatest clinical challenge presented by coronary anomalies is the decision about the treatment. Functional tests such as cardiac MRI or stress echocardiography to detect effort-induced ischemia and myocardial scars are helpful to direct the appropriate treatment. According to the American College of Cardiology/American Heart Association guideline, a surgical revascularization of coronary anomalies with a documented ischemia is recommended.<sup>[9]</sup> However, previous studies revealed that standard clinical stress tests often fail to detect myocardial ischemia resulting from coronary anomalies.<sup>[10]</sup>

In our patient's case, adenosine stress and rest perfusion cardiac MRI revealed stress induced perfusion defect at the anterior wall. However, as the atherosclerotic lesions and occlusion in the branch supplying the anterior wall were considered eligible for neither percutaneous intervention nor bypass graft surgery, the patient was scheduled for intensified medical therapy

and routine cardiac checkups were recommended once in a year.

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### Conflicts of interest

There are no conflicts of interest.

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