

## Anomalous Right Coronary Artery and Sudden Cardiac Death

Brian Greet, MD; Adriana Quinones, MD; Monvadi Srichai, MD; Sripal Bangalore, MD; Robert O. Roswell, MD

### Introduction

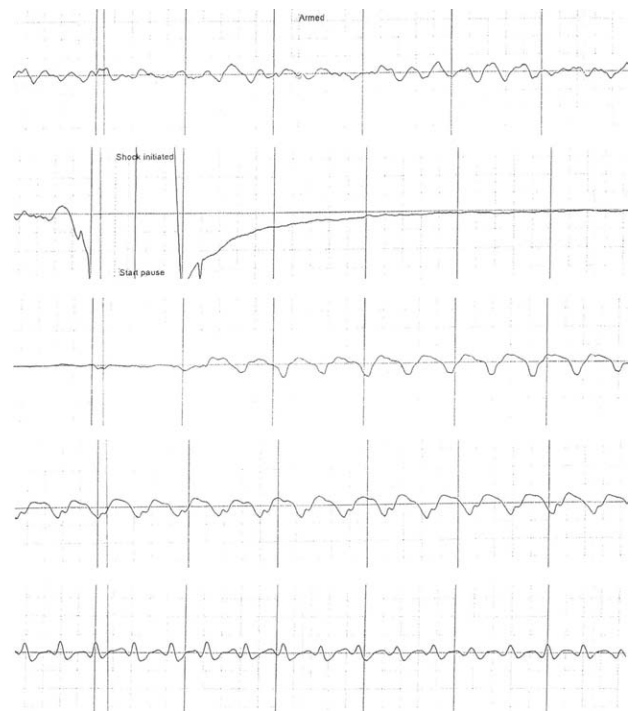
Sudden cardiac death (SCD) in association with anomalous coronary arteries is a rare phenomenon. When described it is most often associated in patients with anomalous left coronary arteries, although anomalous right coronary arteries are incidentally found more frequently.<sup>1</sup> Current theories posit ischemia as the etiology of SCD in these patients. The proposed mechanisms of ischemia induced SCD are as follows: (1) acute angulation at the ostium of the coronary artery and kinking; (2) an abnormal slit-like opening; (3) mechanical compression of the anomalous artery if it courses between the aorta and pulmonary artery during exertion; and (4) vasospasm of the anomalous artery.<sup>2</sup>

### Case Report

A 37-year-old woman with a history of 2 uneventful vaginal deliveries, 2 and 5 years before presentation, experienced a witnessed collapse while in a train station. The patient was found pulseless, and cardiopulmonary resuscitative measures were initiated immediately by bystanders. An automated external defibrillator was applied, and the patient eventually received 1 shock via the automated external defibrillator (Figure 1). Analysis of the automated external defibrillator showed ventricular fibrillation that converted to normal sinus rhythm with elevation of the ST segment with defibrillation and then resolution of the ST segment elevations.

The electrocardiogram obtained on initial hospitalization revealed normal sinus rhythm without significant ischemic changes. Transthoracic echocardiogram initially revealed diffuse left ventricular hypokinesis with a depressed ejection fraction that, on repeat 48 hours later, had completely normalized. Peak troponin I was measured at 2.154 ng/mL. Coronary angiography was performed and revealed no obstructive lesions but revealed an anomalous origin of the right coronary artery (RCA), arising from the left coronary cusp (online-only Data Supplement Movie I).

Further imaging with cardiac computed tomography angiography (Figure 2) confirmed an anomalous origin of the RCA, arising from the left cusp via a separate ostium off of the left coronary cusp. In addition, the RCA had an acute angulation



**Figure 1.** Telemetry recordings from the automated external defibrillator.

with a slit-like opening at the ostium measuring 4×1 mm. The RCA demonstrated an interarterial proximal course between the pulmonary artery and aorta. Further workup as to the etiology of her arrest, including cardiac magnetic resonance imaging and pulmonary artery computed tomography angiography, was unrevealing. The patient unfortunately did not have a favorable neurological outcome and was palliatively extubated and discharged to hospice where she expired 2 weeks later.

### Discussion

The incidence of anomalous origin of the RCA arising from the left coronary cusp that courses between the great vessels varies between 0.026% and 0.250%. Three subtypes of

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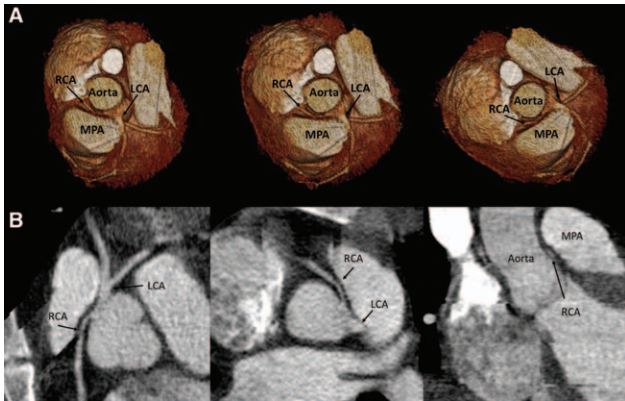
The online-only Data Supplement is available at <http://circep.ahajournals.org/lookup/suppl/doi:10.1161/CIRCEP.112.978635/-/DC1>.

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**Figure 2.** **A**, Coronary artery computed tomography (CT) angiography with 3-dimensional reconstruction demonstrating the anomalous origin of the right coronary artery from the left coronary cusp. **B**, Multiplanar and curved multiplanar reformatted view again showing the anomalous course of the right coronary artery between the aorta and main pulmonary artery. RCA indicates right coronary artery; LCA, left coronary artery; and MPA, main pulmonary artery.

anomalous RCA have been described: (1) a high interarterial course between the pulmonary artery and the aorta; (2) a low interarterial course between the right ventricular outflow tract and the aorta; and (3) a hypoplastic anomalous RCA orifice. An anomalous RCA with a high interarterial course between the pulmonary artery and aorta is at risk for being compressed during exercise or routine activities that can cause angina, malignant arrhythmias, and SCD. Exercise nuclear stress testing revealed reversible defects consistent with ischemia in a patient with an anomalous RCA coursing between the aorta and pulmonary artery with episodic chest pain.<sup>3</sup>

In young athletes with anomalous coronary arteries and SCD, it is not uncommon to find that patients experienced chest pain or syncope before the sentinel event. Routine testing with resting or exercise electrocardiography is not sufficiently sensitive to rule out this congenital anomaly, nor can it predict the likelihood of SCD.<sup>2</sup>

The timing of symptoms is also unpredictable, and the first symptom of this rare disorder may be SCD. This patient was

asymptomatic and successfully delivered 2 children vaginally 5 and 2 years before presentation. Aortic dimensions increase with age and correlate well with height, weight, and body surface area.<sup>4</sup> When an anomalous artery courses between the great vessels, the onset of symptoms probably occurs when the ischemic threshold has been surpassed. This is likely to occur when the increasing size of either the aorta or pulmonary artery combines with an increased adrenergic surge to produce mechanical compression of the anomalous coronary artery. The challenge for providers is to predict when that ischemic threshold will be met to offer potentially lifesaving treatments, such as surgical correction. Anomalous coronary arteries can be surgically corrected by bypass grafting, reimplantation of the artery to its proper coronary sinus, or by an unroofing procedure.<sup>3</sup> Weighing the risks and benefits of a surgical procedure for this rare disorder remains challenging.

This case is the first to demonstrate the exact mechanism of SCD in patients with this rare coronary anomaly. The evidence presented here lends credence to the hypothesis that transient ischemia, in humans with anomalous coronary arteries, leads to malignant arrhythmias causing SCD.

## Disclosure

None.

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