

# ANOMALOUS ORIGIN OF RIGHT CORONARY ARTERY

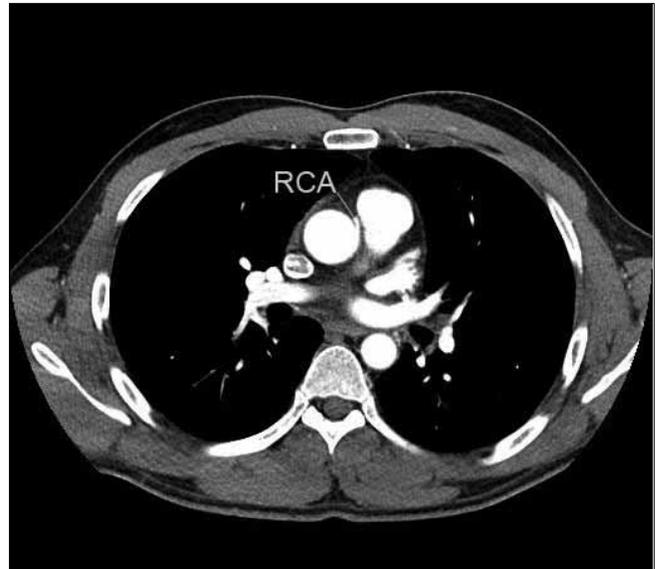
Neerav G. Sheth MD, Siva K. Kumar, MD

## Introduction

Anomalous coronary arteries are rare but potentially life threatening abnormalities of the coronary circulation. Many of the variations can be benign in nature; however, some may lead to myocardial ischemia and/or sudden cardiac arrest.<sup>1</sup> The most common anomalies described in the medical literature involve branches of the left coronary artery, with the left circumflex artery originating from the right sinus of Valsalva being the most prevalent. Patients may present at any age with symptoms ranging from syncope to sudden cardiac death. Here we present a case of a patient with anomalous right coronary artery circulation (RCA) who presented with atypical chest pain.

## Case Report

A 38-year-old African American male with a past medical history of hypertension, COPD, and cocaine abuse presented with a three day history of atypical chest pain. Initial ECG showed Normal Sinus Rhythm, non-specific ST changes with premature ventricular contraction and a positive urine drug screen for cocaine metabolites (Figure 1). He underwent a Cardiac CTA from emergency room which revealed an anomalous origin of the RCA approximately 8mm above the sinotubular junction above the left coronary sinus of Valsalva. Furthermore, the proximal portion of the vessel traversed between the aorta and right ventricular outflow tracts (Figure 2). Given the patient's anomalous RCA course, cardiothoracic surgery was consulted for surgical repair of the anomalous vessel. The rationale for surgical repair is based on a significantly increased risk for sudden cardiac arrest and myocardial infarction. His particular anatomy places him at an extremely high risk during exercise secondary to dilatation of the pulmonary artery and aorta resulting in compression of the proximal portion of the anomalous vessel thereby inducing ischemia and ventricular arrhythmia leading

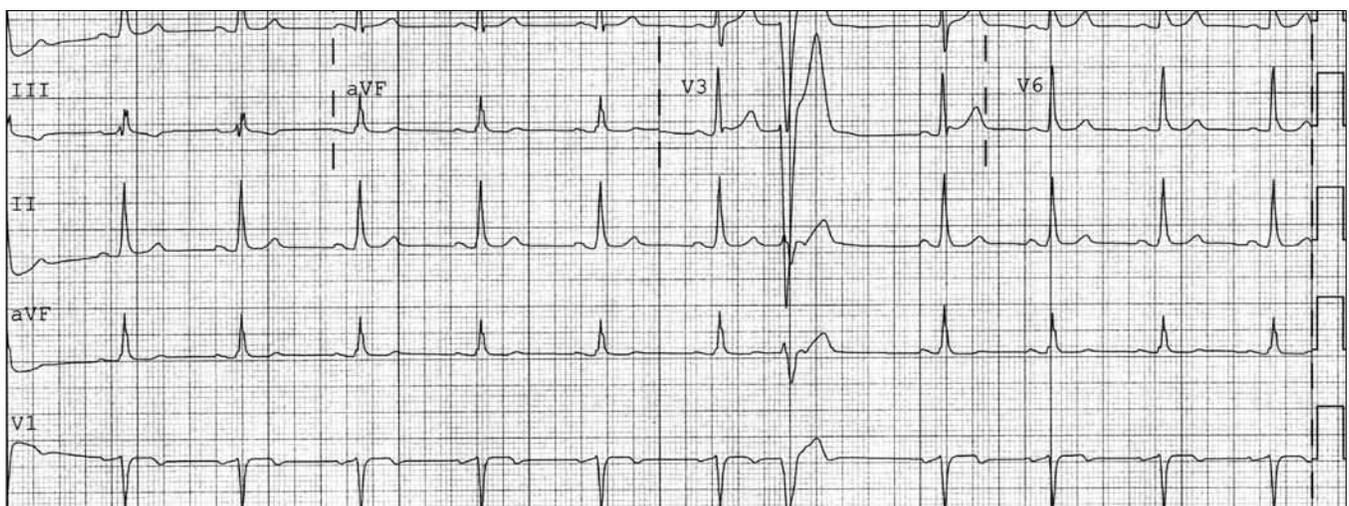


**Figure 2.** Cardiac CTA showing the anomalous RCA coursing between the aorta and pulmonary outflow tracts

to sudden death. The patient was taken to the operating room for a one-vessel saphenous vein aortocoronary bypass to the RCA at the level of the posterior descending artery (PDA).

## Discussion

Splanchnic mesoderm gives rise to all component of a normal heart. The mesoderm differentiates into the cardiogenic area that occurs during week 3 of embryogenesis. The cardiogenic area subsequently forms a pair of endocardial tubes which fuse to form the primitive heart tube.<sup>2</sup> Normal coronary arteries arise from appropriate differentiation of pluripotent cells into their



**Figure 1.** ECG on initial presentation.

respective anatomic and functional components. Anomalies of the coronary circulation result from processes that disrupt the normal differentiation and specialization of the primitive heart tube.<sup>3</sup> In particular, abnormal involution, position of endothelial buds, or septation of the truncus arteriosus may give rise to anomalous origin of coronary arteries.<sup>4</sup>

In general, anomalous coronary arteries can be described as those able to cause interruptions in coronary blood flow or significant or major anomalies, and those that do not, also known as nonsignificant or minor anomalies. Significant anomalies are exceedingly rare, but are responsible for 0.25% - 0.9% of congenital malformations.<sup>2,5</sup>

With respect to our case, anomalous origin of the right coronary artery from the left coronary sinus of Valsalva occurs with an incidence of 0.05% to 0.1% in the general population.<sup>6</sup> While anomalous coronary arteries occur with low frequency, there is a high risk of sudden death due to myocardial ischemia and resultant arrhythmia associated with them.<sup>7</sup> Various mechanisms have been postulated to cause the aforementioned ischemia including: Origin in an acute angle and folding or occlusion caused by the angulation at the point of coronary artery emergence,<sup>8</sup> coronary spasm resulting from its torsion movement,<sup>8</sup> mechanical compression of the anomalous artery between the pulmonary and aortic trunks during physical exertion,<sup>5</sup> and an intramural origin of the coronary artery from within the aortic tunica media.<sup>8</sup> The majority of these complications may be exacerbated during or immediately after exercise, as exercise leads to compression of coronary arteries as well as increasing the preexisting angulation of the proximal portion of the anomalous vessel.<sup>9</sup>

Clinical presentation of these patients ranges from asymptomatic for non-significant anomalies to syncope, chest pain, myocardial infarction and sudden cardiac death in significant anomalies. Diagnosis of anomalous coronary arteries may be suggested by echocardiography and a high index of suspicion. Depending on the course of the anomalous vessel, a nuclear stress test may reveal regional hypoperfusion abnormalities in the myocardium supplied by the vessel.<sup>15</sup> In the current era, there is also a role for Cardiac CTA (CCTA) and Cardiac MRA (CMRA) as non-invasive diagnostic modalities prior to or in lieu of coronary angiography. A few small studies have shown CMRA to show a high degree of correlation to cardiac catheterization<sup>10</sup> and have a sensitivity of 88% and a specificity of 100% as compared with traditional coronary angiography.<sup>11</sup> Similarly, multi-detector row CCTA has also been shown to accurately depict anomalous origin of coronary arteries in those with equivocal findings at cardiac catheterization or echocardiography.<sup>12</sup>

Treatment of significant anomalies should be guided by the nature of the anomalous vessel. The vessel may need surgical reconstruction, decompression, reimplantation in the correct sinus, or bypass with ligation of the native vessel to eliminate

competitive flow in the graft.<sup>13,14</sup> Particularly, if a vessel courses between the aorta and pulmonary trunks, has a narrow ostium, or arises from the pulmonary vasculature, it is highly susceptible to repeated episodes of myocardial ischemia. These patients would benefit from surgical intervention. Those with asymptomatic or non-significant anomalous coronary arteries may be observed and managed clinically.

Our patient did extremely well following a single vessel aortico-coronary bypass grafting to the RCA at the level of the PDA. He had complete resolution of his presenting chest pain upon completion of the bypass. He was medically optimized prior to discharge and had no further symptoms at follow up.

## References

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