



# High-Risk Coronary Artery Anomalies, the Second Most Common Cause of Sudden Cardiac Death in Young People: A Case Study

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## Abstract

It is our intention to present an atypical case of a high-risk coronary artery anomaly to draw awareness and reduce mortality from Sudden Cardiac Death (SCD) syndrome. SCD is the second most common cause of high risk coronary anomalies in young people and nearly half a million lives are taken per year from this condition.

We present a young, sedentary, symptomatic 34-year-old male patient who presented to his Primary Care Physician (PCP) with complaints of worsening chest pain. He is a non-smoker and denied alcohol or drug use but had strong family history of premature coronary artery disease on both his maternal and paternal sides. This case is impactful because it shows a non-athlete profile which is an outlier to the majority of literature which tends to portray only athletes and highly active patients.

**Keywords:** Coronary artery anomaly; Sudden cardiac death; Sudden cardiac arrest

## Introduction

Anatomical variations of the coronary arteries can range from fistulas to degrees of angulation to sites of origin [1]. While most of these anomalies do not pose a significant functional threat, there are a few that have been known to increase risk of death [2]. Providers should be concerned about these high-risk situations known to cause arrhythmias, heart attacks and sudden cardiac death. We will identify those concerning aberrations, understand the population at risk, identify how they present and make recommendations.

## Methods

Chart review of case study patient. Literature review: searched “coronary artery anomalies”, “sudden cardiac death/arrest”, and “malignant coronary artery anatomy”.

## Case Presentation

34-year old male patient presented to his Primary Care Physician (PCP) with complaints of worsening chest pain. He is a non-smoker and denied alcohol or drug use but had strong family history of premature coronary artery disease on both his maternal and paternal sides. His Body Mass Index (BMI) was 36. Electrocardiogram (EKG) showed normal sinus rhythm, normal axis and Interventricular Conduction Delay (IVCD) (Figure 1).

He had a history of essential hypertension and had been recently started on blood pressure lowering medication Hydrochlorothiazide (HCTZ) and cholesterol lowering medication Crestor. Calcium score was ordered and found to be 291, with greatest contribution coming from the right coronary artery, placing him at 90<sup>th</sup> percentile on MESA scale. See calcium score distribution in Figure 2.

Two-dimensional (2D) echocardiogram was done and showed mild concentric left ventricular hypertrophy with normal Ejection Fractions (EF) of 60% to 65%. No valvular abnormalities were detected (Figure 3).

Nuclear Medicine (NM) stress test and Single Positron Emission Computerized Test (SPECT)

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Figure 1: EKG taken at patient's PCP office, showcasing an IVCD.

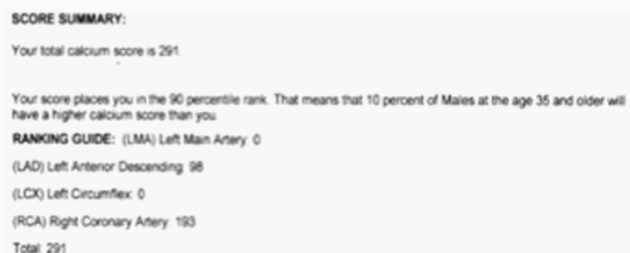


Figure 2: Patient's calcium score distribution.



Figure 3: Patient echocardiogram showcasing normal EF and normal valves.

was ordered and showed appropriate response with exercise. Patient walked 11 min and 30 sec. Metabolic Equivalents (METs) were achieved at 12.8, double work product 35,720 target heart rate achieved at 102, demonstrating normal myocardial perfusion. Computed Tomography Angiography (CTA) of coronary arteries done next with main finding of Right Coronary Artery (RCA) anomalous origin from left coronary cusp between the intra arterial course between the aorta and pulmonary artery in which compression of the proximal RCA occurred. Proximal RCA showed a calcified plaque with mild Stenosis. LAD showed 25% to 49% lesions (Figure 4).

This coronary anomalous finding prompted a referral to a cardiothoracic surgeon and a Left Heart Catheterization (LHC) was done. LHC found RCA anomaly from the left coronary cusp which was identified as the dominant artery. Multivessel disease also confirmed (LAD 75% blockage, first and second obtuse marginal 85% blockage, and RCA 80% blockage). Recommendations for bypass were made (Figure 5).

## Results and Discussion

Coronary artery anomalies have been incidentally identified in 5% of cardiac catheterizations [3]. All do not increase risk or cause

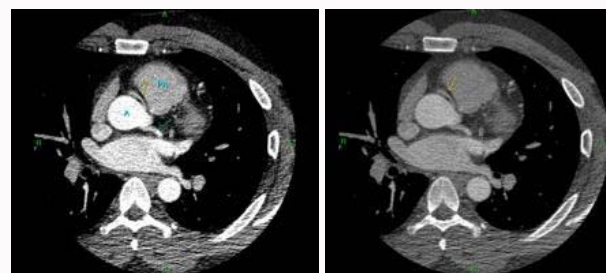


Figure 4.1 and 4.2: CTA Coronary Arteries showcasing the anomalous origin of the right coronary artery (yellow arrow) from the left aortic cusp and coursing between the aorta and pulmonary artery. Left coronary artery is visible as well (blue arrow in first image).



Figure 5.1 and 5.2: Patient Left Heart Catheterization (LHC) study stills. RCA can be seen in the left most side of stills, originating from the left cusp. Second image shows compression of RCA at origin point (scoping at beginning of vessel) between the aorta and pulmonary artery.

symptoms to the patient. However, any coronary anomaly that reduces supply of oxygen to the myocardial tissues is considered high-risk, causing ischemia and Sudden Cardiac Death or Arrest (SCD/ SDA) [4]. These are the top concerns especially in young patients less than 30 years of age.

The National Heart, Lung, and Blood Institute reports that nearly a quarter to almost half a million US citizens die of SCD every year. Generally, these cases result in death within minutes [3]. The most common cause of SCD in young athletes is hypertrophic cardiomyopathy. However, we draw attention to the second most common cause of SCD in young persons, which are high-risk coronary artery anomalies [5]. In fact, over 30% of SCD in young people with exercise have been attributed to coronary artery anomalies post mortem. Symptoms are not always present. Syncope episodes during exercise are considered severe with milder symptoms being shortness of breath, chest pain and fatigue [3]. Most studies tend to exclude non-athletes and patients with low physical activity, but our case give basis to draw concern in non-athletes that show mild symptoms and/or mention family history.

The concerning coronary anomalies every provider should know are: 1) those which originate from the pulmonary artery or from the opposite aortic sinus, thus traversing between the pulmonary artery and aorta 2) a solitary coronary artery and 3) an enlarged coronary fistula. For the first type, when the anomalous coronary arteries run between the two great vessels and engorgement occurs, most often during exercise, compression can occur. This compression causes ischemia which then causes sudden cardiac death [6]. The second type can also cause oxygen demand issues and the third type can be a site for stasis.

Those at highest risk are 30 years of age or younger and are

physically active. Inactive individuals are still at high risk however. Those with a comparable anomaly over 30 years of age are considered at reduced risk, but still higher than the average population for SCD [3].

Literature review appears to point to poor work-ups and missed anomaly diagnoses [7]. Currently no screening guidelines and low awareness means providers are possibly overlooking symptoms, especially in inactive patients. Also, our assessment has led us to question whether high-risk coronary anomalies are more common than currently reported [8]. One study of 242 autopsies found 142 (59%) cardiac deaths resulted from coronary anomalies and of these, 78 (32%) were SCD, of which 64% were exercise-related deaths with minimal symptoms [9]. This highlights a discrepancy between reported prevalence and actual prevalence.

Treatment for symptomatic or asymptomatic patients is the same for those with known high-risk coronary anomalies. Treatment includes lifestyle modification, pharmaceutical management, Percutaneous Coronary Interventions (PCI), and surgery. For lifestyle modification, it is recommended that the patient avoid strenuous activity to avoid great vessel engorgement and subsequent compression on the coronary artery. Pharmaceutical management is recommended for symptomatic relief and prevention of SCD. Recommendations are to slow the heart with beta-blockers, reduce excess fluid with furosemide, ensure a regular rate and provide oxygen therapy. PCI with stent placement has been successful in some coronary anomalies but this is anomaly type-dependent. Surgery is the most invasive option but is often the best option. Limited studies exist on the best surgical techniques [3].

## Conclusion

Increased awareness in patients and providers as well as a high-suspicion screening protocols and at times tighter surgical correction

guidelines are needed to effectively prevent SCD in young patients [10]. Earlier detection at the primary care level and earlier referral to cardiology are highly recommended.

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