

# IMAGES in PAEDIATRIC CARDIOLOGY

**Abbas U<sup>1</sup>, Cossor W<sup>2</sup>. Anomalous Origin of Left Coronary Artery – an unusual case. *Images Paediatr Cardiol* 2018;20(2):1-4.**

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## **Abstract**

Anomalies of the coronary arteries are rare but are an important cause of sudden cardiac arrest in young athletes. Sudden cardiac arrest has been reported in patients with congenital anomalies of the coronary arteries. We present a rare case of sudden cardiac arrest caused by anomalous left main coronary artery originating high from the posterior aspect of left sinus of Valsalva with intramural retroaortic segment, associated with bicuspid aortic valve.

## **Introduction**

Anomalies of the coronary arteries are a rare but important cause of sudden cardiac arrest in young athletes. Coronary anomalies are most often classified into abnormalities of origin, distribution, and association with fistulae. About 80% of coronary anomalies are benign and incidental findings.<sup>1</sup> A study by Yamanaka and Hobbs in 1990 found the overall incidence of coronary artery anomalies in more than 120,000 patients undergoing coronary angiography to be 1.3%.<sup>2</sup> In a retrospective review of Department of Defence data from 1977 to 2001, nontraumatic sudden death occurred in 126 military recruits out of over 6 million cases reviewed. Of these patients, the cause of death was identified as a coronary artery anomaly in 39 patients, with 21 of those having an anomalous origination of the left coronary artery from the right sinus of Valsalva, with an interarterial course between the pulmonary artery and aorta.<sup>3-4</sup>

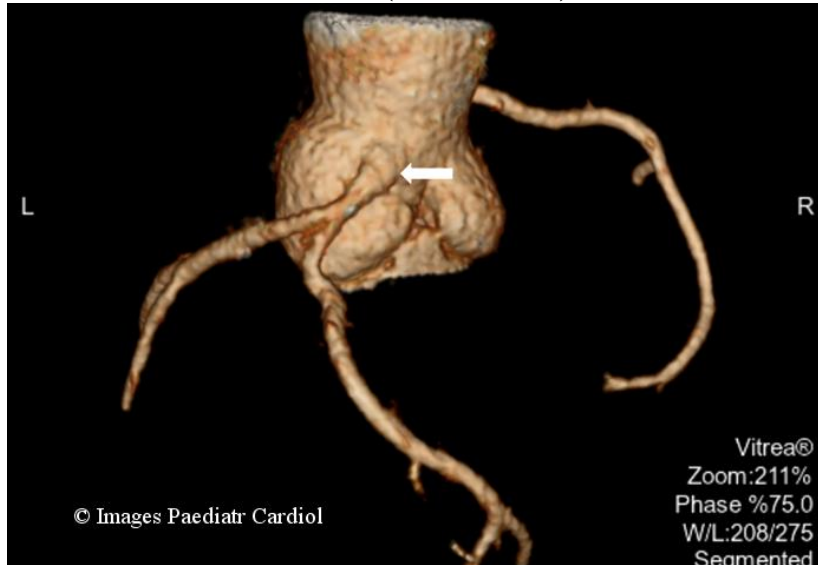
We present a rare case of sudden cardiac arrest caused by anomalous left main coronary artery originating high from the posterior aspect of the left sinus of Valsalva, with intramural retroaortic segment, occurring in association with a bicuspid aortic valve.

## **Patient**

A 16 year old male with congenial bicuspid aortic valve and mild valvar aortic stenosis was transferred from an outside hospital for cardiac evaluation following aborted sudden cardiac death (SCD). The patient had been playing water polo when he lost consciousness in the water and became cyanotic. An AED on site showed polymorphic ventricular tachycardia (Torsade de pointes). Shock was advised and delivered and the patient regained consciousness.

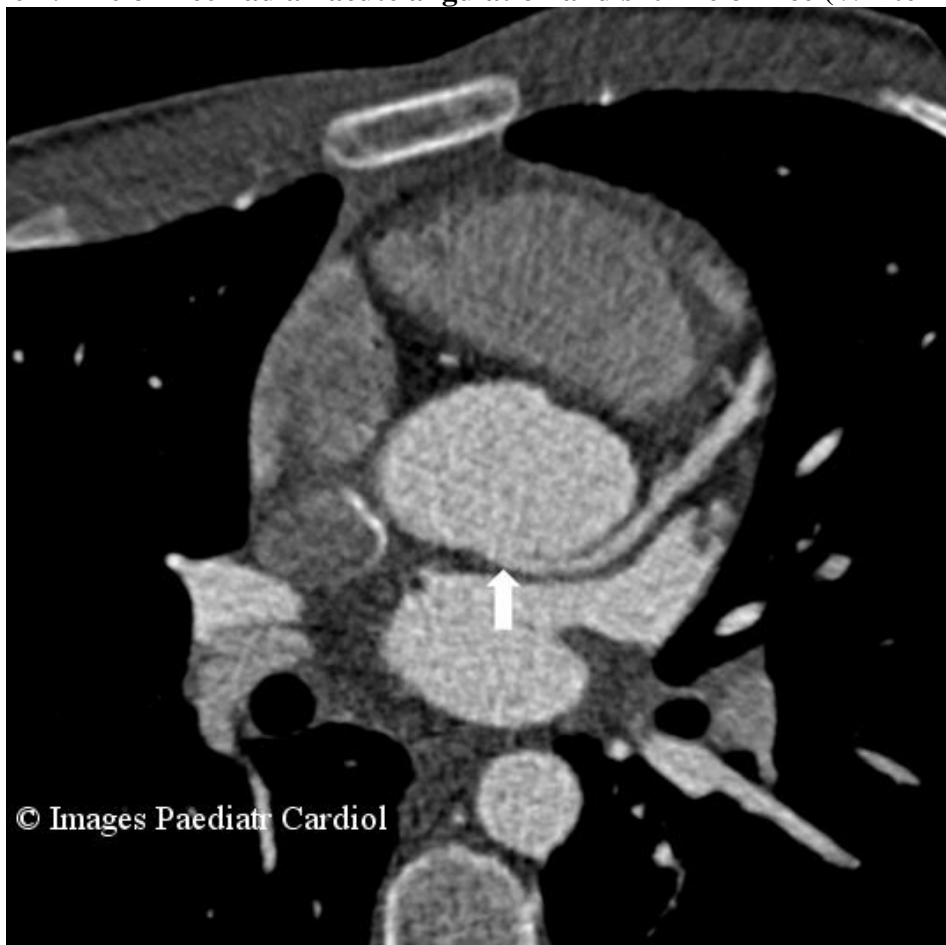
CT angiography showed the left main coronary artery arising high from the posterior aspect of the left sinus of Valsalva at the level of the sinotubular junction (figure 1).

**Figure 1: Anomalous left coronary artery originating from the posterior aspect of left sinus of Valsalva (white arrow).**



The orifice had an acute angulation and slit-like orifice (figure 2). There was an intramural retroaortic segment which coursed leftward and toward the annulus, before exiting the aortic wall and branching into the left main and circumflex arteries.

**Figure 2: The orifice had an acute angulation and slit like orifice (White Arrow)**



The proximal segment had an ellipsoid cross-section typical of intramural coronaries (figure 3). The left main and circumflex coronary arteries had no evidence of stenosis, and normal branching patterns. The right coronary artery originated normally from the right sinus of Valsalva and coursed normally into the right atrioventricular groove with no stenosis. A bicuspid aortic valve with fusion of the right and noncoronary leaflets was noted.

**Figure 3: Left main coronary artery cross section. Ellipsoid cross-section typical of intramural coronaries (White Arrow)**



Cardiac MRI demonstrated normal biventricular systolic function. Left ventricular ejection fraction was 62% and right ventricular ejection fraction was 52%. There was no evidence of myocardial scar or fibrosis.

The patient underwent complete unroofing of the left main coronary artery, aortic commisurotomy and implantable cardiac defibrillator placement. The postoperative course was uncomplicated. Echocardiography on postoperative day three showed normal cardiac function.

### **Conclusion**

We present a rare case of left coronary artery originating high from the posterior aspect of left sinus of Valsalva with intramural retroaortic segment. Congenital coronary artery anomalies remain an important cause of sudden cardiac death. The true prevalence of coronary artery anomalies in the general population is unknown. Surgery remains the mainstay of treatment.

## References

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