

## Monomorphic Ventricular Tachycardia as the First Manifestation in a Patient with Anomalous Coronary Artery

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### Introduction

Coronary artery anomalies comprise a heterogeneous group of rare congenital heart defects, being classified according to their origin, course and distal bed, with an incidence ranging between 0.3% and 1.5% in the overall population. Dodge-Khatami et al.<sup>1</sup> subdivided them into seven categories, according to their clinical complexity: coronary arteries originating from the pulmonary artery, coronary arteries with anomalous aortic origin, congenital atresia of the left main coronary artery, coronary arteriovenous fistulas, coronary arteries forming myocardial bridges, coronary artery aneurysms and coronary stenosis. The Texas Children's Hospital classification uses angiotomography with virtual angiography and divides the classification into three topics: origin of the anomalous coronary artery, coronary artery course and ostium morphology. From the viewpoint of anatomical risk, the anomalous left coronary artery with an interarterial course, presence of intramurality and a slit-like ostium are the main predictive factors of severity.<sup>1-3</sup>

A specific type is the anomalous origin of the left coronary artery from the right coronary sinus with an interarterial course, which is associated with hard outcomes in approximately 60% of the cases.<sup>2</sup> Clinically, patients may present with nonspecific symptoms,

ranging from palpitations, chest pain, post-exertion syncope or remain asymptomatic throughout life, with sudden death being the first and only manifestation of this condition.<sup>4</sup>

Most cases have been reported in young male individuals, but there is no scientific evidence yet whether the incidence is actually higher in males, or if this gender is more often diagnosed by performing more intense physical activities, therefore triggering symptoms.

The most common of these conditions is the anomalous origin of the left coronary artery from the pulmonary artery. However, we present herein a case with an anomalous origin of the left coronary artery from the right coronary sinus, with a proximal course between the aortic and the pulmonary arteries in a 31-year-old man.

### Case report

A previously healthy 31-year-old male patient, a mason, was admitted to the Emergency Unit in October 2012, complaining of palpitations and cold sweats, with hypotension (BP = 90 x 50 mmHg), which developed into syncope at the hospital unit. He was submitted to the first electrocardiogram (Image 1A), which disclosed sustained monomorphic ventricular tachycardia (SMVT). He underwent electrical cardioversion and was transferred to intensive care unit (ICU) for clinical stabilization.

Therapy with amiodarone was started in the ICU, with some episodes of slow ventricular tachycardia (VT) and periods of accelerated idioventricular rhythm (Image 1B), albeit asymptomatic. He reverted to sinus

### Keywords

Arrhythmias, Cardiac; Tachycardia, Ventricular; Coronary Vessel Anomalies; Death. Sudden, Cardiac.

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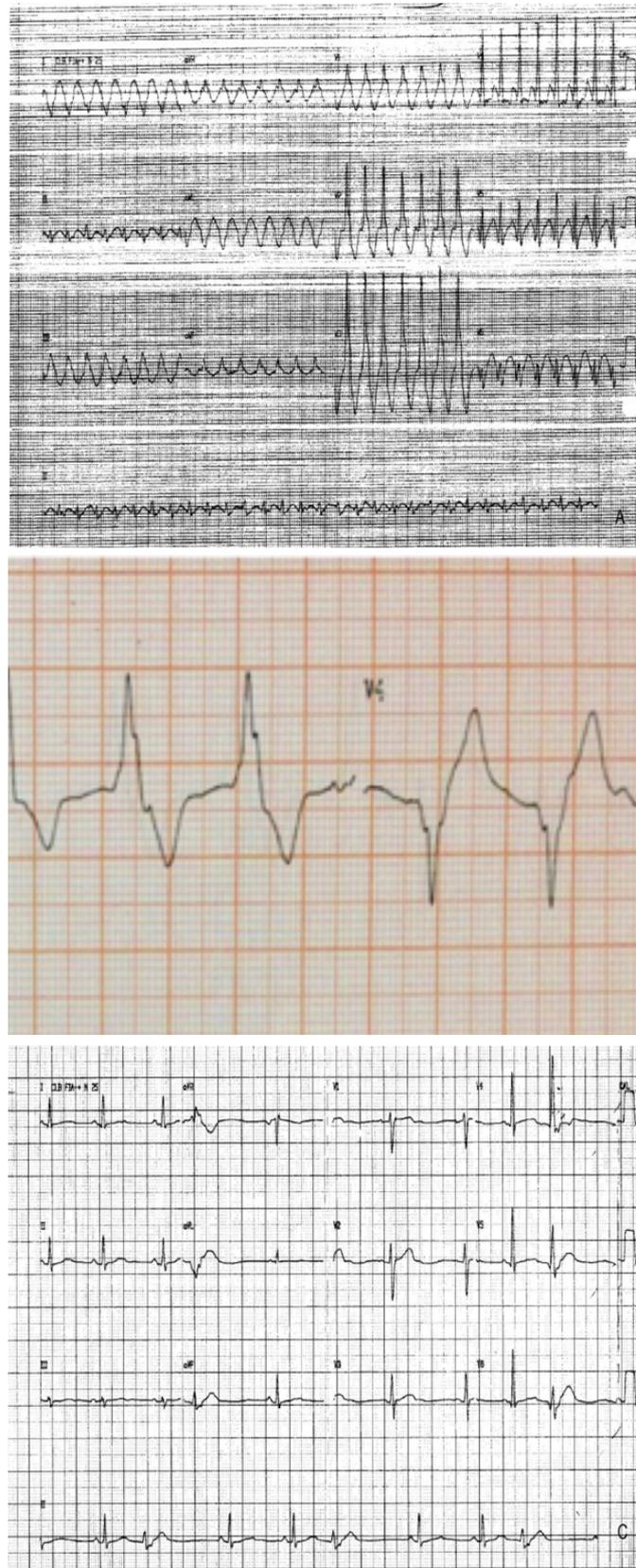


Figure 1 - A - Sustained monomorphic ventricular tachycardia. B - Accelerated idioventricular rhythm. C - Sinus rhythm with isolated ventricular extrasystoles.

rhythm (Image 1C) after 24 hours and the etiological investigation was initiated with a transthoracic echocardiogram, which disclosed only mild and diffuse left ventricular impairment. The exercise testing and magnetic resonance imaging of the heart showed no alterations, and serologic tests for Chagas' disease and cardiac markers were negative.

Acute myocarditis was suspected, which would have lead to the VT and a tachycardiomyopathy caused by the time he remained on sustained VT.

The patient received amiodarone, carvedilol, captopril and spironolactone, remaining asymptomatic and being followed at the outpatient clinic in the following months, without complications.

Ventricular function was normal at the transthoracic echocardiography and exercise testing at 3 and 6 months after the event, without arrhythmias or myocardial ischemia. As there was no symptom recurrence, even after the patient returned to his work routine, amiodarone was withdrawn.

However, without the antiarrhythmic drug, a new event was triggered in December 2014, which led the patient to once again seek the emergency care unit with palpitations and cold sweats. The VT was reverted again with electrical cardioversion, and the patient returned to the cardiology outpatient clinic for assessment. An angiotomography was then requested, which disclosed the anomalous origin of one of the coronary arteries (Figure 2). The diagnosis was confirmed after a coronary angiography, which concluded that the left coronary artery originated from the right coronary artery, coursing between the pulmonary artery and the aorta (interarterial course). With the anatomical definition of the condition, the surgical correction was chosen, using an internal mammary artery graft to the anterior descending and circumflex arteries, with the left main coronary artery ligation being successfully performed. The patient has been followed for 2 years, with no new episodes of VT and no medication.

## Discussion

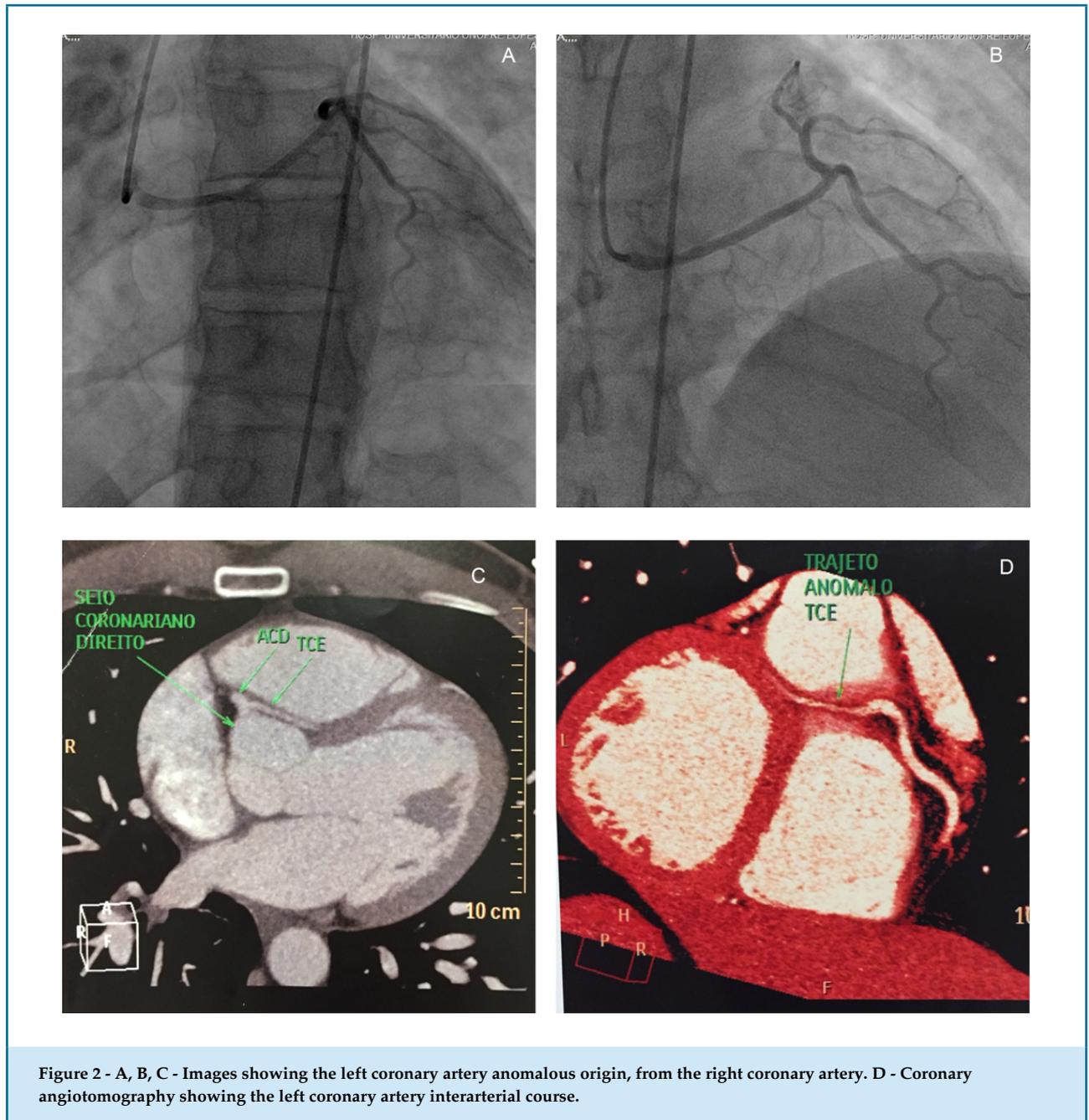
Ventricular arrhythmias comprehend a spectrum that range from ventricular extrasystoles, VT, to ventricular fibrillation. Ventricular fibrillation is the one most frequently associated with acute coronary syndrome. On the other hand, the SMVT has as its electrophysiological mechanism of re-entry related to scarring due to structural heart disease (e.g.,

previous infarction and Chagas' disease). In the younger population - children, adolescents, and young adults - with sustained episodes of VT, the condition is usually due to diseases that manifest early in life, including genetically-determined arrhythmias, acute myocarditis, and congenital heart disease – among the latter, coronary artery anomalies.<sup>5</sup>

Congenital anomalies of the coronary arteries may result in benign or malignant clinical consequences depending on their course and origin.<sup>2</sup> They are usually classified into four groups: according to the coronary origin and course (absent left main coronary artery, anomalous location of the coronary ostium inside or outside the appropriate Valsalva sinus, anomalous location of the coronary ostium in the inappropriate Valsalva sinus and single coronary artery); intrinsic to the coronary anatomy (stenosis or atresia of coronary ostia, coronary aneurysm, coronary hypoplasia and myocardial bridge); anomalies of terminal coronary circulation (fistulas into cardiac chambers, inferior vena cava or pulmonary arteries and veins); and anomalous anastomotic vessels.<sup>2</sup>

The anomaly described in this case is that of the left coronary artery originating from the right coronary sinus, a rare congenital abnormality, with a prevalence of 0.15% in the overall population.<sup>6</sup> This anomalous coronary artery is most commonly related to sudden death (59% of cases), usually preceded by physical activity (in 81% of these cases). The cases in which the coronary artery courses between the aorta and pulmonary (interarterial) arteries, as described in the reported patient, are the ones most often associated with severe outcomes.<sup>6</sup> One of the potential mechanisms that explains this fact is the coronary artery compression by the aorta and the pulmonary trunk during exercise, leading to myocardial ischemia.<sup>2,7,8</sup> When only the origin is anomalous, but the coronary artery does not follow this course, there is no risk of sudden death.<sup>4</sup> In general, the pre-pulmonary, subpulmonary or retroaortic courses are considered benign.<sup>3,4</sup>

In general, most patients remain asymptomatic or exhibit symptoms only after strenuous physical exercise, which may have a fatal outcome in these cases. Clinical presentation ranges from palpitations, dyspnea, chest pain and syncope to sudden death,<sup>4,6,7</sup> being the second leading cause of sudden death in young individuals.<sup>2,6</sup> Basso et al.<sup>8</sup> reported that only ten (36%) of the 27 cases with sudden death (23 anomalous left coronary arteries and four right coronary artery anomalies from the



opposite Valsalva sinus) had symptoms before the event, including syncope, chest pain and palpitations. All cases had an acute-angled outflow and a slit-like ostium.<sup>8</sup>

Due to the varied and nonspecific symptomatology, clinical suspicion and detailed investigation are necessary. Thus, a 12-lead electrocardiogram, exercise testing and an echocardiogram are suggested for the initial approach of symptomatic patients, which in some cases may suggest the diagnosis or disclose another cause for the symptoms.

Subsequently, a coronary angiogram and a coronary angiogram should be performed for diagnostic confirmation.<sup>7</sup> In cases of symptomatic patients, surgical revascularization is the therapeutic indication, especially when the left coronary artery originates from the opposite coronary sinus and courses between the aorta and the pulmonary artery, due to the risk of coronary compression by the larger-caliber vessels.<sup>4,9</sup> There is no consensus regarding the treatment of the anomalous anatomy with no evidence of ischemia or with an intramural course

or ostium anomaly.<sup>9</sup> The management tends to be more conservative, with the use of beta-blockers and changes in lifestyle, aiming to avoid strenuous physical exercises.

Therefore, the anomalous origin of the coronary artery is a group of rare congenital cardiac malformations with variable presentation. Due to the possibility of a lethal prognosis, it is necessary to identify the target population to establish screening methods to attain an early diagnosis of the anatomical alteration. Because it is more frequently associated with sudden death, a protocol should be established for young individuals practicing highly competitive sports or those subject to strenuous physical activities, aiming to prevent such an outcome. The diagnosis should always be recalled in cases of ventricular tachyarrhythmias<sup>10</sup> with no other apparent cause, being a challenge for clinical practice, since this is a silent condition, but with definitive surgical treatment.

### Author contributions

Conception and design of the research: Sousa JCV, Miranda RM, Silva PMN, Madruga GM, Figueiredo

NMS, Farias DC. Acquisition of data: Miranda RM, Silva PMN, Madruga GM, Figueiredo NMS, Farias DC. Analysis and interpretation of the data: Sousa JCV, Miranda RM, Silva PMN, Madruga GM, Figueiredo NMS, Farias DC. Writing of the manuscript: Miranda RM, Silva PMN, Madruga GM, Figueiredo NMS, Farias DC. Critical revision of the manuscript for intellectual content: Sousa JCV. Supervision / as the major investigator: Sousa JCV.

### Potential Conflict of Interest

No potential conflict of interest relevant to this article was reported.

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### Study Association

This study is not associated with any thesis or dissertation work.

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