

## Sudden Cardiac Arrest in Athletes: Do not Miss Suspicious Details

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### Introduction

Sudden cardiac death (SCD) related to sports activities is an unexpected and rare event, usually occurring in young and apparently healthy athletes. The main cause of SCD in young athletes (< 35 years old) is ventricular arrhythmia (VA) associated with arrhythmogenic disorders (e.g. hypertrophic cardiomyopathy, arrhythmogenic right ventricular cardiomyopathy – ARVC, channelopathies). Some of these conditions can be suspected by routine pre-participation clinical evaluation, while others remain undetectable even after careful screening.<sup>1-3</sup> This paper presents the case of a professional athlete with undiagnosed ARVC, whose first manifestation was malignant VA and biventricular dysfunction.

### Case report

A 19-year-old white male professional handball player participated in a regional league since he was 16. He had unremarkable medical or family history. His pre-season medical examination showed electrocardiogram (ECG) with sinus rhythm, inverted T wave in right precordial leads and occasional premature ventricular contractions (PVC) (Figure 1). For more information, his physician ordered 24-hour Holter monitoring, transthoracic echocardiography (TTE) and an exercise test. The 24-hour Holter monitoring revealed periods of sinus bradycardia and 3713 PVC occurring as isolated, pairs or triplets, independently of exertion. TTE and exercise test were described as normal and he was allowed to play.

### Keywords

Spots; Athletes; Physical Endurance; Risk Assessment; Sudden Death; Cardiac Arrest; Arrhythmias Cardiac; Arrhythmogenic Right Ventricular Dysplasia.

Two years later, the athlete collapsed due to cardiac arrest during a handball match, while he was defending an attack. His colleagues started cardiopulmonary resuscitation (CPR) immediately. The prehospital medical emergency team arrived 10 minutes later and detected ventricular fibrillation. After one shock, the patient showed signs of return to spontaneous circulation on the field. Afterwards, the young athlete presented another collapse in the emergency transport, with shockable rhythm. Advanced life support with defibrillation and mechanical compressions were started again and the patient recovered after a total of 20 minutes of CPR. The first ECG (figure 2A) at the emergency room showed sinus rhythm, right axis deviation, dominant R waves in V1 and elevation of ST segment in precordial leads.

Bedside TTE revealed dilated right chambers with biventricular global systolic dysfunction. Emergent coronary angiography showed normal coronary arteries. Lab tests showed high sensitivity troponin I elevation with no other relevant alterations.

Three days after the event, invasive mechanical ventilatory support and vasopressors were suspended and the patient recovered without neurological or cognitive deficits. Serial ECGs showed inverted T wave in right precordial leads (Figure 2B).

TTE was repeated and confirmed dilatation of the right chambers with marked trabeculations in the right ventricle (RV) and biventricular global systolic dysfunction (Figure 3A-D).

For further investigation, cardiac magnetic resonance (CMR) was performed, showing left ventricular (LV)



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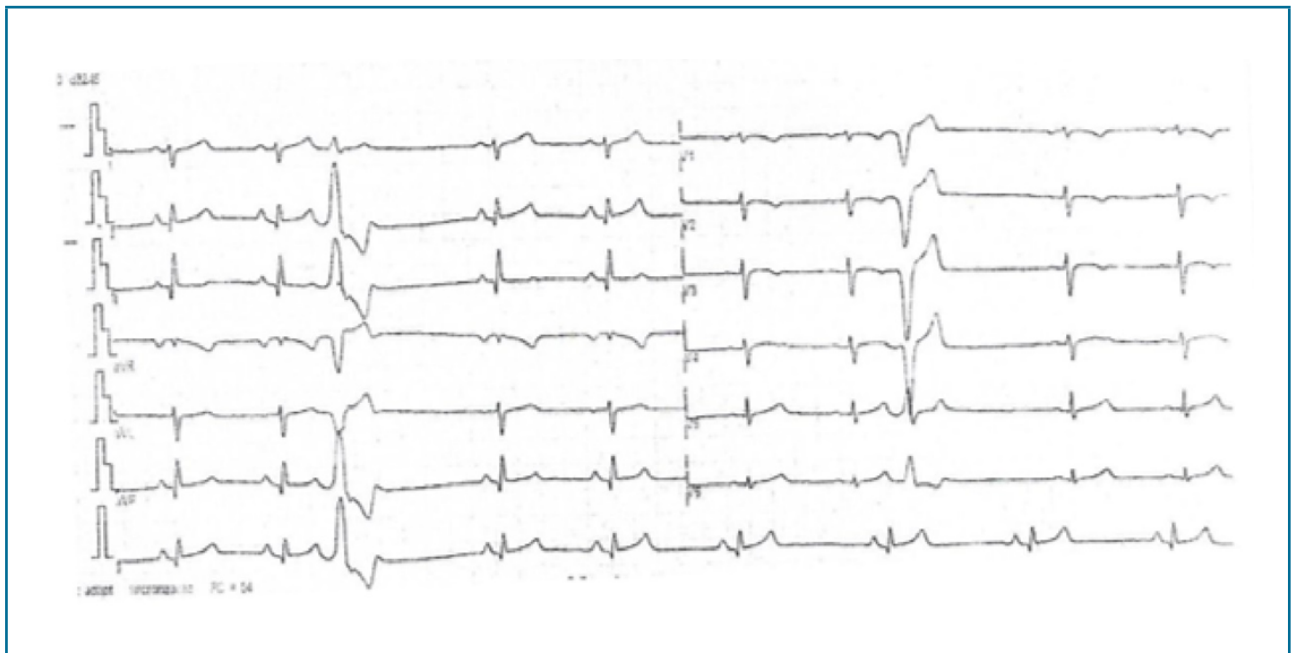


Figure 1 - Pre-seasonal routine electrocardiography.

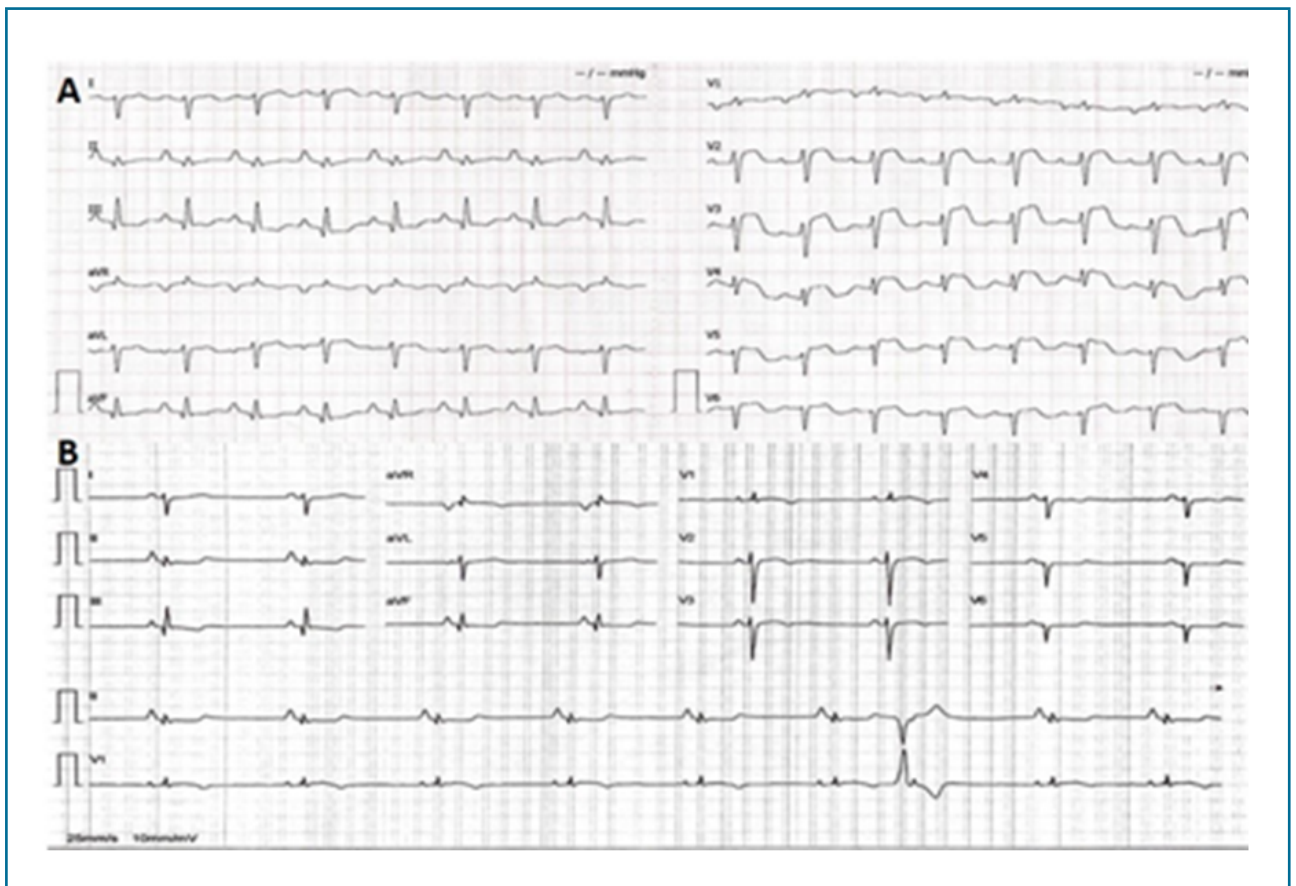


Figure 2 - A) First ECG after return of spontaneous circulation showing sinus rhythm, right axis deviation, dominant R waves in V1 and elevation of ST segment in the precordial leads. B) ECG after total recovery and at hospital discharge. ECG: electrocardiography.

end-diastolic volume in the upper limit of normality (104 ml/m<sup>2</sup>) and mild global LV systolic dysfunction (LV ejection fraction 41%); RV was severely dilated (148 ml/m<sup>2</sup>) with dyskinesia in the outflow tract and severe systolic dysfunction (RV ejection fraction 28%). Late gadolinium enhancement was found in the RV outflow tract (transmural) and in the LV inferolateral wall (subepicardial). These findings suggested diagnosis of ARVC with biventricular involvement (Figure 3E-G). Genetic test revealed desmoglein 2 gene variant of uncertain significance (c.874C > T).

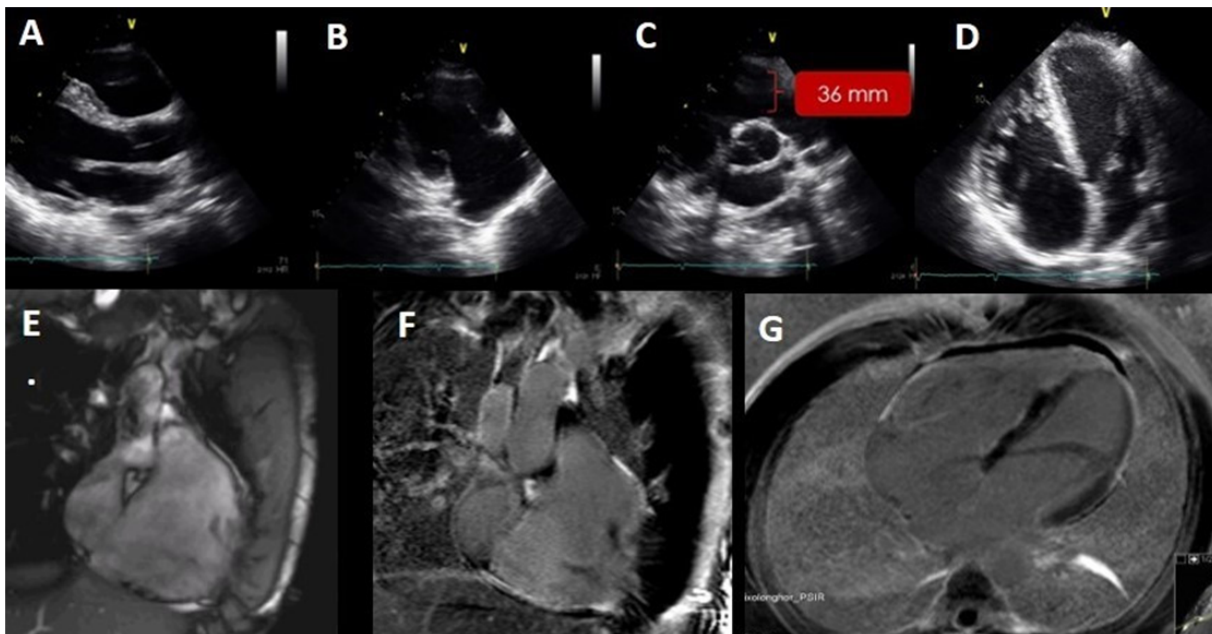
Due to frequent PVC and runs of nonsustained ventricular tachycardia (NSVT) with inferior axis and left block morphology non-responsive to beta-blockers, sotalol was started with success. Implantable cardioverter defibrillator (ICD) was implanted for secondary prevention.

The patient was discharged home on sotalol 160 mg twice a day and lisinopril 5 mg daily. He was advised not to engage in competitive and/or endurance sports.

## Discussion

ARVC is a heritable heart muscle disorder characterized by VA, heart failure (HF) and SCD.<sup>4</sup> ARVC is usually diagnosed in adolescence-young adulthood and is more frequent in male patients. Known disease-causing genes mostly encode desmosomal proteins, although the true prevalence of these genes has yet to be determined. Therefore, a negative genetic test does not rule out the ARVC diagnosis.<sup>5</sup>

The histopathological hallmark of ARVC is the replacement of myocardium by fibrofatty tissue, predominantly in the RV wall. It occurs mostly in the RV inflow tract, outflow tract and apex (“triangle of dysplasia”), from the epicardium to the endocardium. These changes lead to wall thinning and aneurysm formation. More infrequently, though not rare, there is LV involvement, as in our case.<sup>6</sup> Physical exercise may aggravate mechanical uncoupling of myocytes; therefore, exertion is a trigger of disease onset and progression, as well as VA.<sup>4</sup>



**Figure 3 - Top:** TTE with dilated right chambers (A: parasternal view; B: right chambers parasternal view; C: short axis view showing RV outflow tract), hypertrabeculated RV (D: four-chamber view) and biventricular systolic dysfunction. **Bottom:** CMR revealed LV end-diastolic volume in the upper limit of normality with mild systolic dysfunction, severe RV dilatation, dyskinesia in outflow tract and severe RV systolic dysfunction (E). Late gadolinium enhancement was visible in the RV outflow tract (transmural) and in the LV inferolateral wall (subepicardial) (F and G). TTE: transthoracic echocardiography; CMR: cardiac magnetic resonance; RV: right ventricle; LV: left ventricle.

Clinically, three phases have been described in ARVC. In the “concealed phase,” patients are often asymptomatic. In the overt “electrical phase,” they present symptomatic arrhythmias with/without structural abnormalities in imaging tests. Progressive disease may result in RV, LV or biventricular HF combined or not with VA.<sup>5</sup>

Cardiac arrest can be the first manifestation, as in our case, even in the concealed phase.<sup>4,5</sup> Our patient had some abnormalities on the previous ECG and on the 24-hour Holter that could suggest the diagnosis, such as inverted T wave in V1-V3 leads and frequent PVC. Indeed, in recent international recommendations for electrocardiographic interpretation in athletes, anterior inverted T wave in non-black athletes and > 2 PVC per 10 seconds tracing are considered ECG abnormalities.<sup>7</sup> However, these abnormalities are nonspecific and, after normal TTE, our patient was cleared to sport practice.

CMR is the preferred imaging method for ARVC diagnosis. It is useful not only in RV morphological and function evaluation, but also in tissue characterization. Late gadolinium enhancement can be visible predominantly in subepicardial RV wall and/or LV inferolateral (observed in this case) or inferoseptal regions, contributing to the early diagnosis of left-sided disease.<sup>8</sup>

Current task force criteria for ARVC diagnosis<sup>9</sup> includes major and minor criteria concerning morphological RV abnormalities (by TTE, CMR or angiography), pathological abnormalities in RV endomyocardial biopsy (EB), depolarization and repolarization changes in ECG, VA, and family history. In the presence of 2 major criteria, 1 major and 2 minor criteria or 4 minor criteria, ARVC diagnosis is considered definitive.

Our patient fulfilled 2 major and 2 minor criteria: 1 major criteria of morphological RV abnormalities (RV dilatation and RV systolic dysfunction with regional dyskinesia in TTE/CMR), 1 major criteria of repolarization abnormalities (inverted T waves in precordial leads) and 2 minor criteria of VA (NSVT with RV outflow morphology and > 500 PVC in 24-hour Holter monitoring). He had no depolarization abnormalities or family history of ARVC or SCD. Given the results of non-invasive investigation, EB was dismissed.

Advising against competitive/endurance sports is the first step in the treatment of ARVC. Beta-blockers prevent exertion-induced VT/VF and are

recommended in the presence of HF, as well as the remaining standard HF therapy. Antiarrhythmic drug therapy has a role in patients with symptomatic frequent PVC/NSVT or many other appropriate ICD therapies.<sup>10</sup> ICD therapy is recommended in patients with history of aborted SCD and hemodynamically poorly tolerated ventricular tachycardia.<sup>10</sup>

## Conclusion

This case presents a malignant ARVC presentation in a young athlete with previous subtle abnormalities in medical exams. It highlights the missing details on clinical evaluation even when a comprehensive medical examination is performed. All patients' data need to be carefully analyzed in order to early detect cardiac disease, like ARVC, and reduce SCD in this population. Disqualification of affected patients from competitive sports might be life-saving.

## Authors Contribution

Conception and design of the research: Braga M. Acquisition of data: Braga M, Araújo P. Analysis and interpretation of the data: Braga M, Araujo P. Writing of the manuscript: Braga M, Dias P, Vasconcelos M, Almeida R, Maciel MJ. Critical revision of the manuscript for intellectual content: Braga M, Dias P, Vasconcelos M, Almeida R, Maciel MJ.

## 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.

## Ethics approval and consent to participate

This article does not contain any studies with human participants or animals performed by any of the authors.

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