

Sudden cardiac death in athletes: A brief review of literature with a case report

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Abstract

Background

Sudden cardiac death (SCD) in athletes is a rare event; however, it is still 2-4 times more often when compared to non-athletes. The single most important predictor is exercise fainting or near fainting, which should be a warning requiring explanation and investigation. Early identification of athletes at high risk of sudden death imposes drastic decisions such as restriction of competitive sports activity, but also by prophylactic treatment with drugs and implantable cardioverter defibrillator (ICD).

Case Report

The author report here the case of a 43-year-old female triathlete who had recurrent syncope from physical effort (running) caused by a life-threatening ventricular arrhythmia and therefore was declared ineligible for competitive sport. This athlete however, died a few years later suddenly during a cycling event.

Conclusion

Sports cardiologists should never underestimate the life-threatening ventricular arrhythmias when these are associated with recurrent syncope. Relying not only on the results of genetic and cardiovascular imaging, but rather considering clinical observations and making accurate diagnoses when the two hallmark signs are present with vulnerable substrate: syncope associated to malignant ventricular arrhythmias.

Introduction

Sudden cardiac death (SCD) is defined by the World Health Organization (WHO) as that condition which occurs within the first 24 hours after the symptoms' onset. However, several experts shorten this time and describe it as "an unexpected death, marked by abrupt loss of consciousness in an individual within the first hour after symptoms' onset with or without heart disease"¹.

Sudden death occurs in approximately one in 200,000 athletes annually². SCD in athletes is rare; however, it is still 2-4 times more often when compared to non-athletes. Several mechanisms are proposed: myocardial ischemia, repolarization due to potassium channel down regulation and it may also be explained by the presence of multiple

factors such as cardiac hypertrophy (athlete's heart) and/or hypertrophic cardiomyopathy, increased sympathetic tone, genetic defects, drugs, doping agents or food ingredients. These factors together may increase the repolarization homogeneity which sometimes leads to risk of arrhythmias and SCD³. The prevention of sudden death by athletes⁴, remains a challenge. First, the prevalence of any single, associated, condition is very low, probably less than 0.3%. Second, the sensitivity and specificity of tests commonly used to screen, leaves much to be desired. The single most important predictor is exercise fainting or near fainting, which should be considered a warning requiring explanation and investigation⁵. While almost all of the causes relate to congenital or acquired cardiovascular disease, an exception is commotio cordis, in which the heart is structurally normal but a potentially fatal loss of normal rhythm occurs because of the accidental timing a blow to the chest. Survival is about 15% and time to cardiopulmonary resuscitation (CPR) and defibrillation may determine survival. The victim is usually male and, in the US, two-thirds occur in football and basketball, reflecting the popularity of these sports and the number of athletes involved in them. Elsewhere in the world, soccer is the most commonly associated. In statistics derived from National Collegiate Athletic Association (NCAA) data, one death per 25,000 collegiate athletes over a five years period was attributable to medical causes⁶. Over the last several years, sudden deaths of trained athletes, usually associated with exercise, have become highly visible events fuelled by news media reports and with substantial impact on both the physician and lay communities^{7,8,9}. Exercise can trigger an acute cardiac event which may precipitate to arrhythmic sudden cardiac arrest (SCA)¹⁰. In two studies from France and The Netherlands, the incidence and prognosis of exercise-related out-of-hospital cardiac arrest (OHCA) have now been reported^{11,12}. We do know that death during an athletic event can result from direct and indirect causes, but is mainly caused by a malignant ventricular arrhythmia that degenerates into ventricular fibrillation, which can be resolved with a defibrillation¹³. The direct causes of death primarily are traumatic (i.e., a closed head injury). The major indirect cause of death during athletics is from an arrhythmia resulting in sudden cardiac arrest. Preventing such arrests is difficult if the pre-existing cardiac condition has not been recognized. To try and achieve this end, physicians should include a thorough history and cardiac examination as part of the pre-participation physical examination. 12-lead electrocardiogram (ECG), a stress rhythm ECG, or both, should be performed if any risk factors are identified at

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initial assessment¹⁴. Those risk factors include a history of chest pain, shortness of breath, or arrhythmia; a history of cardiac problems requiring medication; and a family history of death due to sudden cardiac arrest at a young age. Findings on clinical examination of bruits, murmurs, high blood pressure, and arrhythmias deserve more thorough evaluation. The vast majority of 'at risk athletes' does not experience premonitory symptoms and, thus, preparticipation screening represents, in spite of everything, the only strategy capable of identifying the underlying cardiovascular disorder¹⁵. The importance of early identification of silent cardiovascular diseases at a sub-clinical stage relies on the concrete possibility of SCD prevention by lifestyle modification, including restriction of competitive sports activity, but also by prophylactic treatment with drugs and implantable cardioverter defibrillator (ICD)¹⁶. Interest in these tragic events has accelerated owing to their increased recognition; awareness that underlying, clinically identifiable cardiovascular diseases are often responsible; and the availability of preventive treatments for avoiding sudden death suffered by high-risk athlete-patients. We present the case report of an athlete who suffered sudden death during physical activity, and who perhaps, in hindsight, could have been saved by being constantly surveyed under a more aggressive invasive treatment. To highlight the ability to recognize athletes at high risk of sudden death when clinical signs such as both syncope and electrocardiographic documentation of threatening cardiac arrhythmia are present.

Case Report

This case report describes the story of a 43-year-old female triathlete who promptly interrupted practicing competitive sports during a preliminary sports pre-participation screening for a triathlon competition in 2010 due to recurrent episodes of palpitations following syncope induced by fast running that was occurred in the last 3 months. The family history was negative for heart disease and sudden death. The physical examination was unremarkable.

Resting ECG showed premature ventricular beat with couplets and T-wave inversion on precordial leads (From V1 to V4) (Figure 1). QTc interval was in the normal range (< 450 ms) with a slight increase at night (<480 ms) on Holter ECG monitoring.

A exercise ECG treadmill testing revealed a polymorphic ventricular tachycardia with R-R interval shorter than 150 ms (Figure 2) symptomatic for dizziness and presyncope. A two-dimensional transthoracic (2D TT) echocardiogram showed the normal size and morphology of the cardiac chambers with good systolic function and no valvular abnormalities (Figure 3). Consequently, the athlete was admitted to two consecutive cardiology centres to determine the precise etiology of the ventricular arrhythmia and the probable genetic cause, e.g. mutations, responsible for some variants of long QT syndrome (LQTS) or catecholaminergic polymorphic ventricular tachycardia (CPVT). However, genetic analysis for the identification of the mutations responsible for some variants of long QT syndrome (LQTS) and catecholaminergic polymorphic

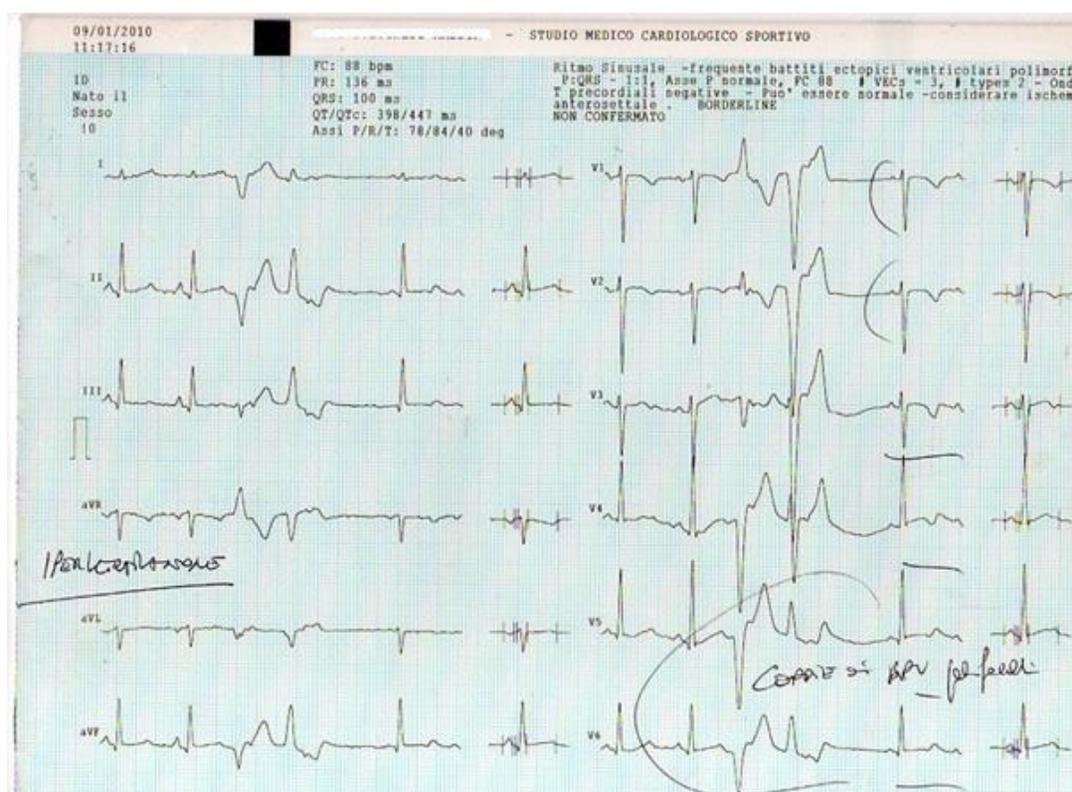


Figure 1: Resting ECG shows premature ventricular beats with couplets and T-wave inversion on precordial leads (From V1 to V4).

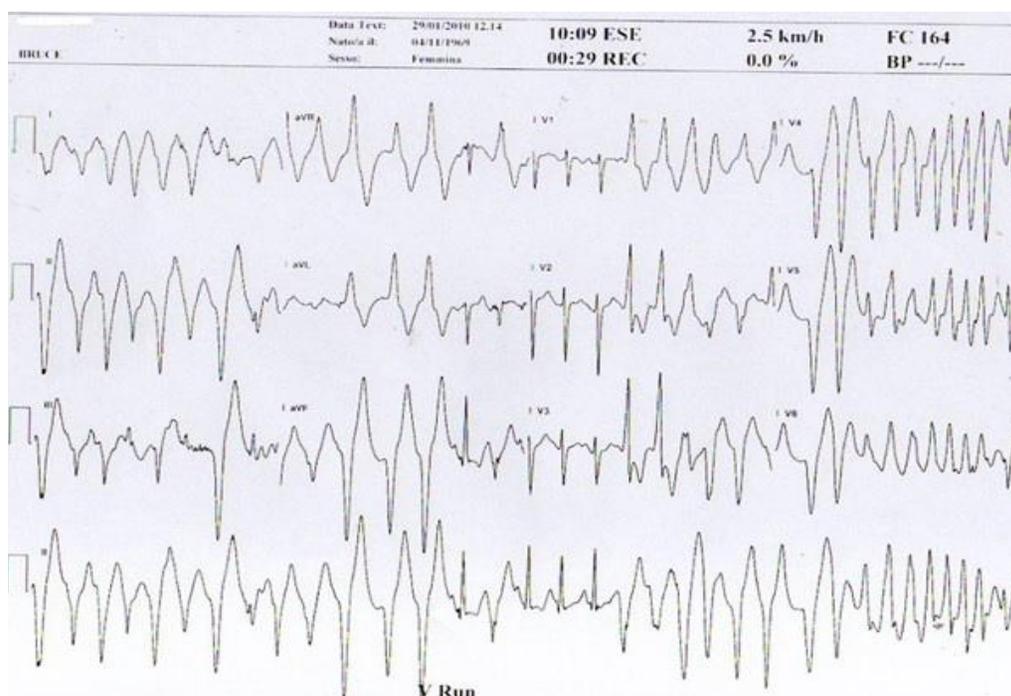


Figure 2: Exercise ECG treadmill test shows polymorphic ventricular tachycardia with R-R interval shorter than 150 ms.

ventricular tachycardia (CPVT) were negative. The screening covered the entire coding sequence of the genes *KCNQ1*, *KCNH2*, *SCN5A*, *KCNE1*, *KCNE2*, *hRyR2* did not identify any mutation in the genes analyzed.

A second cycle ergometer exercise ECG testing, under beta-blocker therapy, showed only a few premature ventricular beats, infrequent and monomorphic, with rare couple. Even a cardiac magnetic resonance imaging with contrast was negative for cardiomyopathy and CARTO mapping showed normal voltages. A coronary angiography and intracardiac electrophysiology studies were not performed. A right ventriculography with endomyocardial biopsy was then made. The histological findings showed

significant but “nonspecific” myocardial abnormalities such as moderate fibrosis and myocellular vacuolization, features that were suggestive of an arrhythmogenic substrate of uncertain nature, so the medical pathologist wrote. After prolonged discussion amongst the cardiological heart team, the triathlete was discharged with the recommendation to refrain from running and undergo beta-blocker therapy (with Nadolol 80 mg die). Unfortunately, this athlete disappeared during follow-up without a trace until received the news of his sudden death three years later during a cycling competition. This work conforms to the values laid down in the Declaration of Helsinki (1964). The protocol of this study has been approved by the relevant ethical committee related to our institution in which it was performed. All subjects gave full informed consent to participate in this study.

Discussion

Several events in all parts of the world show that sudden cardiac death (SCD) in athletes is still a reality which continues to challenge experts in sport and medicine cardiology collaborating with athletes^{17,18}. Warrò et al¹⁹ proposed a mechanism underlying sudden cardiac death in athletes that does not relate to myocardial ischemia but is based on repolarization abnormalities due to potassium channel down regulation and can also be best explained by the concurrent presence of several factors such as cardiac hypertrophy (athlete’s heart), and/or hypertrophic cardiomyopathy, increased sympathetic tone, genetic defects, drugs, doping agents, food, or dietary ingredients. Together these factors can increase the repolarization inhomogeneity of the heart (“substrate”) and an otherwise



Figure 3: Echocardiographic findings shows no structural abnormalities of the heart.

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“harmless” extrasystole (“trigger”) occurring at the wrong time may sometimes induce life-threatening arrhythmias. The case of this unfortunate athlete raises many questions about diagnosis, treatment and follow up. First of all, it must be pointed out that this competitive athlete, physically active from an early age, presented recurrent syncopal and pre-syncopal episodes during physical exertion. The exercise ECG treadmill tests performed in two centres showed very rapid ventricular arrhythmias and especially an episode of polymorphic ventricular tachycardia. Despite cardiovascular examinations resulted negative, image (CARTO mapping and Cardiac Magnetic Resonance), the histological finding of cardiac myocardial biopsy deposed for underlying cardiomyopathy or unrecognized arrhythmogenic heart disease. In fact, findings were clearly present on resting ECG findings suggestive of cardiomyopathy such as inverted (negative) T-waves in anterior precordial leads (V1 to V4). The presence furthermore of polymorphic ventricular beats and inducible polymorphic ventricular tachycardia by exercise testing evoked strong suspicion of Arrhythmogenic Right Ventricular Cardiomyopathy or Catecholaminergic Polymorphic Ventricular Tachycardia. For this cardiac vulnerable substrate the athlete died suddenly. It could, therefore, be assumed that a similar event was foreseeable, but unfortunately, no ICD was applied. The eventual effective prevention of sudden cardiac death requires the development of novel cost effective cardiac electrophysiological screening methods. Athletes identified by these tests as individuals at higher proarrhythmic risk should then be subjected to more costly genetic tests in order to uncover possible underlying genetic causes for alterations in ionic channel structure and/or function. In the case of no evident structural heart disease or genetic cause, the decision should bravely only be made on clinical data and then on the phenotypic expression of the disease. Fate unfortunately exceeds our noble attempts to prevent sudden death in athlete.

Conclusions

The take-home message is: never underestimate the crucial signals suggestive for possible sudden death in athletes such as syncope associated to life-threatening ventricular arrhythmias. This case is emblematic for the lack of pathognomonic data regarding of diagnosing underlying cardiomyopathy, despite the histological evidence of an arrhythmogenic substrate and the phenotypic expression of the disease, perhaps no one had the idea to suggest ICD implantation to the athlete. It is therefore dramatic that an athlete, discovered to be a bearer of a potentially lethal arrhythmia and alerted to the potential risk of sudden death, thus completely ignored this diagnosis and medical recommendations on total abstention from all forms of physical activity. It is therefore a paradox that an athlete, diagnosed with a potentially lethal arrhythmia and warned about the sudden death risk, should ignore this information with relative

recommendations to avoid physical activity. Not rely only on the results of genetic and cardiovascular image, but rather to give prominence to the hospital and make accurate diagnoses when the two hallmark signs are present without clearly anatomic substrate: syncope associated to malignant ventricular arrhythmias. The confirmation to this story is that the athlete's twin sister, who even suffered from the same symptoms and consequently from the same disease, subsequently was treated with ICD.

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