

Sudden Cardiac Death in a Female Triathlete: Complexities of Risk Stratification

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Abstract

The prevention of sudden death in athletes is one the greatest challenges in sports cardiology. Early identification of athletes at high risk of sudden death can lead to difficult decisions such as restriction of competitive sports activity, but also treatment with medications and implantable cardioverter-defibrillator. We report a case of a 41-year-old female triathlete who had recurrent syncope during exercise with documented polymorphic ventricular tachycardia and nonspecific abnormalities on endomyocardial biopsy. Against medical advice, she continued to participate in athletic events and subsequently had a sudden cardiac death during a cycling event.

Keywords: Sudden cardiac death; Athletes; Implantable defibrillators; Ventricular tachycardia

Introduction

Sudden cardiac death (SCD) occurs in approximately one in 200,000 athletes annually [1]. For most, the underlying mechanism is due to an abnormal substrate (i.e, channelopathies, cardiomyopathies, Brugada or Marfan syndrome, coronary artery disease) and a trigger (i.e. myocardial ischemia, premature ventricular contractions, increased sympathetic tone, performance enhancing drugs). Prevention of sudden death due to an arrhythmogenic cause is a very difficult challenge.

Over the last several years, sudden deaths of athletes, usually associated with exercise, have become highly visible events fuelled by news media reports with substantial impact on both lay communities and the physician [2-4]. While there is debate about the use of ECG screening for the asymptomatic patient [5], a thorough evaluation must be completed in those with symptoms, especially syncope with exercise. This case demonstrates that risk stratification to determine implantable cardioverter-defibrillator (ICD) placement can be difficult in those with syncope and that restriction from sports is only effective if the patient agrees.

Case Report

A 41-year-old female triathlete had an episode of syncope during running, for which she did not initially seek medical attention. A year later, she had two episodes of palpitations and presyncope occurring during a race, at which point she saw a physician. Resting ECG showed multifocal premature ventricular contractions (PVC) and T-wave inversion from V1-2, though T wave inversions post-PVC persisted out to V4 (Figure 1). The QT interval was normal and there was no evidence of delta or epsilon waves. ECG in the resting phase after a submaximal cycle ergometer test again showed multifocal PVCs.



Figure 1: Resting electrocardiogram demonstrating T wave inversions in V1-2, though post PVC T wave inversions extend from V1-V4. Multifocal PVCs are seen.

She then underwent a Bruce protocol exercise ECG treadmill test. This showed a self-terminating, rapid polymorphic ventricular tachycardia associated with presyncope (Figure 2).

A transthoracic echocardiogram demonstrated normal chamber size, function, and morphology with no valvular abnormalities. She was then evaluated at two cardiology centers to determine the precise etiology of the ventricular arrhythmia and the probable genetic syndrome responsible.

A cardiac magnetic resonance imaging with contrast was negative for cardiomyopathy with no late gadolinium enhancement. Voltage mapping using the CARTO system (Biosense Webster, Inc., Diamond Bar, CA) showed normal voltages with no evidence of scar, though there was the presence of a small bulging inferior to the pulmonary valve in the septal right ventricular outflow tract of unclear significance. No programmed stimulation was performed.

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A right ventricular endomyocardial biopsy was then made. The histological evaluation showed significant but "nonspecific" myocardial abnormalities of moderate fibrosis and myocellular vacuolization. Genetic testing of KCNQ1, KCNH2, SCN5A, KCNE1, KCNE2, and RyR2 did not identify any mutations.



Figure 2: Exercise treadmill test showing a slow polymorphic ventricular tachycardia (PMVT), followed by a brief period of sinus tachycardia. This is followed by a similar PMVT followed by rapid PMVT.

After prolonged discussion with her physicians, she was started on nadolol, and it was recommended she refrain from exercise and competitive events. A second cycle ergometer exercise ECG test while on nadolol showed only infrequent, monomorphic premature ventricular beats.

The patient did not follow-up with a physician afterward. Three years later, she died suddenly during a cycling competition, presumably due to a ventricular tachyarrhythmia. It is not known if she was taking nadolol at the time. The patient's twin sister also had symptoms of dizziness and later suffered a sudden cardiac arrest, but was resuscitated and treated with an ICD.

Discussion

SCD in athletes is a reality which continues to challenge experts in sports cardiology collaborating with athletes [6,7]. Generally, the term electrical cardiomyopathy refers to LQST, SQTS, Brugada syndrome and catecholaminergic polymorphic VT (CPVT). All of these diseases put the patient at increased risk of SCD. For example, as Jacoviello et al. reported [8], Brugada Syndrome (BRS) is a hereditary channelopathy that can be characterized by mild right ventricular (RV) abnormalities that are not detectable with conventional echocardiography.

While ECG screening has received much debate recently, risk stratification is still difficult in many situations, and whether to implant an ICD is not always an easy decision. Similarly, having patients accept their diagnoses and follow through with recommendations may not always occur.

Our patient did not have a phenotype consistent with a single diagnosis. She developed bidirectional PVCs with exercise and polymorphic VT which can be seen in CPVT, but it is unusual to have T wave abnormalities and an abnormal histologic substrate. Her QT

interval was normal, suggesting this was not long QT or short QT syndromes. There was no early repolarization pattern. There was no Brugada pattern on multiple ECGs, and SCA with exercise is unusual with Brugada syndrome. ECG, MRI, voltage map and biopsy were not consistent with arrhythmogenic right ventricular cardiomyopathy. Echocardiogram did not show any evidence of mitral valve prolapse. It is possible that she had phospholamban cardiomyopathy, as this has been associated with a similar phenotype, but testing for this gene was not completed [9].

This case demonstrates two important points. Risk stratification in patients with unexplained syncope and idiopathic ventricular arrhythmias is difficult, as these are a heterogenous group of undescribed genetic abnormalities and syndromes. In LQT and CPVT, patients thought to be highest risk are those with recurrent syncope while on beta-blocker. She had an improvement in ventricular ectopy on exercise testing while taking a beta-blocker, so an ICD was not implanted during the initial presentation. If she had had appropriate follow-up and she reported non-adherence to her beta-blocker or continued symptoms, it would likely have been reasonable to offer her an ICD at that time. Left cardiac sympathetic denervation may also have been reasonable to perform. Recently there has been a questioning of the Bethesda criteria [10] to allow the return to sports in those with an ICD [11,12], though given that all of her events were with exercise, we would not have recommended this patient participate in sports. The second issue relates to patient autonomy and decision making. In the United States, there is a legal framework for organizations to prevent athletes from returning to sports [13].

In Italy, a 12-lead ECG and limited stress test is routinely obtained as part of a mandatory comprehensive screening program [6]. This national sports pre-participation screening has been performed at regional centers by highly specialized physicians according to the COCIS protocol [14].

This federally mandated and funded systematic screening, coupled with athletic restriction, has been associated with a decline in deaths. Fundamental to the this screening program is the belief that trained athletes represent a special subset of the general population who are at higher risk for sudden death, as well as reported by other recently recommendations [15].

However, there are not medical evaluations of patients prior to recreational events, and it is incumbent on the patient to self-report and self-disqualify for most athletic competitions. Therefore, in this situation, the patient decided not to follow physician recommendations. It is important that patients are fully educated regarding their disease, as best as can be possible, so they can understand the reasoning behind recommendations and prevent SCA events.

Unfortunately, due to the low incidence of electrical cardiomyopathies there is a lack of large randomised trials studying their treatment. However, primary prevention by ICD therapy should be considered in patients at high risk of SCD for recurrent syncope due PCVT.

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