

CAUSES AND PREVENTION OF SUDDEN CARDIAC DEATH IN YOUNG ATHLETES

By

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Introduction

Regular physical activity has several beneficial effects on health and is linked with reduced cardiovascular and all-cause mortality. However, in certain circumstances, an acute bout of exertion may cause dangerous health effects and, in extreme cases, sudden death (the so called “*paradox of exercise*”). Although rare, the sudden death of a young athlete is a dramatic event with a devastating impact on his family, sports community, and medical staff. It was the case of professional soccer player Marc-Vivien Foe from Cameroon (28 years old), who died suddenly while playing in the Confederations Cup, due to a hypertrophic cardiomyopathy, or, more recently, of the Spanish soccer player Antonio Puerta (22 years old) who had a syncope on the field, with spontaneous recovery and a new collapse in the locker room due to a fatal cardiac arrest. No information on the cause of death is available (newspapers suggested an arrhythmogenic right ventricular cardiomyopathy), but, unequivocally, he suffered from other “unexplained” syncopal attacks on effort before the death.

Dealing with exercise-related sudden death (ESD), we usually refer to an *unexpected and witnessed sudden cardiac arrest occurring within 1 hour from an exercise bout in an apparently healthy person*. Following this definition, ESD is *rare*, with an incidence of 1/100'000 to 1/300'000 per year in males. This explains why, still today, experts discuss on the usefulness of a routine medical pre-participation screening in preventing ESD.

Mechanisms and substrates of exercise-related sudden death

It is largely demonstrated that *cardiovascular diseases* play the dominant role in ESD, accounting for 80-85% of cases. Usually, the fatal cardiac arrest occurs either during or immediately after strenuous exercise, suggesting that autonomic nervous system changes related to effort may trigger malignant arrhythmias in subjects with cardiac disease. This aspect is indirectly confirmed by the fact that ESD is more frequent during official events in respect to training sessions, probably because of the greater psychological involvement.

Among cardiovascular diseases, *atherosclerotic coronary artery disease* is the leading cause in older athletes (>35 years). In younger athletes (<35 years), the substrate responsible for ESD is represented by rare, often silent, heart diseases, the most frequent being:

- *Hypertrophic cardiomyopathy (HCM)*, a genetic disease, with a prevalence in the general population of about 1/500. HCM is the dominant cause of ESD among young US athletes, a feature that can be explained both by the greater prevalence of the disease in Blacks, and by the absence of a routine medical pre-participation screening in USA. HCM is characterized by an abnormal hypertrophy of left ventricular walls, with a spatial disarray of myocardial fibers at microscopic level, and propensity to life-threatening arrhythmias, the final path to sudden death. *Idiopathic left ventricular hypertrophy (ILVH)*, accounts for about 10% of all ESD cases among young US athletes. It is usually characterized by an extreme symmetrical left ventricular hypertrophy (wall thickness >13 mm), a reduced cavity diameter and an impaired diastolic filling. ILVH should be differentiated by the “athlete’s heart”, showing moderate symmetric left ventricular hypertrophy, mild-to-moderate cavity dilation and normal (or even “supernormal”) diastolic filling.

HCM may be totally asymptomatic and the physical exam may be completely negative. However, the diagnostic suspicion can arise from the electrocardiogram (ECG), very frequently abnormal. In our experience on 72 young athletes with HCM, an abnormal rest ECG was the cause of referring to our laboratory in 80% of cases, indicating that ECG is a sensitive and specific tool in detecting HCM, especially in post-pubertal



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athletes (*figure 1*). A well-conducted echocardiogram (ECHO), and/or cardiac magnetic resonance imaging (MRI) generally confirm the diagnosis.

- *Congenital coronary artery anomalies (CCAA)* are very rare in the general population. However, young athletes with CCAA have a 70-fold increase in relative risk of dying during effort than at rest (*Corrado D, JACC 2003*). The most common anomaly linked with ESD is a left main artery arising from the right sinus of Valsalva, particularly when its course to the usual position is between pulmonary artery and aortic root. In some cases, ESD may be due to a “malignant” myocardial bridge (intramural course of a coronary artery). Unfortunately, CCAA are rarely diagnosed in lifetime. The physical exam, as well as rest and stress ECG, are usually normal and only about 1/3 of subjects refers symptoms (chest pain, palpitations and/or syncope/presyncope). However, when symptoms are present, especially during or immediately after a strenuous effort, a suspicion of a CCAA should be arisen. ECHO, in experienced laboratories, can be useful as first diagnostic approach and, nowadays, MRI and angio-coronary computed tomography (angio-CT) scans can solve doubts on coronary arteries anatomy (*figure 2*).
- *Arrhythmogenic right ventricular cardiomyopathy (ARVC)* is the leading cause of ESD in young athletes in Italy and Europe. ARVC is a genetic condition causing fibrosis and fatty infiltration of the myocardium, with progressive wall thinning and dilation of the right ventricle, associated with ventricular tachyarrhythmias, particularly in relation to psycho-physical stress. Subjects with ARVC may be completely asymptomatic or may refer palpitations, syncope/presyncope at rest and/or during effort. Cardiac physical exam may be negative, but ECG is often abnormal, showing QRS complex widening (the so-called “epsilon” wave, due to right ventricular activation delay), repolarization abnormalities (negative T waves in right precordial leads), and ventricular premature beats. ECHO and MRI let to better face the diagnostic suspicion (*figure 3*).
- *Myocarditis (MYOC)* is an inflammatory condition of the myocardium, more frequently due to a viral infection. In our experience, athletes with MYOC may be fully asymptomatic or may show subtle symptoms (exercise intolerance, palpitations, tachycardia at rest and excessive heart rate increase on exercise, etc.), which appear days or weeks after a viral illness. For this reason, MYOC incidence in athletes is



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probably underestimated. ECG may be abnormal and dysrhythmias are frequently present when investigated with stress ECG and Holter monitoring. Malignant ventricular arrhythmias may be responsible for ESD in presence of either active or healing myocarditis. Thus, an early clinical diagnosis and full recovery after MYOC, are mandatory for a safe return to sport.

- *Wolff-Parkinson-White (WPW) syndrome* is a disease of the cardiac conduction system, characterized by the congenital presence of one (or more) additional electrical pathways. Depending on the electric characteristics of these abnormal pathways, atrial impulses, bypassing the atrio-ventricular node, can reach the ventricles more rapidly and, in case of atrial tachyarrhythmias (atrial fibrillation) a desynchronization of ventricular activity, deteriorating in a ventricular fibrillation (VF), is possible. The WPW prevalence ranges from 0.15% to 0.2% of the general population, but the risk of ESD regards only the 0.1% of these subjects. Invasive or transoesophageal electrophysiological study is mandatory in symptomatic subjects to assess the ESD risk.
- *Primitive electric heart diseases* are genetic anomalies of ion channels (“*channelopathies*”). They include some rare, malignant conditions as long and short QT syndromes, Brugada syndrome, polymorphic catecholaminergic ventricular tachycardia, etc. An accurate analysis of rest and stress-ECG may arise the diagnostic suspicion in most of cases, allowing withdrawal of these subjects from competitive sports.
- *Pharmacologic treatments and doping*: stimulants as epinephrine, ephedrine, cocaine and related sympathetic drugs all have a pro-arrhythmic effect, particularly in presence of dehydration, dyselectrolytemia (both very frequent in boxers) or in association with other heart abnormalities. Deaths have also been linked to performance-enhancing agents, as erythropoietin, mainly due to blood hyper-viscosity and increased thrombogenesis. Anabolic steroids may cause cardiac hypertrophy, myocardial fibrosis, and accelerated atherosclerosis and may be responsible for acute cardiac events and ESD.
- *Commotio cordis (CC)* is an electrophysiological event caused by precordial chest impact occurring in individuals usually free from heart disease. When the chest impact



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is delivered within a narrow, electrically vulnerable period of the cardiac cycle, it may start a VF. CC is a not rare cause of ESD in contact sports and one case was documented in soccer: after a penalty kick, a goal-keeper playing in the Argentinean league, received a violent ball-hit on his chest and fall unconscious to the ground. A recent study of our group (*Bianco M, Intern J Sports Med 2005*), however, showed that no clinical and humoral signs of myocardial damage are found after official amateur boxing matches, although ventricular repolarization abnormalities can be found on ECG in 20% of boxers, to be likely related to sympathetic hyper-activity linked to the agonistic event.

Conclusions

Overt or concealed heart diseases are responsible for 80-85% of ESD in athletes. Sports physicians should be able to recognize clinical symptoms (especially palpitations and presyncope/syncope during exercise) of potentially fatal conditions in young athletes. This is the first step in preventing the rare, but tragic, occurrence of ESD among young boxers and athletes.

Moreover, since the large majority of the diseases responsible for ESD in young athletes are hereditary, and are already evident in post-pubertal age, pre-participation screening should be applied to young players starting competitive amateur career. The efficacy of screening depends mostly on the experience and competence of visiting physicians, but a simple and *low-cost protocol, consisting of a careful medical and familial history taking, a thorough physical examination, a rest ECG* and, when possible, a stress ECG can identify a large number of subjects with potentially lethal diseases. Further instrumental investigations (ECHO, stress ECG if not done before, Holter, MRI, angio-CT and so on) may be useful when any diagnostic doubt arises.



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Figures.

Fig. 1

A

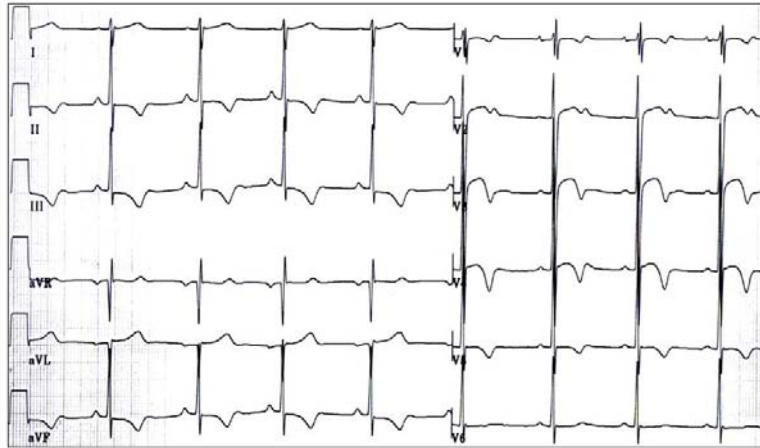


Fig. 1

B

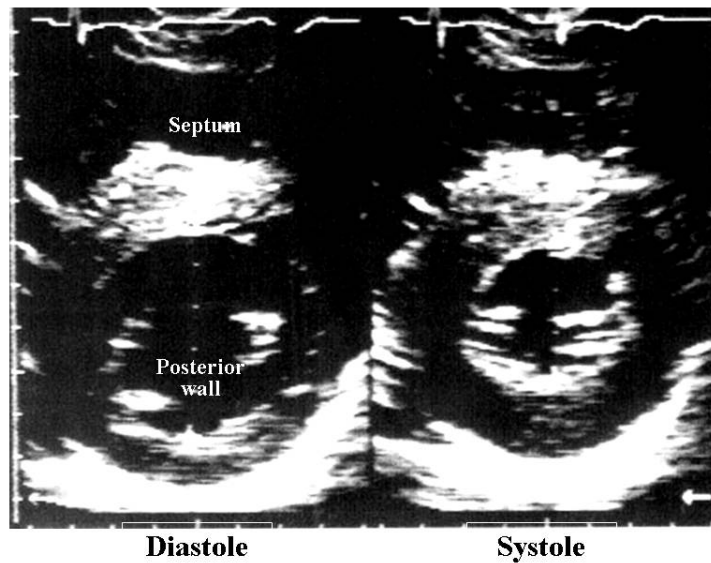


Figure 1. A 16 years old Nigerian football player. A) At the first medical examination in Italy, the ECG revealed high QRS voltages and diffuse negative T waves in limb and precordial leads. B) ECHO showed “abnormal” concentric hypertrophy (septum thickness 17-18 mm, posterior wall 14-15 mm) largely beyond the physiological limits for young football players (<13 mm). Hypertrophic cardiomyopathy was confirmed by cardiac invasive investigation, including



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myocardial biopsy. The athlete was disqualified from competitive football in Italy, but he is playing in another European country.

Fig. 2

A

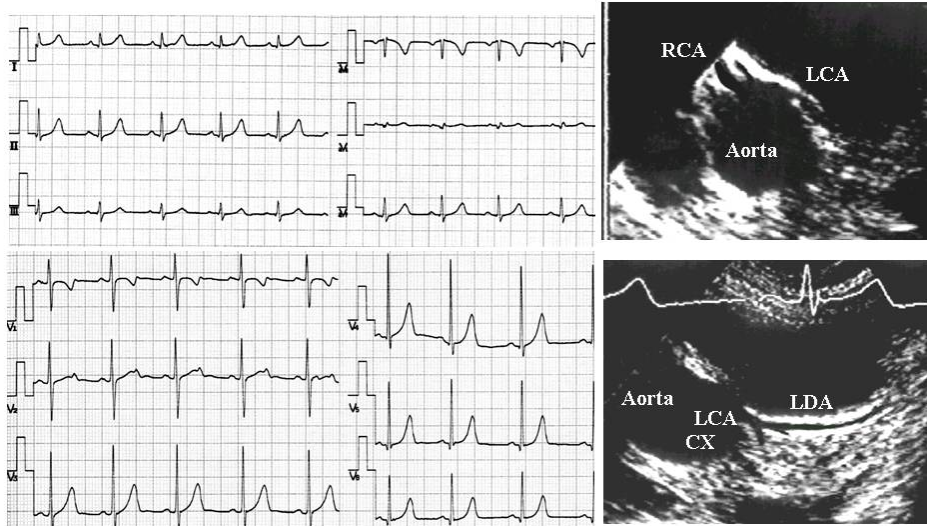


Fig. 2

B

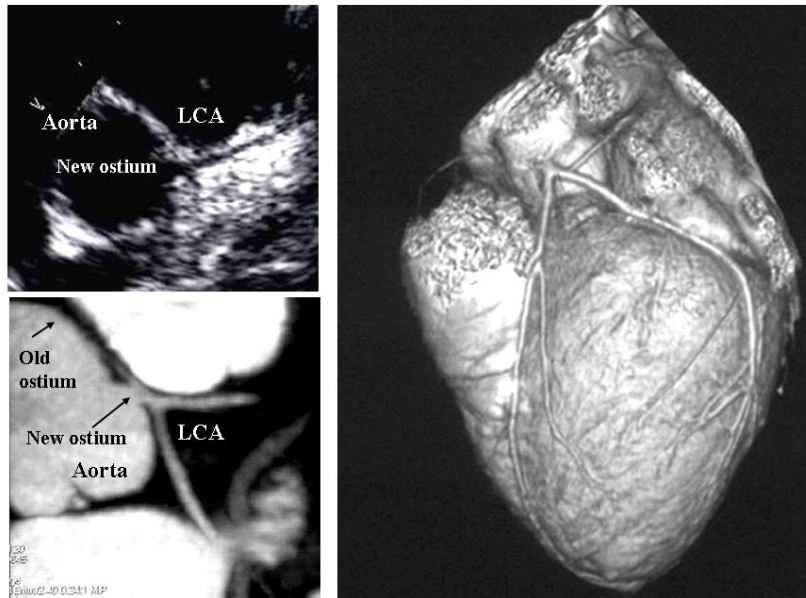


Figure 2. 13 years old Italian football player, referred for an unexplained syncope during a match. A) Rest ECG was completely normal. ECHO suggested an abnormal origin of left coronary artery (LCA) from the right sinus of Valsalva, near the ostium of right coronary artery (RCA). LCA had a course between the aorta and pulmonary artery giving rise to left descending (LDA) and circumflex branch (CX). Abnormal origin and course were confirmed by angio-magnetic resonance imaging and coronary angiography. B) The ECHO (top-left) performed after surgical correction shows the new ostium created by the surgeon in the left sinus of Valsalva. Angio-coronary computed tomography (bottom left and right) clearly shows the new (corrected) origin and course of LCA. The young player resumed recreational football without symptoms over two year of follow-up.

Fig. 3

A

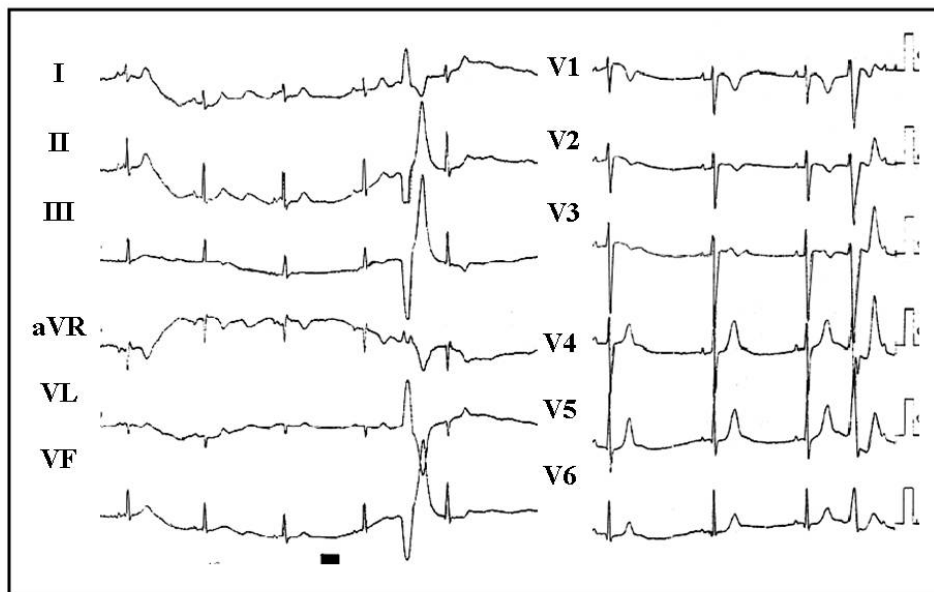


Fig. 3

B

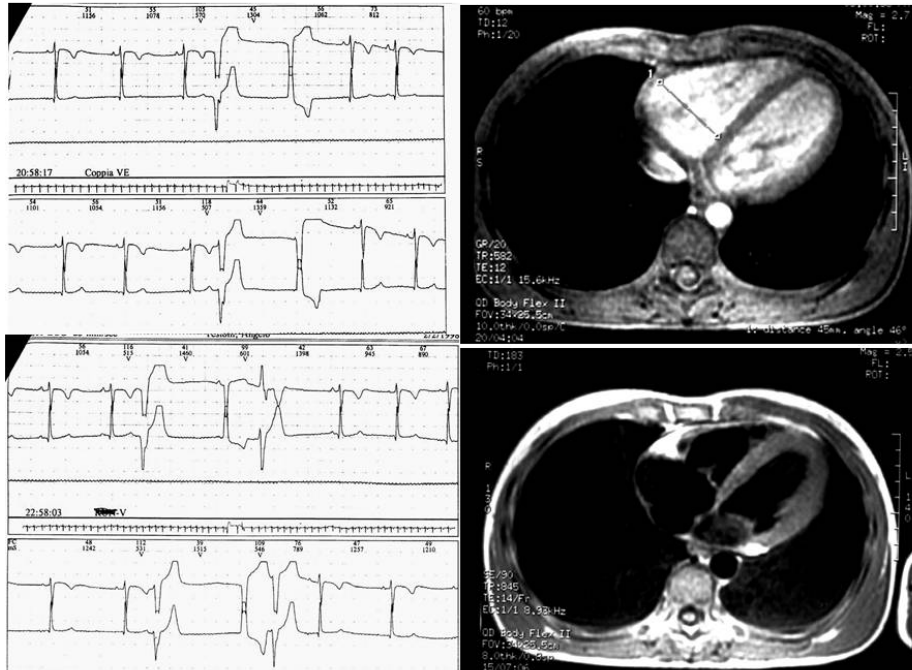


Figure 3. 14 years old Italian football player. A) The rest ECG, at the pre-participation screening, showed negative T waves in right precordial leads and frequent premature ventricular beats (PVB) with left bundle branch morphology and left axis deviation. B) Holter monitoring (left) revealed complex PVBs. Magnetic resonance imaging showed dilation and possible fatty infiltration of the basal wall of right ventricle. Despite the advice of stopping football, he continued playing until died suddenly during a match. Autopsy revealed a severe form of arrhythmogenic right ventricular cardiomyopathy.