Sudden Cardiac Death in Athletes: Still Much to Learn

Joanna Sweeting, BSc, MAppSci (Env. Sci)[a,b],
Christopher Semsarian, MBBS, PhD, MPH, FRACP, FHRS, FCSANZ[a,b,c,*]

KEYWORDS

• Sudden cardiac death • Athletes • Genetics

KEY POINTS

- Sudden cardiac death (SCD) in athletes is a rare but tragic event with an incidence of approximately 1 in 50,000.
- Common causes of SCD in athletes include the structural and arrhythmogenic genetic heart diseases, which are inherited as autosomal dominant traits.
- The process following an SCD in an athlete involves a comprehensive postmortem examination, clinical family screening, and genetic testing.
- It is vitally important to identify, where possible, the cause of death in these cases. If a genetic heart disease is suspected, family members should be thoroughly examined to identify other individuals with disease who are therefore at risk of SCD.
- Many questions remain unanswered, including differences in the causes of SCD in athletes globally, and establishing the best approaches to identify those athletes at highest risk of SCD.

INTRODUCTION

The sudden cardiac death (SCD) of any individual is a tragic event, particularly when there has been no sign of disease before death. In athletes, the impact may be felt more acutely because these individuals are often young and widely thought to be the epitome of health and fitness.[1] This perception is often exploited in media coverage of SCD, further amplifying the impact of these tragedies, not just for the family of the individual but for the entire community.[2,3] Because of the emotive nature of these events, it is imperative that those who encounter the individuals and families affected by these tragedies are well informed of the potential causes and course of action required following an SCD.

To be classified as an SCD, the death must occur within 1 hour from the onset of symptoms in witnessed cases, and within 24 hours of the individual last being seen alive and well in unwitnessed cases.[4] In athletes, SCD can occur at any time during competition, in training, or even occasionally at rest.[5] Other possible causes, such as respiratory, cerebrovascular, and drug-related causes, must be excluded. An athlete may be defined as an individual who engages in regular, intense physical activity through competition and training, with an emphasis on striving to improve and achieve.[6] This definition encompasses paid professional sportspersons as well as university, college, and high school students, and may be extended to include children and adolescents involved in organized sporting activities. Competitive athletes include those who exercise for more than 10 hours per week, whereas those engaged in sports for recreational activities usually participate for less than 10 hours per week.[7]

[a] Agnes Ginges Centre for Molecular Cardiology, Centenary Institute, Locked Bag 6, Newtown, New South Wales 2042, Australia; [b] Sydney Medical School, Edward Ford Building, Fisher Road, University of Sydney, New South Wales 2006, Australia; [c] Department of Cardiology, Royal Prince Alfred Hospital, Missenden Road, Camperdown, New South Wales 2050, Australia
* Corresponding author. Agnes Ginges Centre for Molecular Cardiology, Centenary Institute, Locked Bag 6, Newtown, New South Wales 2042, Australia.
E-mail address: c.semsarian@centenary.org.au

Cardiol Clin 34 (2016) 531–541
http://dx.doi.org/10.1016/j.ccl.2016.06.003

This article briefly summarizes the current literature regarding the incidence of SCD in athletes and the possible underlying causes, with a focus on inherited heart diseases. The course of action following an SCD is discussed with respect to the postmortem examination; genetic testing, such as a molecular autopsy; and the implications for family members, including clinical and genetic screening. In addition, the areas in which there is still much to learn are discussed, including the difficulties associated with postmortem examination and genetic testing in these individuals, and the issue of divergence in the most common causes of SCD in athletes identified in different countries.

INCIDENCE OF SUDDEN CARDIAC DEATH IN ATHLETES

SCD is the most common medical cause of death among athletes, with a wide range of incidence rates reported; from 1 per 3000 to 1 per 917,000, depending on ethnicity and type of sport.[8] A large proportion of available incidence data comes from studies of athletes in the United States, both at high school and college level. A recent article from Harmon and colleagues[9] reported results from a study of 10 years of data from the National Collegiate Athletic Association (NCAA) in the United States. SCD accounted for 79 out of 514 deaths (~1 per 54,000 athlete years) over the 10-year period, with the incidence in men 3.2 times that of women.[9] Basketball players in general were found to have the highest risk of SCD, for both men and women, with a rate of ~1 per 15,000 athlete years. For men participating in division 1 basketball, the incidence increased to 1 per 5200 athlete years.[9] Black athletes were also found to be at higher risk of SCD than white athletes, with a rate of 1 per 21,491 athlete years compared with 1 per 68,354 athlete years. Another study, combining data from the NCAA database and the US National Registry of Sudden Death in Athletes database, found that 64 of 182 sudden deaths (35%) had cardiovascular causes and that cardiovascular deaths were 5 times more common in African American athletes than in white athletes over a period of 10 years.[10] This finding translates to an incidence of 1.2 per 100,000 athlete years, with black athletes again found to have a higher risk of SCD.

Elsewhere in the world similar studies have been conducted in an attempt to establish the incidence of SCD in athletes. A 7-year retrospective study in Denmark, of athletes aged 12 to 35 years, found an incidence rate of 1.2 per 100,000 athlete years or 1 per 82,645 sports-related SCDs in this specific age range.[11] A second retrospective study from Denmark incorporating data from a 3-year period showed a difference in the rate of SCD in athletes depending on age, with a rate of 0.47 per 100,000 athlete years in those aged 12 to 35 years and a rate of 6.64 per 100,000 athlete years for those aged 36 to 49 years.[12] This study also showed that incidence rates in competitive and noncompetitive or recreational athletes were similar and that the rate of SCD in the general population (10.7 per 100,000 person years) was much higher than sports-related SCD. A prospective study in Italy from 1979 to 1999 assessed rates of sudden death in athletes in the same age range of 12 to 35 years.[13] In athletes, the rate of SCD by cardiovascular causes was 2.1 per 100,000 athletes per year, giving an incidence of 1 per 47,600.

There are challenges in determining the true incidence of SCD in athletes because of differences in rates according to age, ethnicity, and chosen sport. Difficulties are also encountered in determining an accurate denominator for calculations and ensuring accurate reporting of sudden death events on databases and through the media.[8] Taking this into account, and based on the worldwide data from these and other retrospective and prospective studies, it is generally accepted that the incidence of SCD in athletes is within the range of 1 to 3 per 100,000, with the rate in the comprehensively studied cohort of NCAA athletes approximately 1 per 50,000.[9,14] Tragically, in many of these cases the first sign of disease is the SCD event, with a limited number of individuals aware of a preexisting heart condition.

CAUSES OF SUDDEN CARDIAC DEATH IN ATHLETES

The causes of SCD in athletes can be dichotomized into acquired and genetic causes. Acquired causes of SCD in athletes include myocarditis and coronary artery disease, in addition to those with external causes such as commotio cordis and drug misuse. In individuals 35 years of age and older, the most commonly identified cause of an SCD event is coronary artery disease, whereas for those aged less than 35 years the genetic heart diseases are more common.[1] The genetic conditions include structural diseases such as hypertrophic cardiomyopathy (HCM) and arrhythmogenic right ventricular cardiomyopathy/dysplasia (ARVC), and arrhythmogenic disorders including long QT syndrome (LQTS) and catecholaminergic polymorphic ventricular tachycardia (CPVT). A summary of the major features of these and other genetic heart diseases is shown in **Table 1**. In all these diseases, high-intensity and competitive physical activity is thought to increase the risk of