Sudden cardiac death in young competitive athletes

ABSTRACT: When an athlete suffers a sudden cardiac death, particularly while participating in a sport being viewed by thousands of spectators, it is a rare and catastrophic event that generates a great deal of discussion among the general public and medical community. Many cardiovascular conditions can predispose an athlete to such an event. The most common causes of sudden cardiac death in athletes are hypertrophic cardiomyopathy and arrhythmogenic right ventricular cardiomyopathy. Other causes include myocarditis, coronary artery anomalies, valvular disease, aortic dissection, commotio cordis, and electrical disorders such as Wolff-Parkinson-White syndrome. Multiple studies have tried to determine the incidence of sudden cardiac death in athletes, but this has proven to be difficult because of differing definitions, inconsistent identification of causes of death, and geographic variation. Further studies are needed to determine the cost-effectiveness of pre-participation screening programs for competitive athletes and to inform strategies that can reduce the burden of sudden cardiac death in this population.

Athletes are thought to represent the pinnacle of health and fitness. When a young athlete suffers a sudden cardiac death (SCD), it is a rare but catastrophic event that garners attention from the media, the general public, and the medical community. SCD in athletes is often the result of lethal arrhythmias caused by genetic disorders that have gone undiagnosed because patients tend to be asymptomatic. In addition to inherited disorders, other risk factors and conditions can predispose athletes to cardiac events.

Risk factors
In competitive athletes younger than age 35 (the focus of this article), the majority of sudden cardiac death cases are due to inherited heart diseases, whereas in masters athletes older than 35, most cases are due to coronary artery disease. More men than women experience SCD, with a male-to-female ratio of 10:1. In the United States, basketball is the highest risk sport followed by football, swimming, lacrosse, and cross-country skiing. In Europe, soccer has the highest incidence of SCD. This may be the result of the greater popularity of certain activities and the varying intensities of different sports and the training required.

Risk can be classified further by levels of play and ethnicity. Athletes participating in more elite divisions and those with an African background appear to have increased risk. The highest risk athletes, irrespective of ethnicity, are National Collegiate Athletic Association (NCAA) Division I male basketball athletes, who have an annual risk of 1:3126. Black male basketball athletes have a greater than threefold increased risk when compared with white male basketball athletes in all divisions. How-

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ever, extrapolation of these results is not straightforward given the limited number of reports in narrowly defined groups and the lack of validation and common data definitions.

During exercise, numerous physiological changes occur that may predispose the susceptible athlete to a malignant arrhythmia and SCD. Intense exercise is associated with increased sympathetic activity and increased cardiac oxygen demand, which may lead to ischemia and increased ventricular ectopy via modulation of cardiomyocyte properties. Similarly, catecholamine surges and electrolyte imbalance may result in myocardial irritability and subsequent ventricular ectopy. In the immediate postexercise period, a reduction in cardiac output and venous return combine with arterial vasodilation to cause a drop in blood pressure. This can lead to a transient reduction in coronary perfusion, and may initiate an ischemic cascade with arrhythmic consequences. Practically speaking, the complex convergence of factors that leads to sudden cardiac death is poorly understood, since athletes are typically exercising at levels common for them, and there is no unique circumstance in the vast majority of cardiac arrests and sudden deaths.

### Incidence

Sudden cardiac death is the leading cause of mortality in young athletes on the playing field. However, the exact occurrence of SCD is unknown and widely debated. Rates differ from one study to another because of varying methodologies for case ascertainment, data definitions, and regions studied. Accurate calculation of incidence requires a reliable number of sudden cardiac deaths (numerator) as well as an exact number of sport participants (denominator). Obtaining data on numbers of SCD has been difficult because of the retrospective nature of most studies, and underestimation has been common because researchers have relied on media reports, catastrophic insurance claims, and other electronic database sources.

Studies conducted in the United States, Italy, Israel, and Canada have found incidence rates ranging from 1:9000 to 1:300 000 deaths per year in young competitive athletes. One of the largest studies examined the incidence of both sudden cardiac arrest and SCD in the US Registry for Sudden Death in Athletes, and estimated that American athletes between age 8 and 39 had a risk of 1:164 000.

In Canada, studies by Pilmer and colleagues have looked at the rates of SCD in athletes and nonathletes age 1 to 19 years and age 2 to 40 years in Ontario. These studies found the highest incidence of SCD beyond early childhood was seen in those age 15 to 19. Most of the SCD events occurred during sleep, but the likelihood of the event occurring during activity increased with age. For those age 2 to 40, subjects younger than 30 were more likely to have a malignant arrhythmia. In this older subgroup, more than 70% of

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**Table 1. Incidence rates for sudden cardiac death from multiple studies.**

<table>
<thead>
<tr>
<th>Study population</th>
<th>Methods and reporting system</th>
<th>Incidence</th>
</tr>
</thead>
<tbody>
<tr>
<td>High school and college athletes age 13–24 years (United States)</td>
<td>Public media reports, other case reports</td>
<td>1:300 000</td>
</tr>
<tr>
<td>Competitive athletes age 12–35 years (Veneto, Italy)</td>
<td>Mandatory registry for SCD</td>
<td>1:27 800</td>
</tr>
<tr>
<td>High school athletes age 13–19 years (Minnesota, United States)</td>
<td>Catastrophic insurance claims</td>
<td>1:200 000</td>
</tr>
<tr>
<td>High school athletes age 14–17 years (United States)</td>
<td>Cross-sectional survey</td>
<td>1:23 000</td>
</tr>
<tr>
<td>All athletes in sportive activity at any level of physical endurance (Israel)</td>
<td>General media reports in two national newspapers</td>
<td>1:38 000 (average yearly)</td>
</tr>
<tr>
<td>SCD in persons age 2–40 years (Ontario, Canada)</td>
<td>Retrospective population-based cohort</td>
<td>0.7:100 000 (2–18 years)/1:142 800</td>
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<tr>
<td></td>
<td></td>
<td>2.4:100 000 (19–29 years)/1:41 700</td>
</tr>
<tr>
<td></td>
<td></td>
<td>5.3:100 000 (30–40 years)/1:18 900</td>
</tr>
<tr>
<td>College athletes age 17–23 years (United States)</td>
<td>NCAA resolutions database, public media reports, catastrophic insurance claims</td>
<td>1:43 770</td>
</tr>
<tr>
<td>Military recruits age 18–35 years (United States)</td>
<td>Mandatory registry, autopsy-based</td>
<td>1:9000</td>
</tr>
<tr>
<td>Competitive athletes age 12–35 years (United States)</td>
<td>Public media reports, other electronic databases</td>
<td>1:166 000</td>
</tr>
</tbody>
</table>
the patients who suffered SCD had underlying structural heart disease, with coronary artery disease being the major contributor, followed by dilated cardiomyopathy. When data were available, SCD was related to exertion in only 9% of decedents age 2 to 40, but increased to 24% in those age 10 to 19.12,13

In a US study, Van Camp and colleagues reported very low incidence rates, which were likely the result of underestimation.8 An Italian study is believed to provide the most reliable results and to support the criteria for an accurate calculation of incidence. The study was prospective in nature and all cases of SCD were recorded in a mandatory registry.1 The researchers found an incidence of 1:28,000 before implementing a screening program that included an ECG, and an incidence of 1:250,000 afterwards (89% decrease). Similarly, a study of military recruits relied on a mandatory registry of deaths with confirmation by autopsy, and discovered an incidence of 1:9,000.13 An Israeli study found a sudden cardiac death rate similar to that of the Italian study, with an incidence of 1:38,000.11 Although the Italian and Israeli rates are similar, there is a strong possibility of underestimation in the Israeli incidence rate because the data used came only from two national newspapers.

A study of NCAA athletes provided a higher incidence rate (1:43,770)14 than some of the initial reports in the United States—a rate that came closer to that in the Italian study, perhaps because the NCAA used data for athletes age 17 to 23 playing 40 different sports. This rate is still an imperfect estimate, however, because it is based solely on media reports and insurance cases. Drezner and colleagues were the first to examine the rate of SCD in a cross-sectional study and found an incidence of 1:23,000, consistent with the rate in the Italian study.10 The variability in reporting sudden cardiac deaths makes comparing data difficult and further emphasizes the need for agreed-upon data definitions and a reliable reporting system.

### Cardiovascular causes of SCD

The consensus definition of a competitive athlete from the Summit on Electrocardiogram Interpretation in Athletes is “an individual who engages in regular exercise or training for sport or general fitness, typically with a goal of improving performance.”16 In addition to SCD, other common causes of sudden death that may occur in young athletes should be considered. A recent study of sudden death in student athletes found that in approximately 65% of cases the cause was not cardiovascular in nature but involved suicide, trauma, or drug use. The number of drownings and heat strokes causing sudden death was also not insignificant.5

The leading reported cause of SCD in the United States is hypertrophic cardiomyopathy (HCM), while in Italy the leading cause is arrhythmogenic right ventricular cardiomyopathy (ARVC). Other causes of SCD include myocarditis, coronary artery anomalies, valvular disease, aortic dissection, commotio cordis, and electrical disorders such as Wolff-Parkinson-White (WPW) syndrome, long QT syndrome (LQTS), and Brugada syndrome (see Table 2). In addition, SCD may occur in up to 12% of young patients in the absence of any apparent structural heart disease.21 These cases are presumed to involve primary arrhythmia disorders that are often inherited, and there is evidence that genetic testing of the SCD victim may yield informative results that explain the event and can be useful for family screening.

### Hypertrophic cardiomyopathy

Hypertrophic cardiomyopathy is primarily a familial condition with an incidence estimated to be about 1:500 in athletes.22 HCM commonly involves a mutation in genes that encode sarcomeric proteins. The presentation of HCM is varied, ranging from left ventricular hypertrophy seen on imaging, exertional syncope, and ventricular arrhythmias without warning.23 SCD from HCM is usually secondary to ventricular tachycardia or ventricular fibrillation, but syncope may result from left ventricular outflow tract obstruction leading to decreased cerebral perfusion. Subendocardial ischemia may also develop and cause chest pain. The diagnosis is usually made on echocardiography, but classic signs can be found on

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**Table 2. Common causes of sudden cardiac death in athletes.**

<table>
<thead>
<tr>
<th>Young competitive athlete</th>
<th>Masters athlete</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>• Hypertrophic cardiomyopathy</strong></td>
<td><strong>• Coronary artery disease</strong></td>
</tr>
<tr>
<td><strong>• Arrhythmogenic right ventricular cardiomyopathy</strong></td>
<td><strong>• Dilated cardiomyopathy</strong></td>
</tr>
<tr>
<td><strong>• Idiopathic dilated cardiomyopathy</strong></td>
<td><strong>• Hypertensive heart disease</strong></td>
</tr>
<tr>
<td><strong>• Myocarditis</strong></td>
<td><strong>• Myocarditis</strong></td>
</tr>
<tr>
<td><strong>• Coronary artery anomalies</strong></td>
<td></td>
</tr>
<tr>
<td><strong>• Aortic dissection</strong></td>
<td></td>
</tr>
<tr>
<td><strong>• Commotio cordis</strong></td>
<td></td>
</tr>
<tr>
<td><strong>• Arrhythmic heart disease</strong> (including Wolff-Parkinson-White syndrome, long QT syndrome, short QT syndrome, Brugada syndrome, and catecholaminergic polymorphic ventricular tachycardia)</td>
<td></td>
</tr>
</tbody>
</table>
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Physical exam. These signs include systolic murmur that increases from squat to stand, brisk carotid impulse, and prominent apical pulsation. Furthermore, common ECG findings include a left ventricular hypertrophy pattern, prominent Q waves, or deeply negative T waves. At present, given the phenotypic variability of HCM, the 2005 Bethesda Conference guidelines and the 2011 American College of Cardiology Foundation/American Heart Association guidelines recommend that patients with a probable diagnosis of HCM should be excluded from competitive sports, with the possible exception of low-intensity sports such as billiards, golf, and bowling. These recommendations are irrespective of age, symptoms, degree of obstruction, and the presence or absence of an implantable defibrillator. However, there is no evidence to suggest that patients with genotype-positive but phenotype-negative HCM should be excluded from sporting activities.

Arrhythmogenic right ventricular cardiomyopathy
While HCM is the most common cause of SCD in the United States, a 21-year prospective study found arrhythmogenic right ventricular cardiomyopathy was the most common cause of SCD in young people in the Veneto region of Italy. This finding, which may be due in part to greater recognition of ARVC through advances in imaging and genetic screening, has not been reproduced in other studies, particularly in North American populations, suggesting the variability of the causes of SCD in different ethnic groups and populations.

ARVC is generally an inherited disorder whereby the right ventricular myocardium is gradually replaced by fibro-fatty tissue that can subsequently lead to malignant ventricular arrhythmias and SCD. The risk of SCD may increase as much as five-fold when patients with ARVC participate in competitive sports. Patients with ARVC are more likely to have preceding symptoms of palpitations and presyncope. If these symptoms are found in conjunction with ECG changes such as T wave inversion in leads V1 to V3, an epsilon wave in V1 or V2, or premature ventricular contractions with a left bundle branch block pattern, ARVC must be strongly suspected. Due to the nonspecific nature of ARVC, magnetic resonance imaging is often recommended to aid in diagnosis.

As in athletes with HCM, athletes with ARVC should be advised to avoid competitive sports with the possible exception of low-intensity activities. Recent compelling evidence from both animal models of accelerated disease and family surveys supports the assertion that exercise is an accelerant in individuals predisposed to ARVC.

Myocarditis
Myocarditis is an inflammatory condition commonly caused by a viral infection that leads to cardiac dysfunction, and may account for up to 7% of SCD cases related to athletics. and may range from benign effects to heart failure requiring heart transplantation. The inflammation may resolve spontaneously but may also require standard therapies for heart failure. Patients can return to normal activity 6 months after clinical presentation as long as ECG and echocardiogram results and serum marker levels normalize.

Coronary artery anomaly
Congenital coronary artery anomalies account for up to 20% of SCD in athletes, and in the United States these anomalies are the second most common cause of SCD in young athletes. The most common anomaly occurs where the left main coronary artery arises from the right sinus of Valsalva, coursing between the pulmonary artery and aorta. Although most patients are asymptomatic, syncope, palpitations, and angina may occur. When such
Marfan syndrome. In general, athletes with Marfan syndrome without evidence of aortic root dilatation (generally greater than 40 mm), significant mitral regurgitation, or a family history of aortic dissection or SCD can participate in low and moderate static/low dynamic sports, but decisions need to be made on an individual basis.23,25

**Aortic dissection/ Marfan syndrome**

Aortic dissections are more likely in patients with Marfan syndrome, a connective tissue disorder inherited in an autosomal dominant pattern and characterized by multi-organ dysfunction. Common systems affected include the musculoskeletal, ocular, and cardiovascular systems.20 From a cardiovascular perspective, the aortic root may dilate abnormally and lead to rupture or dissections. In addition, patients may develop mitral valve prolapse and progressive ventricular dysfunction leading to an increased risk of arrhythmias.

Marfan syndrome may be unrecognized until a family member is diagnosed or a clinical presentation leads to diagnosis. Hundreds of mutations affecting the gene encoding fibrillin-1 result in an estimated prevalence of 1:5000 to 1:10000 for Marfan syndrome.23 Given that tall stature is a phenotypic trait of the syndrome, it is not surprising that athletes with Marfan syndrome are overrepresented in sports such as volleyball and basketball. Echocardiography is used to measure aortic dimensions and mitral valve kinetics, and to provide follow-up measurements once an abnormality is identified. In general, athletes with Marfan syndrome may be unrecognizable until a family member is diagnosed or a clinical presentation leads to diagnosis. Hundreds of mutations affecting the gene encoding fibrillin-1 result in an estimated prevalence of 1:5000 to 1:10000 for Marfan syndrome.23

**Commotio cordis**

Commotio cordis means “concussion of the heart” and is a rare condition whereby a mechanical blow to the precordium leads to an arrhythmia.23,30 These cases typically affect athletes who participate in contact sports, with 10 to 20 cases being reported annually.31 The timing of the blow is critical as it must occur on the upstroke of the T wave as seen on an ECG, which accounts for only 1% of the entire cardiac cycle. At present, there are no effective screening tests to identify individuals susceptible to commotio cordis. Instead, the focus has shifted to preventing the condition by promoting the use of chest protectors and softer age-appropriate equipment. Resuscitation attempts must be made promptly, otherwise the survival rate for commotio cordis is low, with the best estimate of survival being 35%.23,31 Given the rarity of this condition there are no consensus guidelines, and survivors of commotio cordis must be assessed for a return to competition on an individual basis.23

**Arrhythmic heart disease**

Several conditions that affect the electrical system of the heart can lead to SCD in athletes. Patients with Wolff-Parkinson-White syndrome have an accessory pathway that leads to pre-excitation and potential arrhythmias. The prevalence of WPW in athletes ranges from 0.1% to 0.3%, which is comparable to prevalence in the general population. On rare occasions, a benign re-entrant arrhythmia degenerates into atrial fibrillation with a very rapid ventricular rate conducted over the accessory pathway, triggering ventricular fibrillation and cardiac arrest.32 Characteristic ECG findings include pre-excitation and a short PR interval.30 Symptoms are common and patients often undergo curative radiofrequency ablation of the accessory pathway. For those who are asymptomatic, evaluation by an electrophysiologist is recommended, with restriction from sport typically applying to those who have concurrent atrial fibrillation or those who have a short refractory period.23 Conversely, asymptomatic athletes without any structural heart disease can likely be permitted to participate in all sports.

Long QT syndrome may be responsible for up to 8% of SCD cases. A long QT interval can be caused by many extrinsic factors, including medication use, but it may also be hereditary and caused by mutations in genes related to cardiac ion channels.23 There is a prolonged ventricular repolarization period that may lead to ventricular tachycardia and sudden cardiac death. Congenital long QT syndrome is present in 1:2500 individuals, half of whom will have a normal or borderline resting QT interval.31 The most common form of LQTS (type 1) is associated with exertion, particularly swimming. Exercise testing is the diagnostic tool of choice when long QT syndrome is suspected.33 An individual discussion with a cardiologist or electrophysiologist is warranted for patients with long QT intervals.23 While guidelines differ regarding allowable activities for patients with LQTS, patients are generally restricted to low-intensity activities. However, there is no evidence to restrict patients with genotype-positive but phenotype-negative
LQTS from competitive sports.25

Brugada syndrome is a channelopathy affecting cardiac sodium channels. The syndrome can lead to ventricular fibrillation and is seen commonly in people of Southeast Asian ancestry. In European studies of the general population, the incidence of SCD is 1.34 per 100,000 per year, and some of these cases are known to involve inherited arrhythmia conditions such as Brugada syndrome.34 ECG findings for the syndrome show characteristic elevations of the ST segment in the anterior leads that are followed by negative T waves.23 With the newer ECG criteria, there are two characteristic patterns of Brugada syndrome: the classic and more concerning type 1 coved pattern, and the combined previously designated type 2 and 3 saddleback patterns.35 Individuals with Brugada syndrome more commonly experience SCD during sleep, which is thought to be related to increased nocturnal vagal tone.36 There is also an association between Brugada syndrome and hyperthermia.37 The Bethesda Conference recommendations limit athletes with Brugada syndrome to low-intensity activities regardless of whether the patient has an implantable cardioverter defibrillator.25 This is a matter of contention and not fully adhered to by experts in the field, given that most arrhythmic events occur during sleep rather than during activity.34

Conclusions

Sudden cardiac death in young athletes is fortunately a rare event. Worldwide, hypertrophic cardiomyopathy and arrhythmogenic right ventricular cardiomyopathy are the most common causes. Multiple studies have tried to determine the incidence of SCD in athletes, but this has proven to be difficult because of differing definitions, inconsistent identification of causes of death, and geographic variation. An accurate incidence rate for SCD is needed to determine the cost-effectiveness of pre-participation screening programs for competitive athletes. Without an accurate incidence rate, pre-participation screening in the young competitive athlete will remain a controversial topic within international sport and medical societies. We cannot, however, ignore the fact that athletes with unrecognized heart disease may increase their risk of SCD by participating in sport. Further studies are needed to inform strategies that can reduce the burden of SCD in this population.

Competing interests

None declared.

References

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