Sudden Cardiac Death

Sudden Cardiac Death in the Athlete
Bridging the Gaps Between Evidence, Policy, and Practice

Mark S. Link, MD; N.A. Mark Estes III, MD

Sudden cardiac death (SCD) in a young athlete commonly brings to the forefront the many gaps in knowledge regarding how to predict and prevent these rare tragic events.1–3 Although the number of athletic sudden deaths is relatively small, with ≈100 to 150 competitive deaths during sports in the United States annually, they represent an important and emotionally charged public health issue, perhaps out of proportion to the relative risks of other pediatric deaths (Figure 1).1–5 Despite identification of the cardiovascular conditions that predispose to these events, much remains unknown regarding many fundamental issues related to athletic sudden death. The precise frequency with which these events occur in the United States remains unclear because of the absence of athletic death registries (and indeed any SCD registry) with mandatory reporting requirements.1–8 In fact, whether these events are more common in athletes is not at all certain. Additionally, significant gaps in evidence exist related to effectiveness of preventing sudden death in the athlete with preparticipation screening strategies. There are many limitations to the available evidence supporting the notion that athletic restriction improves outcomes. The effectiveness of cardiopulmonary resuscitation and automated external defibrillator (AED) programs, evident in casinos and airports, has not necessarily been shown in athletes.9 The ongoing debates related to prediction and prevention of athletic sudden death persists because the standards of evidence-based medicine have not been fulfilled with appropriately designed randomized controlled trials.

Is the Risk of SCD Higher in the Athlete?

On average, every 3 days in the United States a competitive athlete experiences a SCD, and many of these deaths are nationally noted. However, this same intense media speculation is not given to a nonathlete who experiences SCD or indeed to a competitive athlete who has SCD off the athletic field. On the basis of media reports it would appear that SCD deaths in athletes are much more common than in nonathletes. However, the data supporting such a claim have many limitations. In fact, the only comparative data supporting the increased risk of SCD in athletes comes from the Padua, Italy region, in which they found the risk of SCD in athletes (2.3 in 100 000 per year) was higher than that in nonathletes (0.9 in 100 000 per year).7 Earlier studies have shown an increased risk of SCD in basketball players, with rates as high as 3.6 per 100 000 person-years in male basketball players.10 More recently, this was confirmed in a survey of deaths in National Collegiate Athletic Association colleges in which the risk of SCD in male Division I basketball players was as high as 1 in 3100 person-years.11

This increased risk of SCD in athletes is not seen in other epidemiological series, in which the general population often has a higher risk of SCD in comparison with an athletic population (Table 1).6,10,12–17 In a comprehensive study of 11 emergency department systems in the Resuscitation Outcomes Consortium, the incidence of sudden cardiac arrests in all children under age 19 is 8 per 100 000 patient-years. There are many more sudden cardiac arrests in those under age 1 (72 per 100 000 person-years, likely because of Sudden Infant Death Syndrome), but still a significant number in those between 1 and 11 years (3.73 per 100 000 person-years), and those between 12 and 19 years (6.37 per 100 000 person-years).17 With a US pediatric population of >79 million (US Census data, 2009: www.census.gov/popest/national/asrh/NC-EST2009-sa.html), one could extrapolate these data to an incidence of 3000 and 5000 sudden cardiac arrests per year in the US population between ages 1 and 19 years. It is estimated that 100 to 150 of these sudden cardiac arrests occur in competitive athletes who have their arrest during sports.18 Thus, it appears that only a small fraction of the SCDs in the young occur during competitive athletics.

Data from other countries are similar to those in the United States and Canada. In Hamburg, Germany, there were 48 335 fatalities between 1997 and 2006.19 Of those, only 176 were identified as sports related. In Denmark, there were 5662 deaths in individuals between 12 and 35 years in the years between 2000 and 2006.14 The number of competitive athletes between the ages of 12 and 35 was estimated to be 177 070. Fifteen cases of SCD in competitive athletes during or within 1 hour of sports activity were found, corresponding to an incidence rate of 1.21 per 100 000 athlete person-years, much lower than the 3.76 per 100 000 person-years in the general population. In Japan, a national screening program for all students was instituted in 1973, and the results were published in 2006.15 After this screening program was insti-
tuted, the incidence of SCD was 1.32 per 100,000 person-years. In military recruits, a sudden death rate of 13 per 100,000 person-years has been documented from an autopsy series.16

Thus, much of the data indicate a similar or lower incidence of SCD in athletes compared to nonathletes, although the data are far from conclusive. This arises from such fundamental issues as lack of standard definitions of a competitive athlete and athletic SCD. The lack of mandatory reporting requirements for SCD also hinders accurate assessment of the incidence of athletic SCD. Accurate incidence and prevalence data are essential for assessing strategies for primary prevention through preparticipation screening and secondary prevention with emergency response systems. It is evident that precise data are essential to assess the efficacy of both primary and secondary prevention. Although the underlying causes of sudden death in the athlete are known, the frequency with which sudden death occurs remains to be precisely defined by sex, age, race, nationality, and sport. Currently in the United States, the incidence is derived from identification of cases from media reports and estimated participation rates. Accordingly, the precise number of deaths (numerator) is unknown as is the exact number of athletic participants (denominator).

Screening Athletes and the Prevention of SCD

Although there is universal acceptance of the goal of prevention of sudden death in the athlete, outcomes, risks and benefits of screening, and cost data remain sufficiently incomplete to resolve ongoing debates related to screening and intervention strategies.5,20–28 Only with additional research to address these knowledge gaps can one of the fundamental tenets of evidence-based medicine be fulfilled by ensuring that healthcare policy and practice, including screening, is based on robust evidence from appropriately designed clinical trials.27

Development of an effective preparticipation screening strategy with a sensitivity and specificity to detect cardiovascular conditions predisposing to SCD in athletes is an intuitively appealing public health priority. One major objective of preparticipation athletic screening is detection of potentially lethal cardiovascular diseases likely to manifest with SCD during athletics. However, identification of a potentially lethal disease is only important if an effective preventive strategy reduces the risk of SCD. Subsequent intervention in a fashion that unequivocally improves outcomes is a critical aspect of screening. Identification of cardiac abnormalities is not the only criterion for benefit of screening; there must be the ability to modify risk and outcome for screening to be effective.

In Italy, a 12-lead ECG and limited stress test is routinely obtained as part of a mandatory comprehensive screening program.6,28 This national screening program with ECGs, stress tests, and selective use of echocardiograms has been performed at regional centers by highly specialized physicians with legal liability for errors since 1982. This federally mandated and funded systematic screening, coupled with athletic restriction, has been associated with a decline in deaths.6,28 Fundamental to 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. Accordingly, a higher priority is assigned to the detection of cardiovascular disease in athletes in comparison with nonathletes. With this screening, 2% of athletes were excluded from

![Figure 1. Causes of death in the US population aged 1 to 21 years. Injury, homicide, and suicide dwarf the number of sudden deaths in athletes. SCD indicates sudden cardiac death. Data from the Centers for Disease Control and Prevention (http://webappa.cdc.gov/sasweb/ncipc/leadcaus10.html).](http://circ.ahajournals.org/)

<table>
<thead>
<tr>
<th>Country</th>
<th>Reference</th>
<th>Population</th>
<th>Years</th>
<th>Incidence per 100,000 Person-Years</th>
</tr>
</thead>
<tbody>
<tr>
<td>Italy</td>
<td>Corrado6</td>
<td>Athletes</td>
<td>1980–1981</td>
<td>3.6</td>
</tr>
<tr>
<td>Italy</td>
<td>Corrado6</td>
<td>Athletes</td>
<td>2007–2008</td>
<td>0.4</td>
</tr>
<tr>
<td>Israel</td>
<td>Steinvil12</td>
<td>Athletes</td>
<td>1985–1997</td>
<td>2.54</td>
</tr>
<tr>
<td>Israel</td>
<td>Steinvil12</td>
<td>Athletes</td>
<td>1998–2009</td>
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</tr>
<tr>
<td>United States</td>
<td>VanCamp10</td>
<td>Athletes</td>
<td>1983–1993</td>
<td>0.33</td>
</tr>
<tr>
<td>United States</td>
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<td>Athletes</td>
<td>1985–2006</td>
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</tr>
<tr>
<td>Denmark</td>
<td>Holst14</td>
<td>Athletes</td>
<td>2000–2006</td>
<td>1.21</td>
</tr>
<tr>
<td>Denmark</td>
<td>Holst14</td>
<td>All children</td>
<td>2000–2006</td>
<td>3.76</td>
</tr>
<tr>
<td>Japan</td>
<td>Tanaka15</td>
<td>All children</td>
<td>1989–1997</td>
<td>1.32</td>
</tr>
<tr>
<td>United States and Canada</td>
<td>Atkins17</td>
<td>All children ages 1–11</td>
<td>2005–2007</td>
<td>3.73</td>
</tr>
<tr>
<td>United States and Canada</td>
<td>Atkins17</td>
<td>All children ages 12–19</td>
<td>2005–2007</td>
<td>6.37</td>
</tr>
</tbody>
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*There is a wide range of incidences of sudden death in both athletes and the general population.*
sports participation. This long-term evaluation of the epidemiology of SCD in Italian athletes demonstrated, concurrently with this screening and restriction strategy, a decrease in SCD comparing the athletic sudden death rate in the 2 years before the implementation of screening (3.6 per 100,000 person-years) with the rate 2 decades later (0.4 per 100,000 person-years).

Whether the Italian experience can extend to a more diverse population, especially one with a multitude of races and ethnicities remains, as yet, unproven. Two recent studies from the United States and Israel raise questions about improved outcomes with screening and athletic restriction (Table 2). Using a similarly sized but much more heterogeneous cohort of athletes who did not undergo ECG screening, Maron and colleagues derived mortality rates for student athletes over a 23-year period. This study of high school athletes demonstrated a relatively constant rate of sudden death that was similar to the rate noted by the Italian study after 2 decades of screening and restriction.

An analysis of a mandated screening program of all Israeli athletes with ECG and stress tests also assessed the rate of sudden death among competitive athletes over a 24-year period. The average yearly rate of sudden death or cardiac arrest was 2.6 per 100,000 person-year period of observation. The respective averaged yearly incidence during the decade before and after the mandated screening was 2.54 and 2.66 events per 100,000 person-years, respectively ($P=0.88$).

Importantly, had the 2-year period before the implementation of screening in Israel been used as the baseline, the results would have reached conclusions similar to the Italians. However, these 2 years were characterized by an anomalously high sudden death rate not representative of the decade that preceded the screening in Israel. It was this spike in SCD in athletes that directly led to the establishment of a national screening program. Thus, in both the Israeli and Italian screening programs, the possibility that these baseline rates were unusually high and that the subsequent rates represent regression to the mean or the methodological flaw of immortality bias common in observational trials cannot be excluded.

Despite these conflicting data, an increasing number of countries and organizations, including the International Olympic Committee, now mandate medical preparticipation screening that includes a baseline ECG with the intent of detecting cardiovascular conditions that predispose to sudden death. Restriction of athletic activity is then mandated based on the premise that this restriction reduces the incidence of sudden death.

In the final analysis, the incremental utility of screening with the ECG in comparison with screening with the history and physical examination alone remains debatable because of the absence of sufficiently robust evidence in different populations to conclusively resolve the issue. Furthermore, the evidence regarding athletic screening and restriction as a strategy to reduce incidence of sudden death is limited to 3 observational trials. Passionate, emotional, and compelling arguments have been advanced for implementation of national screening programs such as the Italian approach. However, these arguments have not yet met many fundamental tenets of evidence-based medicine. These tenets include ensuring that clinical practice, including screening, is based on robust data from appropriately designed clinical trials. Screening is appropriate when several conditions are satisfied. Sensitive, specific, practical, cost-effective screening strategies for a condition that has significant morbidity or mortality are required. The condition being screened for should have a proven treatment that can affect its outcome and not merely prematurely identify the inevitable. Treatment afforded by early detection should produce superior results to the early treatment of symptomatic results. A final requisite is that there must be a sufficiently high prevalence of the condition in the population to merit screening in a cost-effective fashion. Based on these considerations, it is evident that there remain considerable gaps in the evidence supporting the notion that preparticipation screening with an ECG and athletic restriction reduce sudden deaths in athletes.

### Table 2. Preparticipation Athletic Screening and Athletic Restriction in Italy, the United States, and Israel

<table>
<thead>
<tr>
<th>Country</th>
<th>Years</th>
<th>Screening</th>
<th>Initial</th>
<th>Examiners</th>
<th>Sudden Death</th>
</tr>
</thead>
<tbody>
<tr>
<td>Italy</td>
<td>1981–2008</td>
<td>Mandatory</td>
<td>History, PE, ECG, ETT</td>
<td>Sports medicine MD</td>
<td>Decrease</td>
</tr>
<tr>
<td>United States</td>
<td>1985–2006</td>
<td>Recommended</td>
<td>History, PE</td>
<td>MD and non-MD</td>
<td>No decrease</td>
</tr>
<tr>
<td>Israel</td>
<td>1985 to 2009</td>
<td>Mandatory</td>
<td>History, PE, ECG, ETT</td>
<td>Certified MD</td>
<td>No decrease</td>
</tr>
</tbody>
</table>

PE indicates physical examination; ETT, exercise tolerance test.

**Inherent Risk in Sport**

It is evident that SCD is not the sole cause of death in sports. In a National Collegiate Athletic Association survey recently published, 51% of deaths were accidental (Figure 2). In addition, 9% of the deaths were suicides, 8% homicides, a
combined number higher than cardiac causes (16%). In addition, there are sports in which there is a high risk of injury or death intrinsic to the sports, yet society allows these sports. These sports include mountaineering, hang gliding, parachuting, boxing, mountain hiking, scuba diving, and even swimming. One of the leading causes of death in the United States in the pediatric population is drowning. In 2006 ≈ 11 000 individuals <20 years of age died of drowning.33 Drowning and near drowning is also a common cause of death and injury in other countries. In Victoria, Australia, the rates of death were 0.6/100 000 participant-years.34 In Denmark, there is an incidence of 0.7/100 000 years of childhood drowning.35 In Singapore, the drowning rate ranged from 0.88/100 000 per year to 1.72/100 000 per year.36 Thus, drowning is a major cause of death in the United States and other countries and has an incidence quite similar to the incidence of SCD in athletes.57

In mountaineering, there is a very high rate of death due predominantly to falls. Indeed, the American Alpine club has documented 1373 fatal accidents between 1951 and 2003.38 In more extreme climbs, it has also been shown that with mountain hiking above an altitude of 7000 m there is a mortality rate of 4.3 per 100 mountaineers.39 The mortality rate for mountaineers above base camp in Mt. Everest is 1.3%.40,41 In less extreme sports such as recreational hiking the mortality rate is 4 per 100 000 annually.42

Marathons have also been associated with sudden death. In an analysis of US marathons with >1000 participants for the years 1975 to 2005, 26 sudden deaths were observed for a participant rate of 0.8/100 000.42 In the London Marathon with 650 000 runners there were 8 cardiac arrests for a death rate of 1 in 8000.43 Deaths are also common with all-terrain vehicles and bicycle crashes.45 Bicycle accidents are estimated to kill ≈ 400 children per year.

Although there are fundamental differences in accidental and traumatic deaths related to sports, these data provide an important context for considering many of the issues related to athletic SCD. Some sports such as basketball, football, and tennis may have a higher risk of SCD in comparison with traumatic death. By contrast, other sports such as mountaineering, swimming, biking, and skiing have a higher risk of traumatic death in comparison with SCD. Individuals and society are frequently uninformed of the cardiac and noncardiac mortality risks inherent in sports. Society does not prohibit these high-risk sports even though death risk in these sports is quite high. Indeed, an individual with hypertrophic cardiomyopathy playing basketball likely has a risk of sudden death that is less than that of an individual who climbs mountains. Currently, there are multiple perspectives on an individual’s autonomy in deciding to undertake athletic activity that may be fatal.

Public Access to Defibrillation and Emergency Action Programs

Public access to defibrillation and emergency action programs are a critical component to sports programs and too often are not taken seriously enough.46 Emergency action programs encompass not only cardiac events, but trauma, asthma, and any other kind of medical emergency. Relying on public emergency programs (ie, emergency medical system) is unlikely to improve resuscitation because athletic facilities are often distant from public roads.46 Data supporting AEDs and early action programs comes from casinos,47 airlines,48 and cities.49 Yet data to support that AED and emergency action programs will have an appreciable effect on resuscitation in athletes are currently limited to retrospective data. Early data demonstrated a very poor survival of National Collegiate Athletic Association athletes in which only 1 of 9 athletes was resuscitated.50 It is not clear how advanced the emergency action programs were at those schools. However, it appears that survival is improving, both in athletes4 and nonathletes.51 Essential elements of a emergency action plan include training and certification of athletic trainers and sports participants (a minimum of basic life support and AEDs). Other essential components include prompt access to AEDs, an institutional plan of how to activate the emergency medical system quickly, integration of the on-site responders and emergency medical system, and finally practice and review of the response plan.46 One could argue that knowledge of how to perform basic cardiopulmonary resuscitation and to operate an AED is a critical component of school health requirements. All individuals, especially those involved in competitive athletics, should know the basics of resuscitation.52 It is likely that recognition of SCD and prompt activation of the emergency medical response by all students and athletes would increase the survival rate in SCD.

Potential Negative Consequences of Prohibiting Competitive Athletics

The interest in athletic sudden death arises in part from both the undue importance of sports in most societies and the paradox that physical activity can have both a positive and negative impact on an individual’s health.5,53 It is clear that vigorous exertion transiently increases the risk of SCD in individuals with cardiovascular disease.5,54 It is evident also, but frequently overlooked, that one unintended consequence of athletic screening is depriving athletes of the many benefits of exercise as a consequence of prohibition of competitive athletics.5,53 There are deleterious physical and psychological sequelae of athletic restriction. There are multiple studies that demonstrate the benefits of continued athletics, and, in general, the more vigorous the continued activity the greater the long-term benefits.5,53 Indeed, it has been argued that low cardiovascular fitness constitutes the largest attributable risk for all-cause mortality.55 Nearly all epidemiological studies show a decreased risk of overall mortality, cardiac mortality, and the development of coronary artery disease with regular exercise.53,55,56 Indeed, American Heart Association physical activity guidelines call for 30 minutes of moderate-intensity aerobic activity 5 days a week or vigorous-intensity aerobic activity 3 days a week.57

In addition to a reduction in physical disease, there is also evidence that sport participation, including but not restricted to competitive sports, reduces the risk of suicide.58,59 In 2007, there were 2358 suicides in the United States in the age group from 1 to 21 (http://webappa.cdc.gov/sasweb/ncipc/leadcaus10.html). Data from the Centers for Disease Control and Prevention show that suicide is the third leading
cause of death in children and adolescents (after injury and homicide) (Figure 1).

Further research is needed to assess both the physical and psychological consequences of restriction from athletic activity. This issue merits careful consideration when considering athletic screening programs. Given the prevalence of cardiovascular conditions predisposing to SCD and the current specificity of screening strategies, it is likely that a significant number of athletes would be unnecessarily restricted having been falsely identified as having a cardiac condition predisposing to SCD.

Putting it All Together

SCD is a very real public health issue and personal issue for not only the athlete, but also for the nonathlete. SCD in a young individual is devastating. Appropriate measures of prevention and treatment are critical. However, based on the best available evidence, it is evident that measures conclusively effective in reducing athletic SCD remain unknown. Nearly all available data are limited by their observational nature. Standard definitions of competitive athletes and athletic sudden death are needed. Robust national registries of athletic sudden death are also essential. Randomized trials of screening and restriction prospectively collecting cost and outcomes data are needed. The ethics of selectively screening athletes for cardiovascular conditions that predispose to SCD without broader screening of the nonathletic population merit careful consideration. Additional studies are needed to evaluate the long-term consequences of athletic restriction. The considerable controversy related to many of the issues regarding athletic sudden death stems from the lack of evidence to resolve them. Experience indicates that when there is disagreement among experts, there is a dearth of reliable data. The best available data indicate that the total number of athletic deaths is relatively small. Furthermore, proven strategies to prevent athletic sudden death are not currently available. Based on these considerations, the most reasoned approach currently is advancing the available evidence by obtaining data from appropriately designed studies rather than prematurely advancing well-intended yet unproven strategies.

Disclosures

Dr Estes has received payment as a consultant from Boston Scientific (modest). Dr Link reports no conflicts.

References


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