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Incidence of sudden cardiac death in athletes: a state-of-the-art review

Kimberly G Harmon,[1] Jonathan A Drezner,[2] Mathew G Wilson,[3] Sanjay Sharma[4]

[1]Departments of Family Medicine and Orthopaedics and Sports Medicine, University of Washington, Seattle, Washington, USA
[2]Department of Family Medicine, University of Washington, Seattle, Washington, USA
[3]Department of Sports Medicine, Aspetar Orthopaedic and Sports Medicine Hosptial, Doha, Qatar
[4]Cardiovascular Sciences Research Center, St George's University of London, London, UK

Correspondence to
Dr Kimberly G Harmon, Departments of Family Medicine and Orthopaedics and Sports Medicine, University of Washington, 3800 Montlake Boulevard, Seattle, WA 98195, USA; kharmon@uw.edu

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ABSTRACT
Sudden cardiac death (SCD) is the leading medical cause of death in athletes; however, the precise incidence is unknown. The objectives of this review were to examine studies on the rate of SCD in athletes, assess the methodological strengths and weaknesses used to arrive at estimates, compare studies in athletes with estimates in similar populations and arrive at an approximation of the incidence of SCD based on the best available evidence. A comprehensive literature search was performed in PubMed using key terms related to SCD in athletes. Articles were reviewed for relevance and included if they contained information on the incidence of SCD in athletes or young persons up to the age of 40. The reference list from each manuscript was reviewed for additional relevant articles. The methods for case identification were examined, as well as the inclusion and exclusion criteria and the precision of the population denominator studied. Thirteen studies were found investigating the rate of SCD in athletes who ranged in age from 9 to 40. An additional 15 incidence studies were located examining the rate of SCD in other populations under the age of 40. Rates of SCD varied from 1:917 000 to 1:3000. Studies with higher methodological quality consistently yielded incidence rates in the range of 1:40 000 to 1:80 000. Some athlete subgroups, specifically men, African-American/black athletes and basketball players, appear to be at higher risk. The incidence of SCD in athletes is likely higher than traditional estimates which may impact the development of more effective prevention strategies.

INTRODUCTION
There is widespread agreement that sudden cardiac death (SCD) is the leading medical cause of death in athletes. Current estimates of the incidence of SCD in athletes range from almost one in a million[1] to 1:23 000[2] athletes per year, while some subpopulations of athletes are reported at even higher risk with an incidence of 1 in 3000.[3] The most commonly acknowledged incidence of SCD in athletes is 1 in 200 000, although recent studies and a closer examination of methodologies have challenged this estimate as too low.[3–5] Contrasts in current estimates are largely due to differing methodology and heterogeneous population comparisons. In order to accurately estimate incidence, a precise numerator (cases identified) and denominator (population) are required. Several factors affect case identification including the definition of an athlete, methods of data acquisition, lack of mandatory reporting requirements in most settings, the inclusion or exclusion of cases based on time and location of the event, inclusion of all cardiac events (including survivors) versus only those resulting in death and the population examined. Selection and calculation of the denominator also varies widely in precision. An accurate understanding of the incidence of cardiovascular events in athletes is absolutely critical to the development and evaluation of screening guidelines. This review will critically examine and compare incidence studies in order to better understand the incidence of SCD in athletes.

METHODS
A comprehensive literature search was performed in PubMed using the terms: incidence, sudden cardiac death, sudden death, sudden cardiac arrest, etiology, pathology, registry, athlete, young, children and adolescents. Articles were reviewed for relevance and included if they contained information on the incidence of SCD in athletes or young persons up to the age of 40. The reference list from each manuscript was reviewed for additional relevant articles. SCD was defined as a sudden unexpected death due to cardiac causes, or sudden death in a structurally normal heart that had no other explanation and a history consistent with cardiac-related death, that occurred within 1 h of symptom onset in a person without known cardiac disease or an unwitnessed death occurring within 24 h of the person having been alive and symptom free.

UNDERSTANDING THE METHODOLOGY
The key to accurately interpreting any study on incidence is to understand the methods used to identify cases, the criteria for case inclusion or exclusion, and what population is being studied. Incidence and aetiology vary considerably with age, making conclusions within broad age ranges difficult to draw. Investigations of well-defined populations such as high school or college athletes may have more important application. Some reports include only exertional deaths, only those which occur within an hour of exercise, or only while playing for a school-sponsored team and reported to insurance companies. This will substantially underestimate the overall incidence of SCD.[3 4 6–9]

Exertional SCD or SCD occurring at any time?
The primary objective of prevention programmes is early detection of disease, in order to prevent adverse health and catastrophic events from ever occurring. While cardiovascular screening in athletes is generally thought justified due to an increased risk of cardiovascular events during exertion, programmes designed to prevent SCD are not intended to prevent SCD only during sport or only during a school-sponsored activity, but rather to promote athlete health and prevent SCD at any

time. Many of the pathologies associated with SCD in young athletes may present with lethal arrhythmias at rest or during sleep; therefore, it is the incidence of SCD inclusive of SCD occurring outside of sport, not just the incidence of SCD occurring during exertion, which is necessary to accurately identify athlete risk.

Case identification: mandatory reporting, media reports, registries and insurance claims

Mandatory reporting systems afford the most reliable results, although few of these exist. Registries depend on passive collection methods and are subject to ascertainment and selection bias. Many studies rely on the use of media reports to identify cases; however, this results in underestimating SCD incidence. In one retrospective cohort study from Denmark, only 20% of athlete deaths identified by death certificates were found through an extensive media search.[6] In another retrospective cohort study of a high profile US college athlete population, media reports identified only 56% of total cases.[3] Media reports are likely to be less reliable prior to widespread use of the Internet and in lower profile athletes (younger athletes, female athletes and non-revenue sports). Studies using catastrophic insurance claims as the only source of case identification also are likely to underestimate the incidence of SCD. Insurance claims provide information only on deaths which occur during school-sponsored sporting events, and do not include cases that occur during individual activity, unofficial practices or deaths which occur outside of sport. In fact, catastrophic insurance claims missed 80% of SCD cases in a study using an internal reporting structure for the NCAA. Consequently, incidence assessments based on media reports and catastrophic insurance claims should be considered a minimum estimate.

The importance of the denominator

Incidence calculations are based on a ratio, thus if the population being examined (the denominator) is not well defined the final estimate can be highly variable. The most accurate incidence estimates will be in a distinct group with well-defined demographics. Estimating the overall number of athletes based on population statistics, in large age ranges or multiple demographic areas without an overarching authority is fraught with potential error. Additionally, some studies may examine sudden deaths in a population as opposed to all deaths in a population.[7 8] When comparing the relative risk of death, eliminating subtypes such as accidental deaths from the overall analysis significantly changes the ratios and can skew results making entities appear more or less common. Consequently, an accurate denominator is critical to precise incidence estimates.

Impact of age on SCD incidence

The rate of SCD varies considerably with age.[10–13] In young athletes (<30 years old), the aetiology of SCD usually involves a genetic structural or primary electrical cardiac disorder. In older athletes, the rate of atherosclerotic heart disease increases rapidly, particularly after the age of 25–30. Thus, studies inclusive of large age ranges or a substantial proportion of older participants can vary significantly and may disproportionately affect incidence rates.[14 15]

Should SCA be included in incidence calculations?

Finally, some incidence calculations examine only SCD, while others also include sudden cardiac arrest (SCA) victims that survive. SCA is an important endpoint with potential morbidity, but it is more difficult to track using the most commonly employed methods. SCA is often not reported in the media and in most settings there is no requirement to report SCA. In early reports of SCD before public access defibrillation programmes, SCA events likely resulted in death, so the need to recognise SCA survivors and SCD victims in incidence calculations was less apparent. With the implementation of emergency action plans and the widespread use of automated external defibrillators (AEDs), survival rates from SCA in athletes now rival that of other public access defibrillation programmes, with one recent prospective observational study demonstrating an 89% survival rate.[9] Survival from SCA may drastically decrease the overall incidence numbers of SCD. Primary prevention programmes are intended to identify athletes at risk and prevent SCA from ever occurring, so to properly evaluate screening programmes the inclusion of SCA and SCD in the assessment of incidence is warranted.

In fact, the identification of potentially lethal cardiac abnormalities during preparticipation screening appears to be universally supported. The American College of Cardiology (ACC), the American Heart Association (AHA), the European Society of Cardiology and the IOC all endorse that the identification of potentially serious medical conditions including cardiovascular disease that place athletes at risk for safe sports participation is the primary reason for screening.[16–19] The latest edition of the Pre-Participation Physical Evaluation Monograph endorsed by six US medical societies and the AHA also states the primary objective of the preparticipation examination is to 'screen for conditions that may be life threatening or disabling'.[20] With such widespread agreement that the objective of screening is the identification of potentially life-threatening conditions, the effectiveness of preparticipation screening should be understood not just in terms of SCD but also in terms of SCA with survival.

EVALUATING THE STUDIES

Estimates of SCD in general populations of athletes

There are 13 studies reporting on SCD in athletes with 5 reporting on SCD incidence in general populations of athletes, most with broad age ranges from 8 to 40[6 8 21–23] (table 1). The first study to report on the rate of SCD in athletes was a 1995 study which employed a retrospective cohort design and utilised the National Center for Catastrophic Sports Injury Research database from 1983 to 1993 to evaluate sudden deaths in high school and college athletes. An overall SCD rate of 1:300 000 was estimated with a rate of 1:1 300 000 in women.[21] While this effort highlighted an area without much previous research, this study relied on passive collection methods and media reports in the preinternet era for case identification, and undoubtedly cases were missed. In addition, the denominator was imprecise. However, 1:300 000 became the starting point for the incidence of SCD in athletes.

The largest report of SCD in a general population of athletes comes from the US Registry for Sudden Death in Athletes (USRSDA).[8 24] The USRSDA has collected information on sudden death in athletes since 1980. The primary sources of information are media searches, electronic databases and self-reports, that is, next-of-kin. The number of cases identified has increased steadily by approximately 6% annually as media search strategies have improved.[8] SCA and SCD were included which contrasted the previous study. The denominator of the athlete population was set at 10.7 million although there is no descriptor of how this was arrived at; in fact, in two previous reports using the same database incidence rates were not reported due to 'unavoidable selection bias' and the certainty of significantly underestimating the rate.[25 26] In this study, the

Table 1 Incidence studies in general populations of atheltes

Author	Year	Country	Study design	Case identification	Denominator	Exertional deaths of all?	SCD or SCA+SCD	Years studied	Population	Incidence	Number of years	Age range	Mean age	Number of cardiac deaths
Van Camp[21]	1996	USA	Retrospective cohort	National Center for Catastrophic Injury Research and media database	17 most popular sports, participants in NCAA, NFHS, NAIA, NAJC, added together, conversion factor to account multisport athletes used 'based on discussions with representatives from the national organisations'. 1.9 for high school and 1.2 for college	Exertional	SCD	1983–1993	College athletes and high school	1:300 000	10	17–24	17	100
Maron[26]	1996	USA	Retrospective cohort	US Registry for Sudden Death in Athletes	'Unavoidable selection bias and certainly significantly underestimate'	All	SCD	1985–1995	Athletes	_	10	12–40	17	134
Maron[25]	2003	USA	Retrospective cohort	US Registry for Sudden Death in Athletes	Not possible b/c of selection bias	All	SCD	1985–2000	Athletes	_	25	9–40	17	286
Corrado[22]	2003	Italy	Prospective cohort study	Mandatory death reporting	Registered Italian athletes	All	SCD	1979–1999	Athletes and young people	1:47 600 athlete 1:142 900 young people	20	12 –35	23	55
Maron[8]	2009	USA	Retrospective cohort	US Registry for Sudden Death in Athletes	Estimated 10.7 million athletes <39 participating in sports during 2000–2006 (method not described)	All	SCA+SCD	1980–2006	Athletes	1:163 934	27	8–39	18	690
Holst[6]	2010	Denmark	Retrospective cohort	Review of death certificates—then autopsies if available —15 sports related SCD (SrSCD)	Denmark population statistics	All and sports related	SCD	2000–2006	Athletes and young people	1:82 645 SrSCD 1:26 595 general pop	7	12 –35	26	15 SrSCD 470 SCD
Steinvil[23]	2011	Israel	Retrospective cohort	Retrospective review of two Israeli newspapers by two media researchers	45 000 registered competitive athletes in 2009, extrapolated the growth of the Israeli population age10–40 since 1985 based on that figure and allowed for a presumed doubling of the sporting population	All	SCD	1985—19 971 998—2009	Athletes	1st—1:393 702nd—1:37 593	24	12–44	24	24

SCA, sudden cardiac arrest; SCD, sudden cardiac death.

incidence of SCD for US athletes aged 8–39 from 2001 to 2006 was calculated at 1:164 000.[8] The strengths of this retrospective cohort study are the long time frame of the registry and the number of cases identified. The limitations of the incidence calculation include a broad age range, underestimation of events and an approximated denominator.

Estimates of SCD in athletes in other parts of the world are impacted by the use of different methods for case ascertainment. In the Veneto region of Italy, there is a mandatory reporting system for juvenile sudden death with autopsies performed by specialist cardiovascular pathologists. In a prospective cohort study from 1979 to 1999, the reported rate of SCD was 1:47 000 in athletes aged 12–35.[22] This study presented a more accurate case identification and a precise denominator and the rate was higher than in previous studies. In a subsequent report from the same Italian dataset, the initial rate of SCD from 1979 to 1981 was 1:28 000, and after implementation of a national screening programme, the rate of SCD decreased by 89% to 1:250 000 in 2006 suggesting benefit to a cardiac screening programme inclusive of ECG.[27]

The disparity in incidence numbers between Italy and the USA have been attributed to the differing ethnic populations with differing risks for SCD and has led some to suggest that the USA need not consider more intensive screening strategies.[28] It is more likely that the differing SCD incidence is due to the disparate methods of measuring incidence: retrospective cohorts based on media reports, catastrophic insurance claims and registries subject to ascertainment bias with uncertain denominators, versus a prospective mandatory reporting system with a reliable denominator.

Other studies of SCD incidence using a broad age range of competitive athletes were performed in Israel and Denmark.[6 23] The Israeli study was a retrospective cohort study which relied on the retrospective search of two major newspapers for case identification in athletes aged 12–44. The denominator was developed by ascertaining the number of registered athletes in 2009 and then extrapolating retrospectively, based on population statistics and the predicted growth in the percentage of the population engaged in athletic activity over time, the number of competitive athletes from 1985 onwards. The incidence of SCD was estimated to be 1:39 000 from 1985 to 1997 and 1:38 000 from 1998 to 2009.[23] Critical weaknesses of this study include case identification using only a retrospective search of two newspapers, inclusion of a 24-year interval with variable media attention to athlete SCD, the broad age range with rates of SCD largely influenced by inclusion of older athletes and the imprecise denominator. The authors concluded that an ECG screening programme started in 1998 showed no benefit in the prevention of SCD in athletes, basing this claim on a flawed scientific foundation.[4] The retrospective cohort study from Denmark reviewed death certificate and autopsy information for case identification and used population statistics as a denominator.[6] The investigators reported a rate of SCD of 1:83 000 in athletes aged 12–35. The primary weakness in this study was the ability to accurately identify competitive athletes from the available information.

SCD in US college athletes

As the incidence of SCD varies considerably with age, the ability to examine populations with more narrowly defined age ranges is important. All studies on SCD incidence in competitive athletes outside the USA include broad age ranges. In the USA, three retrospective cohort studies specifically investigate the incidence of SCD in college athletes aged 17–24, with rates ranging from 1:43 000 to 1:67 000[3 7 29] (table 2). In the first, a survey

Table 2 Incidence studies in college athletes

Author	Year	Country	Study design	Case identification	Denominator	Exertional deaths of all?	SCD or SCA +SCD	Years studied	Population	Incidence	Number of years	Age range	Mean age	Number of cardiac deaths
Drezner[29]	2005	USA	Retrospective survey	Survey answered by 244/326 (75%) Div I NCAA institutions	Number of athletes at surveyed schools	All	SCD		College athletes	1:67 000	3.3			5
Harmon[3]	2011	USA	Retrospective cohort	Parent Heart Watch database, NCAA Resolutions list, insurance claims—capture–recapture analysis demonstrated 90–100% of deaths were likely identified	Participation data from NCAA	All	SCD	2004–2008	College athletes	1:43 000	5	18–26	20	37
Maron[7]	2014	USA	Retrospective cohort	US Registry for Sudden Death in Athletes and NCAA resolutions list for cardiac cases	Participation data from NCAA	All	SCD	2002–2011	College athletes	1:83 000—confirmed 1:62 000—presumed	10	17–26	20	64

SCA, sudden cardiac arrest; SCD, sudden cardiac death.

Table 3 Incidence studies in high school students

Author	Year published	Country	Study design	Case identification	Denominator	Exertional deaths of all?	SCD or SCA +SCD	Years studied	Population	Incidence	Number of years	Age range	Mean age	Number of cardiac deaths
Maron[31]	1998	USA	Retrospective cohort	Insurance claims	Minnesota State High School League	Exertional during school sponsored sport	SCD	1985–1997	High school athlete	1:217 000 overall 1:129 000 male 0 for female	12	16–17	16.5	3
Drezner[2]	2009	USA	Cross-sectional survey	1710 high schools with AEDs surveyed for SCA or SCD	Number of student athletes reported by schools	SCA or SCD occurring on campus	SCA +SCD	2006–2007	High school athletes	1:23 000 SCA +SCD 1:46 000 SCD	Within 6 months of survey	14–17	16	14
Maron[32]	2013	USA	Retrospective cohort	US Registry of Sudden Death in Athletes	Minnesota State High School League	Exertional	SCD	1986–2011	High school athletes	1:150 000	26	12–18	16	13
Roberts[1]	2013	USA	Retrospective cohort	Insurance claims data	Minnesota State High School League	Exertional during school sponsored sport	SCD	1993–2012	High school athletes	1:416 666 over last decade 1:917 000	19	12–19	_	4
Toresdahl[34]	2014	USA	Prospective observational	2149 schools followed for SCA+SCD which occurred on school campus	Number of student athletes reported by schools	SCA or SCD occurring on campus	SCA +SCD	2009–2011	High school athletes	1:87 719 SCA +SCD 1:57 000 male SCA+SCD	2	14–18	_	18 SCA+SCD 2 SCD
Drezner[5]	2014	USA	Retrospective cohort	Media reports	NFHS	All	SCA +SCD	2003–2013	High school athletes	1:153 846 SCD 1:71 428 SCA 1:21 277 male basketball	10	14–18	–	6 SCD 7 SCD
Harmon[35]	2014	USA	Retrospective cohort	Media reports	NFHS	All	SCA +SCD	2007–2013	High school athletes	1:63 988 SCA 1:41 662 male 1:33 815 male basketball	5	14–18	_	74 SCD 35 SCA

SCA, sudden cardiac arrest; SCD, sudden cardiac death.

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was sent to NCAA institutions regarding SCD and AED use over an average of 3.3 years and an SCD rate of 1:67 000 was calculated based on 244 responding institutions.[29] In the second, a database of NCAA college athlete deaths was compiled from the Parent Heart Watch media database, the NCAA Resolutions database and NCAA insurance claims. This database looked retrospectively at 5 years of deaths from 2004 to 2008, identifying approximately nine cases of SCD per year,[3] with the cardiovascular cause of these deaths confirmed by autopsy review in a subsequent study.[30] NCAA participation statistics for race, sex and sport were used for the denominator. In this study an overall SCD rate of 1:43 000 was calculated; additionally, high-risk subgroups were identified such as men with a risk of 1:33 000, African-Americans with a risk of 1:18 000, basketball athletes with a risk of 1:11 000 and Division I male basketball players with an alarming risk of 1:3000.[3] A similar study reviewed sudden deaths in NCAA athletes over a 10-year period from 2002 to 2011.[7] In this study the USRSDA and NCAA Resolutions database was used for case identification, reporting approximately six cases of SCD per year with an incidence of 1:62 000.[7] Thus, these three different studies using variable methodology all arrive at reasonably comparable figures for an incidence of SCD in college athletes of approximately 1:50 000, with some subgroups at higher risk.

SCD in US high school athletes

High school athletes aged 14–18 provide another well-defined age subgroup to determine incidence. Seven US studies examined high schools athletes with wide variation in the reported rate of SCD from 1:23 000 to 1:917 000[1 2] (table 3). An initial incidence estimate of 1:217 000 in high school athletes came from Minnesota in a retrospective cohort study with case identification reliant on catastrophic insurance claim data.[31] A recent follow-up study also limited to catastrophic insurance claims reported an incidence of 1:917 000 over the past decade.[1] Catastrophic insurance claims only capture deaths which occur during school-sponsored events or practices and significantly underestimate the total incidence of SCD. Highlighting this point was a retrospective review of the very same population of Minnesota high school athletes but using media reports for case identification. Media reports identified 13 cases in Minnesota high school athletes (all in boys), including 6 cases of SCD and 7 additional cases of SCA in student athletes who survived. Catastrophic insurance claims detected only one of six deaths (17%) that occurred in athletes during the same study period and only one of four deaths which occurred while playing sports.[5] The three exertional deaths missed by catastrophic insurance claims occurred in athletes when participating in club sports or unofficial practices. In a recent retrospective cohort study of Minnesota state high school athletes using the USRSDA, 13 cardiovascular related sudden deaths were identified over a 26-year period with an estimated incidence of 1:150 000.[32] On closer examination and comparison of studies, catastrophic insurance claims identified only four deaths over 19 years and during an overlapping time period the USRSDA identified 13 deaths over 26 years, again emphasising the limitations of using catastrophic insurance claims as the sole means for case identification. Furthermore, insurance claims data limited only to deaths and not SCA will vastly underestimate the rate of major cardiovascular events in athletes.

Other studies have found higher rates of SCD in high school athletes. In a cross-sectional survey of 1710 high schools, an SCD rate of 1:46 000 was found, with a rate of 1:23 000 when SCA was included.[2] While this study is limited by the potential for reporting bias, these numbers may underestimate risk as only events which occurred on high school campus were captured; events which occurred away from campus or while sleeping were not included. In two retrospective population-based cohort studies, up to one-third of SCD occurred while sleeping, illustrating the limits of only including school-based events.[6 33] A recent 2-year prospective cross-sectional study of 2149 high schools demonstrated an overall SCA rate of 1:88 000.[34] While the prospective monitoring provided more rigorous surveillance, this study also examined events only occurring on school campus. Lastly, a preliminary study examined SCA and SCD in high school athletes in six US states, representing 36% of the US high school population and almost 20 million athlete seasons over 5 years from 2007 to 2013.[35] Media reports were used for case identification and the denominator was defined using data from the National Federation of State High School Associations (NFHS), with an SCA/SCD incidence rate of 1:63 000 calculated.[35] The incidence in male high school athletes was 1:41 000 and in male basketball athletes was 1:33 000.[35]

In conclusion, studies of incidence in high school athletes are more difficult to interpret with a wide range of variables and methods accounting for different incidence rates. The methodological limitations of incidence studies in high school athletes must be recognised. Current assessments of incidence either focus on a fraction of the day (school hours),[1 2 31 32 34] only account for SCD occurring with certain activities,[1 31 32] include only SCD and not SCA with survival[1 31 32 34] or rely on media reports for case identification.[5 36] All these issues result in incomplete case identification and an underestimation of the incidence of SCD. Based on available studies, we believe that 1:50 000 to 1:80 000 should be considered a minimum estimate range for the incidence of SCD in high school athletes with male athletes at higher risk.

Comparisons to similar non-athlete populations

Fifteen studies report on the incidence of SCD in young, non-athlete populations, primarily generated by retrospective cohort studies using death certificates, coroner's databases and autopsies for case identification. SCD estimates range from 1:22 000 to 1:128 000, depending on the age of the population examined.[6 10 14 33 36–40] There are several prospective population-based studies using emergency medical services databases which include SCA and SCD and provide more rigorous methodology to consider[13 41–43] (table 4). The most comprehensive of these prospective studies spanned 30 years and reported an SCA/SCD rate of 1:69 000 in 14–24 year olds and 1:22 000 in 25–35-year olds in King County, Washington.[43]

Four studies have compared the relative risk of SCD or SCA in competitive athletes with the general population. It is generally accepted that intense physical exertion increases the likelihood of SCA in those with underlying cardiovascular disorders. An Italian prospective cohort study reported a 2.5 times relative risk for SCD in competitive athletes compared with age-matched non-athletes.[22] In a prospective cohort study in France, a 4.5 times relative risk of sports-related SCD was reported in competitive athletes compared with recreational sports participants.[32] Similarly, a prospective observational study in US high schools with over 4.1 million total student-years and over 1.5 million student athlete-years of surveillance found a 3.6 times relative risk for SCA on campus in high school student athletes versus student non-athletes.[13 34] In contrast, in a retrospective review of death certificates and media reports in Denmark, investigators reported a rate of SCD in athletes aged 12–35 that was 3.3 times lower than the general population.[6]

Table 4 Incidence studies in the military and general population

Author	Year	Country	Study design	Case identification	Denominator	Exertional deaths of all?	SCD or SCA +SCD	Years studied	Population	Incidence	No. of years	Age range	Mean age	No. of cardiac deaths
Eckart[44]	2004	USA	Retrospective cohort	Mandatory reporting of all deaths to DOD recruit mortality registry with autopsy data	Numbers from DOD	All	SCD	1977–2001	Military recruits	1:10 000	25	18–35	19	108
Gunnarson[36]	2006	Iceland	Retrospective cohort	Death certificates and autopsy reports reviewed	Iceland population statistics	All	SCD	1974–2004	Young people	1:68 000	30	12–35		45
Atkins[41]	2009	USA and Canada	Prospective cohort study	EMS responding to confirmed cardiac cases	Population statistics	All	SCA +SCD	2005–2007	Young people	1:49 000	3	12–19	16.4	193
Chugh[42]	2009	USA	Prospective population based	EMS responding to confirmed cardiac cases	Population statistics	All	SCA +SCD	2002–2005	Young people	1:58 828	3	10–14	_	2
Papdakis[37]	2009	UK	Retrospective cohort	Review of death certificates	Population statistics	All		2002–2005	Young people	1:55 000	4	1–34	_	3409
Holst[6]	2010	Denmark	Retrospect cohort	Death certificates and autopsy reports reviewed if available (67%)	Population statistics	All	SCD	2000–2006	Young people	1:35 000	7	1–35	26.4	15
Solberg[14]	2010	Norway	Retrospective cohort	Norwegian cause of death registry—validated with autopsy and medical records	Population statistics	Exertional	SCD	1979–1998	Young people	1:111 111	20	15–34	27	23
Cooper[40]	2011	USA	Retrospective cohort	Automated data from 4 health plans—study carried out to compare ADHD users to non-users	Number of those 2–24 in health plan	All	SCA +SCD	1986–2005	Young people	1:32 258	19	2–24	11.1	81
Eckart[12]	2011	USA	Retrospective cohort	Mandatory reporting of all deaths to DOD mortality registry with autopsy data	DOD statistics	All	SCD	1998–2008	Military	<20–1:2 967 320–24 1:4 098 325–29– 1:3 012 030–35–1:25 000	10	18–34	_	298
Margey[39]	2011	Ireland	Retrospective cohort	Death certificate review with autopsy confirmation	Population statistics	All	SCD	2005–2007	Young people	1:350 001:20 000 males 1:77 000 female	3	15–35	25	116
Marjion[13]	2011	France	Prospective	EMS responding to confirmed cardiac cases	Population statistics	Sports related	SCA +SCD	2005–2010	Young people	1:102 000 competitive athletes (10–35)	5	12–75	46	820
Meyer[43]	2012	USA	Prospective population based	EMS responding to confirmed cardiac cases	Population statistics	All	SCA +SCD	1980–2009	Young people	14–24 1:69.00 025 –35 1:22 700	30	14–24 25–35	_	14–24: 60 25–35: 245
Pilmer[10]	2013	Ontario, Canada	Retrospective cohort	From coroner's database	Population statistics	All	SCD	2008	Young people	1:34 000 (19–29)	1	2–40	_	174
Pilmer[38]	2014	Ontario, Canada	Retrospective cohort	From coroner's database	Population statistics	All	SCD	2005–2009	Young people	1:128 000	5	1–19	12	116
Winkle[33]	2014	Denmark	Retrospective cohort	Death certificates, autopsy reports and health records reviewed	Population statistics	All	SCD	2000–2006	Young people	1:90 909	7	1–18	13	62

SCA, sudden cardiac arrest; SCD, sudden cardiac death.

The US military is perhaps the best source to calculate incidence rates of SCD in active individuals, due to the mandatory reporting of deaths with standard autopsy protocols and access to premortem medical records and clearly defined denominators. Although individuals in the military do not meet standard definitions of a competitive athlete, military personnel undergo regular, intense physical training, especially for recruits during basic training. Given the rich source of quality data and the similar median age to college athletes (19 years), these studies in military recruits are a good comparator. In a retrospective cohort study of 6.3 million recruit years over 25 years, the rate of SCD in military recruits was 1:10 000.[44] In a follow-up study examining the rate of SCD in active duty military personnel who undergo regular physical activity but not necessarily the intense activity experienced during basic training by recruits, the rate of SCD was approximately 1:30 000 in persons under the age of 30, with the expected increase in overall incidence and death secondary to atherosclerotic heart disease in older military personnel.[12] These military studies in exercising young adults with better methodology also support a higher SCD incidence in athletes than initial estimates.

LOOKING AHEAD

Better quality studies with mandatory reporting of SCD and SCA are required to acquire more accurate information in athletes. Ideally, when SCD does occur there would be a standardised autopsy from a cardiac pathologist accompanied by a molecular autopsy. A pilot study of such a system in five US states with required reporting of sudden death in youth followed by death investigation is currently being funded by the National Heart Lung and Blood Institute, with data collection to begin in 2015. Likewise, in January 2014 the NCAA passed legislation requiring all NCAA institutions to report cases of catastrophic injury (including SCA) or death beginning August 2014. Both these initiatives will provide a rich source of prospective data in a defined population to give more accurate information regarding the incidence and aetiology of SCA/SCD in athletes.

CONCLUSIONS

It is widely acknowledged that SCD is the leading medical cause of death in athletes although its exact incidence remains unclear. Use of media reports or catastrophic insurance claims as the primary method for case identification underestimates the risk of SCD in athletes. The best available evidence together with a close examination of reporting methods for case identification and population definitions, suggests that the incidence of SCD in athletes is higher than initial estimates. An overall incidence of 1:50 000 in young athletes is a reasonable estimate based on a synthesis of existing information, primarily retrospective cohort studies and prospective observational or cross-sectional studies, and when compared with similar populations such as the military where the quality of available data is greater. Male athletes are consistently found to be at greater risk, and there appears to be a disproportionately higher risk among male basketball athletes which requires further investigation. Variability in incidence rates is largely due to differences in methodology including the reliability of case identification, the accuracy of population denominators and the cases which are excluded or included. The goal of cardiovascular screening programmes is to identify potentially lethal cardiovascular disorders, and all SCA and SCD should be included in incidence calculations if a valid assessment of prevention strategies is to be considered. Accurate incidence numbers are important in understanding if screening programmes limited to athletes are warranted, cost effective and effectual. It must be acknowledged that unrecognised limitations and misinterpretations of current data have perpetuated an underestimation of the incidence of SCD and perhaps impeded progress towards the evaluation of more effective screening programmes.

Summary points

- Sudden cardiac death (SCD) is the leading medical cause of death in athletes.
- There are widely varying estimates of SCD due to differences in methodology including the reliability of case identification, age range of athletes studied, the accuracy of population denominators and the cases which are excluded or included.
- There is widespread agreement that athletes should undergo cardiovascular screening prior to participation in sport, and the primary objective of this screening is the identification of potentially life-threatening disorders.
- To fully understand the cardiovascular risks in athletes and the potential to prevent major cardiovascular events through screening, incidence estimates should include sudden cardiac arrest (SCA) survivors and SCD cases occurring at any time (sleep, rest, exertion).
- Based on a synthesis of best available information, an SCD incidence rate of 1:50 000 in college athletes and 1:50 000–1:80 000 in high school athletes should be considered a minimum.
- Some subpopulations such as males, black/African-Americans and basketball athletes appear to be at higher risk.

Research questions

- What is the incidence of SCA and SCD in high school and college athletes based on prospective studies with mandatory reporting requirements and defined populations?
- What is the aetiology of SCD in different athlete populations utilising expert cardiac forensic pathologists and molecular autopsy?
- Why are basketball athletes at higher risk of SCA and SCD compared with other athletes?
- Are athletes at greater risk of SCA and SCD than non-athletes using prospective methodology and mandatory reporting mechanisms?

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