

Data from a nationwide registry on sports-related sudden cardiac deaths in Germany

Philipp Bohm, Jürgen Scharhag and Tim Meyer

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Abstract

Background: Prospective national registries examining the incidence and aetiology of sports-related sudden cardiac death (SrSCD) not only in competitive athletes but also in recreational sports participants are uncommon. In May 2012, a prospective registry on SrSCD was installed to examine the incidence and particularly the aetiology of such events in the general population in Germany.

Methods: The registry consists of a web-based platform to record SrSCD cases. Media-monitoring and cooperation with 15 institutes of forensic medicine complemented the search. SrSCD was defined as death occurring during sports activity or up to 1 hour after its cessation, regardless of successful resuscitation. We included subjects at all levels of competition as well as recreational athletes.

Results: After 30 months of observation, 144 SrSCDs were recorded (mean age 46.8 ± 16.2 years). The overall incidence was 1.2–1.5/million/year, with 97% being male. Most of the cases occurred in the context of non-elite competitive or recreational sports. Football and running were the most common disciplines. In subjects ≤ 35 years, myocarditis prevailed, whereas in athletes ≥ 35 years, CAD predominated by far. Few cardiomyopathies were observed.

Conclusions: In Germany, the largest proportion of SrSCDs occurs in middle-aged men during recreational sports or non-elite competitive sports. The distribution of cardiac diseases responsible for SrSCD seems to vary among European countries. Our findings may indicate the need for a larger focus on myocarditis prevention in the young as well as widening the screening scope to younger athletes below the ‘elite’ level and to senior athletes.

Keywords

Athletes, cardiac screening, prevention, sudden death

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Introduction

The sudden death (SD) of an apparently healthy athlete is a tragic event which attracts an abundance of media attention, especially when elite athletes are involved. However, the incidence of sports-related sudden cardiac death (SrSCD) in competitive young athletes is considered very low, ranging from 0.7 to 3.0 per 100,000 athletes per year.^{1–5} On the contrary, in recreational sports participants >35 years, the incidence of SrSCD is higher and may even rise in the future, as more and older individuals take part in organised sports.^{6,7} Most nontraumatic deaths in sport are attributed to underlying cardiac disorders. In young athletes under 35 years of age, hereditary or congenital cardiac abnormalities such as hypertrophic cardiomyopathy (HCM), arrhythmogenic right ventricular

cardiomyopathy (ARVC) or coronary artery anomalies (CAA) are most common.^{2,3,8,9} In athletes older than 35 years, coronary artery disease (CAD) accounted for the vast majority of SrSCD.^{6,10,11}

Prospective national registries examining the incidence and aetiology of SrSCD not only in competitive athletes but also in recreational sports participants are rare.¹⁰ However, establishment of prospective national registries is warranted to investigate regional distribution patterns of cardiovascular causes of SD (under

Institute of Sports and Preventive Medicine, Saarland University, Germany

Corresponding author:

Philipp Bohm, Institute of Sports and Preventive Medicine, Saarland University, Campus, Building B8 2, 66123 Saarbrücken, Germany.
 Email: philipp.bohm@gmx.de

consideration of genetic and environmental factors) as well as their impact on screening programmes. Therefore, we established a prospective registry on SrSCD to examine the incidence and particularly the aetiology of SrSCD and survived sports-related cardiac arrest (SrCA) in the general population in Germany.

Methods

This prospective study started in May 2012 under the auspices of the German Society of Cardiology (Deutsche Gesellschaft für Kardiologie) and is funded by the German Heart Foundation (Deutsche Herzstiftung). The observation period for this analysis was 30 months (May 2012–October 2014). The study was approved by the ethical review committee and by the independent Data Protection Centre Saarland, Germany.

Definitions

SrSCD was defined as death occurring during sports activity or up to 1 hour after its cessation. Survived SrCA was defined as cardiac arrest that occurred during sport or within 1 hour after its cessation with survival of the subject after defibrillation and/or cardiopulmonary resuscitation (CPR). Sports participants who survived cardiac arrest after defibrillation and/or CPR were considered to have experienced SrSCD. SrSCDs related to trauma – apart from commotio cordis – were excluded.

We included subjects aged 10–79 years because physical activity is very difficult to assess in the very young, and those older than 79 years rarely participate in sport. We defined three different levels of sport:

1. Elite competitive athletes were defined as subjects younger than 40 years who participate in an organised team or individual sport that requires extensive and intensive systematic training and regular competition against others and that gives large relevance to athletic excellence and achievement.
2. Non-elite competitive athletes were defined as subjects of any age who participate in organised competitive events, either in a team or individually.
3. A recreational athlete was defined as an individual who participates in informal recreational sport without competition against others, on either a regular or an inconsistent basis.

Case detection

Three different methods of case detection were used. Firstly, we established a confidential web-based data

platform which records sudden cardiac death (SCD) cases and survived sudden cardiac arrests (SCAs) occurring during sports activity or up to 1 hour after its cessation (www.scd-deutschland.de). Reporting of cases was possible for observers such as relatives, other athletes, coaches, and journalists as well as for medical doctors via online access. Following the entry, contact was established with the reporting person to get more precise information on the circumstances of the incident.

Additionally, we used systematic web-based media monitoring via Press-Monitoring-Screening (PMG), the largest German-speaking press database, on a weekly basis. With a combination of keywords including heart-related terms or activities, cases could be filtered out that occurred in relation to sports activity.

Finally, we cooperated with 15 regional institutes of forensic medicine in Germany.

A definitive cause of death could be fixed only in cases in which an autopsy had been performed or in which the diagnosis could be obtained with clinical certainty (e.g. via coronary angiography). In Germany, an autopsy is not legally binding and is only carried out if the death is regarded as potentially suspicious by the physician certifying the death and/or by the Public Prosecution Service.

Statistical methods

We referred to the GEDA-study (German Health Update) by the Robert Koch Institute (RKI) to get precise information about the typical physical activity of the German population.¹² According to their nationwide surveys, 66% of the general population in Germany ≥ 18 years perform sport (22.3% < 2 h/week, 22.2% 2–4 h/week and 21.3% > 4 h/week). Of the adolescents (< 18 years), 77.5% perform at least 2.5 h sport per week. According to a second nationwide representative study of private households, conducted by the German Socio-Economic Panel (SOEP), 53% of the German population ≥ 17 years perform sport at least once in a month.¹³ The German Federal Statistical Office provided us with exact data on the total population within Germany including a subdivision into year-groups. We calculated the incidence by dividing the average number of SrSDs reported per year by the total number of persons who perform sport according to both, the survey of the RKI (66%) and the SOEP (53%), taking into account the specific age groups.

Results

After 2 years and 6 months of observation, a total of 144 cases were recorded of which 38 subjects survived. Forty-four cases were reported via online access and 101 cases were found via Press-Monitoring-Screening

(one double case-detection). No SrSCD was reported by the different regional institutes of forensic medicine. The overall incidence of SrSCD for sports participants ≥ 18 years was 1.2 per million per year (RKI data). The incidence for male sports participants ≥ 18 years was calculated as 1.6 per million per year, whereas female athletes had a significantly lower incidence with 0.06 per million per year. Adolescents under the age of 18 years had an incidence of SrSCD of 0.4 per million per year. When calculating with the SOEP data, the overall incidence of SrSCD for sports participants ≥ 17 years was 1.5 per million per year.

One-hundred and eight (75%) SrSCDs occurred in public sports facilities (such as a stadium, sport club, or organised running event) and 125 (87%) were witnessed. Prompt CPR was initiated in 118 subjects (82%). The initial cardiac rhythm was ventricular fibrillation (VF) or pulseless fast ventricular tachycardia in 48 cases and asystole in seven cases (with 89 cases remaining unclear). Ninety-six subjects (67%) were transferred to hospital. SrSCD usually occurred during sports activity (122 cases, 85%). Only 22 cases (15%) occurred up to 1 hour after sports cessation. The majority of cases occurred in non-elite competitive or recreational athletes ($n=142$, 99%). Only two cases occurred in young professional elite athletes. Most SrSCDs were observed during football or running, which represent the most popular sport disciplines in Germany (Figure 1).

The mean age of the SrSCD subjects was 46.8 ± 16.2 years (age distribution shown in Figure 2). Of these, 97% were men. In most cases in which it was possible to obtain the cause of death with clinical certainty or from forensic investigations ($n=66$), the cause of death was of cardiovascular origin ($n=64$, 97%). Myocardial infarction was the most frequent cause of SrSCD ($n=34$, mean age 48.2 ± 10.6 years). Nine subjects showed severe coronary stenosis at coronary angiography that most likely resulted in a demand ischemia (e.g. ischemia caused by an imbalance between oxygen supply and demand) as a trigger for SrCA. Myocarditis was diagnosed in 11 subjects with a mean age of 24.5 ± 10.4 years. Only five cardiomyopathies were detected (mean age 20.0 ± 11.4 years), of which two were categorised as HCM, one as ARVC, and one as DCM. In one case, the cardiomyopathy could not be further classified. CAAs were identified in three subjects (mean age 21.3 ± 5.5 years). Two cardiac ion channelopathies were found (one long-QT syndrome, most likely drug-induced, and one Brugada syndrome). In 23 cases, the cause of death could not be established by means of autopsy or invasive/non-invasive cardiac imaging. Of those, eight subjects had a known CAD (all with a past myocardial infarction) so an ischemic cardiovascular event can be assumed. Seven subjects had at least one cardiovascular risk factor, which means that the likelihood of a cardiovascular event was certainly increased. Eight subjects had chest pain/dyspnoea temporally

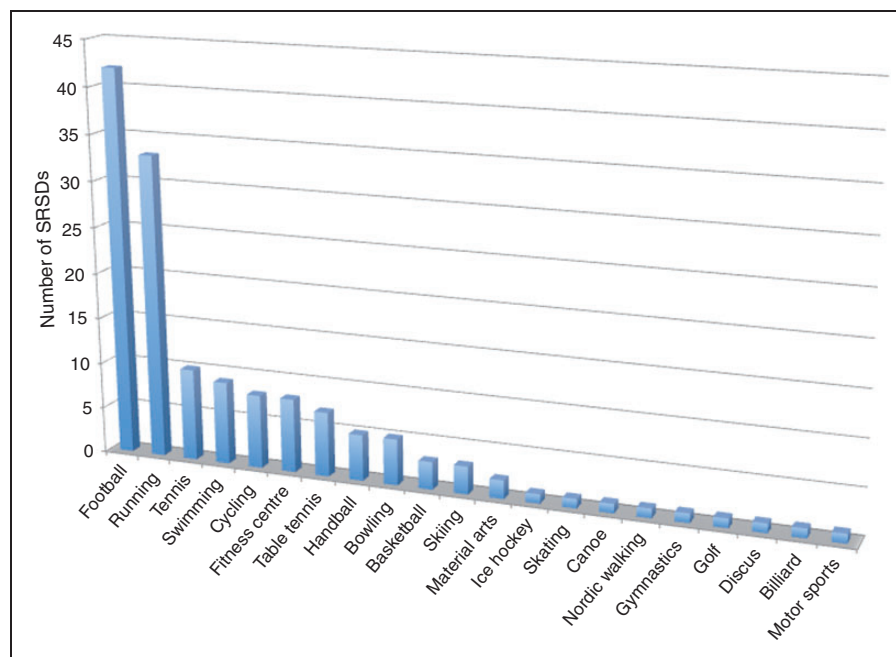


Figure 1. Distribution of sporting disciplines. Bar chart showing the distribution of the different sporting disciplines in which people were engaged at the time of sudden death. SRSDs: sports-related sudden deaths.

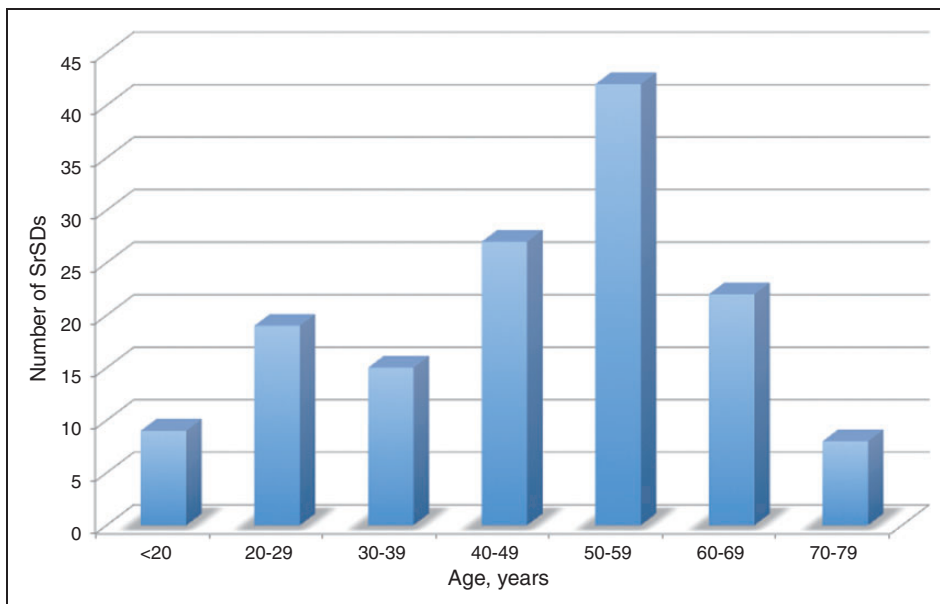


Figure 2. Distribution by age. Bar chart showing the distribution by age of sports-related sudden deaths (SrSDs) in the general population.

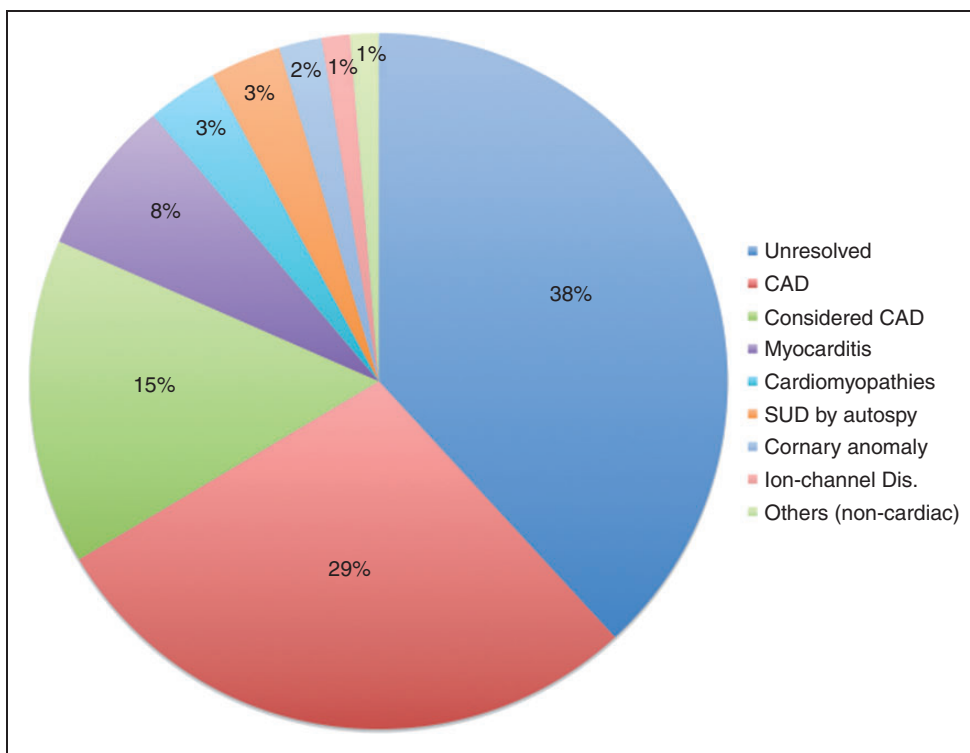


Figure 3. Causes of sports-related sudden deaths (SrSDs) in the general population. Pie chart showing the causes of the 144 SrSDs in the general population.

CAD: coronary artery disease; SUD: sudden unexplained death.

related to the SCA. Therefore, these 23 cases were placed in the category ‘considered CAD’ (Figure 3). The two non-cardiac causes were one pulmonary embolism and one subarachnoid haemorrhage.

Taking into account an age cut-off of 35 years, we observed 37 cases in subjects ≤ 35 years. Myocarditis prevailed in this age group, followed by premature CAD. All cardiomyopathies and CAAs occurred in

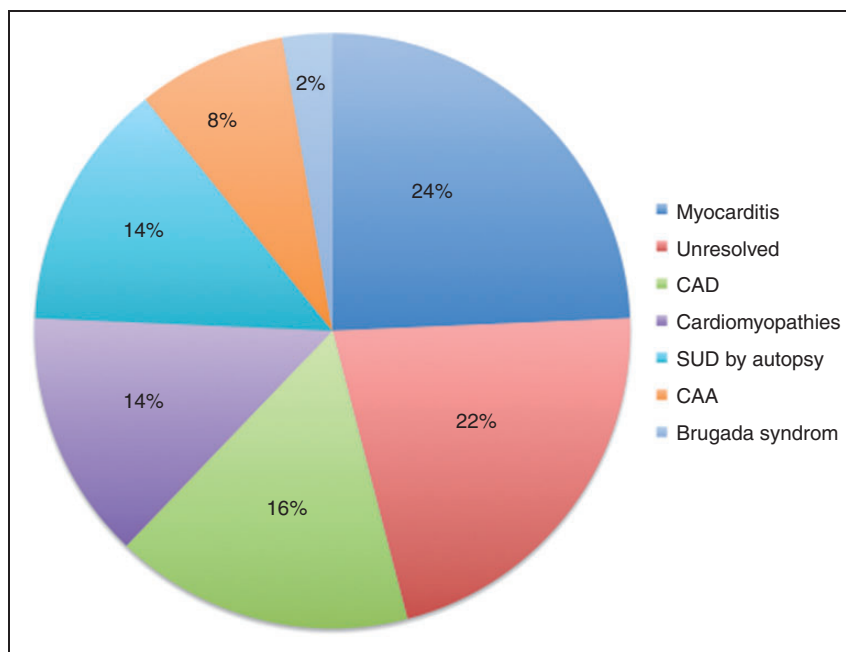


Figure 4. Causes of sports-related sudden deaths (SrSDs) in athletes ≤ 35 years. Pie chart showing the causes of the 37 SrSDs in athletes ≤ 35 years.

CAA: coronary artery anomaly; CAD: coronary artery disease; SUD: sudden unexplained death.

the group of athletes ≤ 35 years (Figure 4). In the age group > 35 years, CAD predominated by far. In total, 28 post-mortem examinations of the heart with detailed histopathological examination were carried out of which 18 had been performed in athletes ≤ 35 years. Five of them - all in the age group ≤ 35 years - remained without precise diagnosis. In those cases, post-mortem molecular analysis had not been performed.

Discussion

In our prospective SrSCD study, most cases occurred in middle-aged athletes performing non-elite competitive or recreational sport. This finding is not surprising as we face a growing number of mainly middle-aged or elderly individuals engaged in competitive sports activity.¹⁴ These data are in accordance with the study by Marijon et al. and Suárez-Mier et al. who also found SrSCD to occur mainly in the general population practising recreational sport.^{10,11} In our study, the most popular sport disciplines football and running were the most commonly affected. This finding does not necessarily mean that these sports represent the highest risks for SCA.⁴ In terms of cardiovascular demand during sport at the time of the event, most SCAs occurred when moderate to high dynamic/moderate static sporting disciplines were performed.

We noted a clear gender inequality with striking male predominance (by more than a factor of 20). Male gender itself constitutes an independent risk

factor for SCA and there is a known higher prevalence of CAD in young and middle-aged men.^{2,6,15} Furthermore, differences in the mode of physical activity and training between men and women might also be relevant, especially in the field of non-elite competitive sport. The fact that most male SrSCD cases during running occurred at the end of competitive long distance running events may indicate that predisposed runners exceeded their normal training pace and pushed themselves too hard above their usual limits, perhaps in combination with a perturbed hydration/electrolyte status.

Regarding the different cardiac disease entities, myocarditis and premature CAD prevailed in young athletes. The mean age in subjects with proven myocarditis was 24.5 ± 10.4 years. Winkel et al. found myocarditis to be the most common structural cause of cardiac death in those aged 1–18 years old.¹⁶ Thus, myocarditis may be more common in young athletes and might also be a triggering co-factor in fatal cases diagnosed with other structural cardiac diseases during autopsy. Interestingly, myocarditis was preceded by upper respiratory tract infections in almost all young myocarditis victims in our study. Therefore, special care should be taken upon return-to-play decisions and educational efforts should be made to spread information about risks from sport with an infection.¹⁷

Our surprising finding of premature CAD being a main cause for SCA in young adults is in line with a recent US study.¹⁸ In our study, even young subjects

without any cardiovascular risk factor were affected. This finding may give rise to questioning of the traditional age cut-off of 35 years separating younger from older athletes. Unsurprisingly, underlying CAD was by far the leading cause in older athletes which confirms the results by Marijon et al.¹⁰ Exercise-induced acute coronary syndromes (EIACS) normally result from atherosclerotic plaque disruption and coronary thrombosis. However, our clinical data suggest that demand ischemia (e.g. ischemia caused by an imbalance between oxygen supply and demand) in subjects with fixed coronary stenosis may also have been the trigger for malignant arrhythmias in EIA events, especially during long-distance running races.

Surprisingly, few cardiomyopathies were found, which confirms that the aetiology of SCD varies greatly in different parts of the world. In studies of US athletes, HCM accounts for the majority of fatal cases, whereas ARVC is responsible for up to one-quarter of SCDs in the Veneto region in Italy.^{9,19} In their retrospective study in the UK, De Noronha et al. found cardiomyopathies to be the most frequent cause of death accounting for 62% of all SCDs.²⁰ However, our results are in line with various other European studies which also showed low numbers of cardiomyopathies.^{10,21,22} These different results, even within Europe, certainly support the hypothesis that the distribution of the different cardiac diseases might be population-specific. Taking into account the ethnic diversity of the populations, the different regional distribution patterns of cardiac diseases may have a profound influence on the appropriate choice of screening procedures. Thus, to optimise targeted screening programmes, national registries are necessary.

In our study, 28 post-mortem examinations had been performed, of which 18 had been carried out in athletes ≤ 35 years. This relatively low number of autopsies can be explained as in Germany autopsies are not mandatory by law and are almost exclusively performed when the death is regarded as potentially suspicious. Therefore, a definite diagnosis in subjects without autopsy who died on site and were not transferred to hospital, could not be established. However, it was still possible to collect helpful indications from some of them. Sudden unexplained deaths (SUD) with a negative autopsy finding accounted for five cases (all in athletes ≤ 35 years) and may be a result of inherited arrhythmia syndromes such as ion channelopathies.²³ Post-mortem genetic testing was not performed in those cases, and should be considered in the future. In case of a SUD, the importance of a close family evaluation also must be highlighted.²⁴ In our study, family history regarding inherited cardiac diseases was unremarkable in all SUD cases.

Implications for preparticipation screening

Only two cases of SrSCD (myocarditis and myocardial infarction) have been observed in young elite competitive athletes so far, which clearly indicates that the absolute number of SrSCD cases in this category seems very low. However, our study revealed 36 athletes ≤ 35 years who experienced SrSCD without ever having benefited from a cardiovascular screening. Therefore, confining screening to 'elite athletes' may have to be reconsidered. On the other hand, one has to admit that some predominating disease entities in the young such as myocarditis, premature CAD, CAAs, and early HCM/ARVC are very difficult to detect, even with a broader cardiac screening programme. Therefore, it remains a difficult task for all organisations designing preventive cardiac screening programmes to adjust the combination of examinations to the current national epidemiological situation.

According to our data, the absolute risk for SrSCD appears to be higher in the general population. That most SrSCDs occurred in non-elite sports participants > 35 years who are usually not screened, indicates the need for a cardiac examination in those athletes. This examination is different from that of younger athletes as the risk of sports-related acute cardiovascular events is almost exclusively related to the development and progression of atherosclerotic CAD. Therefore, cardiovascular screening in athletes > 35 years should be performed according to the individual's cardiac risk profile and the intended level of physical activity.²⁵ Considering the extremely low number of female SrSCD cases in our study, the question arises of whether female subjects need to be screened to the same extent as men.

On-site prevention of SrSCD

Cardiovascular screening will never be able to detect all individuals at risk, regardless of the screening strategy applied. Thus, it is of utmost importance that in the case of a serious adverse event such as SCA, immediate CPR, defibrillation (if needed), and advanced cardiac life support are performed. In our study, 87% of the cases were witnessed and prompt bystander CPR had been initiated in 82% of the cases. However, early rhythm analysis is important as VF may be seen as initial rhythm requiring prompt defibrillation. In our study, 48 of 55 cases with definite information about the initial rhythm had VF, so it can be assumed that most such cases show VF as initial rhythm. Thus, access to early defibrillation through on-site automated external defibrillators (AEDs) and qualified persons is crucial to improve survival.²⁶

Study limitations

We have to acknowledge a certain degree of underreporting which is probably most obvious in the group of sports participants that exercise on an individual basis. Therefore, our estimated incidence of SrSCD in the general population is almost certainly an underestimation. Another limitation is the relatively low number of post-mortem examinations performed at the discretion of the certifying doctor and the Public Prosecution Service.

Conclusions

In Germany, by far the largest proportion of SrSCDs occurs in middle-aged men during recreational sports or competitive sports without elite character. This important finding will be of relevance for the public health in general, given the high participation rates of athletes >35 years in competitive events. The distribution of cardiac diseases responsible for SrSCD seems to vary among European countries. Our findings may indicate the need for a larger focus on myocarditis prevention in the young as well as widening the screening scope to younger athletes below the 'elite' level and to senior athletes. Regarding secondary prevention, educational efforts should be made to optimise safety measures at sporting facilities and to improve the chain of survival.

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