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Swissregard.ch – a prospective registry on sudden death and aborted sudden cardiac death in Swiss athletes

Abstract

Sudden death of an athlete on the field is the most devastating medical event in sports. Incidences of sudden cardiac death (SCD) of 0.5 to 3.0 per 100 000 athletes per year have been reported. Data are derived from reviews of public media reports, available electronic resources and observational studies, and consequently underestimation is likely. The most common causes of SCD in young athletes are underlying congenital cardiac disease or blunt chest trauma (Commotio Cordis) in a structurally normal heart. Male athletes are affected in up to 90% of cases. Competitive team sports like soccer or basketball or endurance sports like marathon running are associated with the highest risk of dying suddenly. Geographical differences in the causes of SCD have been reported and the role of ethnicity still has to be clarified. Therefore there is a need for national registries prospectively recording cases of sudden death in athletes of different regions and ethnicities. The data will be valuable for international comparisons and in the debate of tailored national pre-participation screening programs. In cooperation with regional Institutes of Forensic Medicine we designed a web-based database for recording cases of sudden death in athletes in Switzerland prospectively since 2011.

Zusammenfassung

Der plötzliche Todesfall eines Athleten im Wettkampf ist das verheerendste medizinische Ereignis im Sport. Inzidenzen plötzlicher Herztodesfälle werden mit 0.5 bis 3.0 Fälle pro 100 000 Athleten pro Jahr berichtet. Die Daten stammen aus öffentlichen Medienberichten, vorhandenen elektronischen Quellen und Beobachtungsstudien und eine Unterschätzung der wahren Fallzahl ist daher wahrscheinlich. Bei jungen Athleten sind angeborene Herzkrankungen die häufigste Ursache des plötzlichen Herztodes. Bei strukturell normalem Herzen kann ein Thoraxtrauma (Commotio Cordis) eine ventrikuläre Rhythmusstörung auslösen. In bis zu 90% der Fälle sind männliche Athleten betroffen. Kompetitive Teamsportler wie Fussballer oder Basketballer und Ausdauersportler wie Marathonläufer haben das höchste Risiko, plötzlich zu sterben. Die Ursachen plötzlicher Herztodesfälle zeigen regionale Unterschiede, und die Rolle der Ethnizität ist nicht vollständig geklärt. Aus diesen Gründen besteht ein Bedarf an nationalen Registern, die prospektiv plötzliche Todesfälle von Athleten aus unterschiedlichen Regionen und Kulturkreisen erfassen. Die so gewonnenen Daten sind nützlich für internationale Vergleiche und in der Debatte wie und für wen nationale Vorsorgeuntersuchungen gestaltet werden sollten. In Kooperation mit den regionalen Instituten für Rechtsmedizin haben wir eine webbasierte Datenbank erstellt, in der wir seit 2011 prospektiv plötzliche Todesfälle im Sport in der Schweiz erfassen.

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Background

Sudden death of an athlete on the field is the most devastating medical event in sports. Public interest in sudden cardiac death (SCD) arises from both the importance of sports in many societies and the paradox that physical activity can have both a positive and a negative impact on an individual's health [17].

Vigorous exertion transiently increases the risk of SCD [1]. Inherited cardiovascular conditions are responsible for SCD in young athletes, whereas in older athletes (>35 years of age) atherosclerotic coronary artery disease is the primary pathological finding [17]. Blunt chest trauma may cause ventricular arrhythmias

in a structurally normal heart [14]. The true incidence of sudden death in athletes remains unknown. Estimates from the United States (US) suggest a figure between 0.5 to 0.6 deaths per 100,000 athletes per year [14, 15]. A prospective population-based study in the Veneto Region of Italy reported an incidence of sudden death of 2.3 per 100 000 athletes per year from all causes, and of 2.1 per 100 000 athletes per year from cardiovascular diseases [6]. The discrepancy between the Italian and US figures may be explained in part by the different mechanisms used to identify cases of SCD in young athletes [3]. In US studies, case identification has been predominantly through review of public media reports and other available electronic resources, making underestimation of the true

incidence of SCD likely. A recent US study analyzed the National Collegiate Athletic Association Athletes database. The authors reported an incidence of SCD among collegiate athletes of 2.3 per 100 000 athletes. Only 56% of cases were identified by public media reports [11]. The Italian data were collected by a prospective registry of juvenile sudden death and showed similar results but in a relatively limited geographic area [3, 5].

There are several issues with the Italian study. The most salient points are that these results are from an observational study, including only white athletes from a relatively small area in the Veneto region of Italy [18].

Therefore there is a need for national registries prospectively recording cases of sudden death in athletes of different regions and ethnicities, making international comparisons possible. Switzerland is situated in the middle of Europe, spanning an area of 41 285 km². There are 7.8 million people living in Switzerland, nearly twice the population of the Veneto region. More than 75% of the young population is participating in leisure time activities on a regular basis, and 20% take part in competitions [9].

We formulated the following hypothesis: The cause of sudden death in athletes is of cardiovascular origin in more than 50% of cases. More than 75% of cases will occur in male athletes. Competitive team sports like soccer or basketball or endurance sports like marathon running place the athletes at highest risk of dying suddenly. A significant proportion of cases occur during leisure time activities. Two thirds of cases of SCD or aborted SCD could have been detected by an appropriate pre-participation screening, including history, physical examination and an ECG.

Methods

In cooperation with regional Institutes of Forensic Medicine in Switzerland we designed a web-based database for recording cases of sudden death in athletes. Athletes at an age of 10 to 39 years are included.

Retrospective analysis of case records

In a pilot study, we reviewed case records from the Institute of Forensic Medicine in Bern. In the period from 1999 to 2008 there were 89 cases of SCD in young people aged 10 to 39 years and 13 cases (15%) were associated with sports participation (see Fig. 1). Mean age was 28.9 years in athletes and 29.9 years in non-athletes. In athletes, 5 of 13 cases (38%) occurred in the 36 to 39 year age group. Interestingly, only 4 of 13 cases (31%) were associated with competitive sports. There was a clear male predominance (12 of 13 cases, 92%).

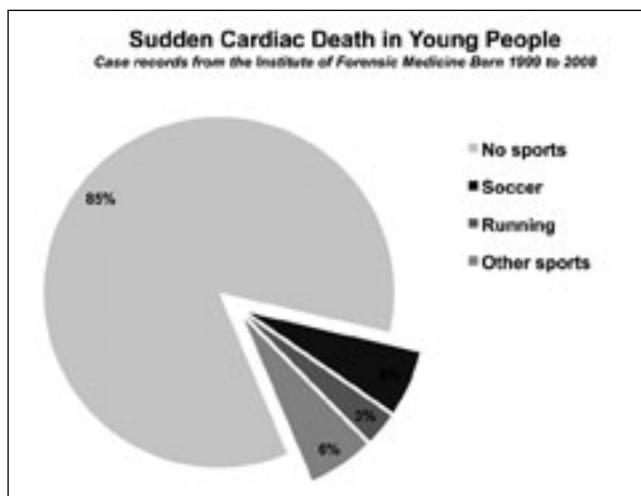


Figure 1: Distribution of cases of Sudden Cardiac Death in young people (10 to 39 years). Of 89 cases, 13 (15%) were associated with sports participation. Most deaths occurred in soccer, followed by running.

Prospective registry

Our prospective registry on sudden death started in 2011. In cooperation with Departments of Cardiology/Divisions of Electrophysiology we plan to extend the registry to cases of aborted sudden cardiac death in 2012. In 2015 we will perform the first evaluation. Online submission of cases is possible via our website www.swissregard.ch (Swiss REGistry of Athletic Related Death) [16]. Example was the U.S. Registry www.suddendeathathletes.org from the Minneapolis Heart Foundation [19]. Database structure of the U.S. Registry was provided by courtesy of Prof. Barry Maron, making international comparisons possible. Personal data, like name or address of the deceased athlete will not be recorded. Subjects will be classified by age, race, gender and sports participation. For this study, we consider competitive athletic as individuals who are engaged in a regular fashion in exercise training as well as participating in official athletic competitions [7]. We consider leisure-time athletes as individuals who are not participating in competitions. Events will be classified in relation to exercise: a) not related to exercise, b) related to non-competitive exercise, c) related to non-elite competitive exercise, d) related to elite competitive exercise.

SCD is defined as an unexpected death as a result of cardiac causes in which a loss of all functions occurred instantaneously or within one hour of the onset of collapse symptoms [5]. If SCD occurred during or 30 minutes after vigorous exertion it is defined as related to exercise [1]. Sudden non-cardiac death is defined as sudden death related to trauma or other non-cardiac conditions.

Autopsies are performed in the regional Institutes of Forensic Medicine following current guidelines for autopsy investigation of sudden cardiac death [2]. In difficult cases, cardiovascular pathologies may be evaluated in the reference center in Lausanne. Results of the pathological analysis are reported in four broad categories, according to the macroscopic and microscopic appearance of the heart: a) morphologically normal hearts, b) cardiomyopathies with a morphological substrate, c) coronary artery pathologies, d) other cardiac pathologies [8]. Common findings are presented in Table 1.

Since probably not all cases of sudden athletic death in Switzerland present at Institutes of Forensic Medicine, reporting of cases is possible for non-professional observers like relatives, other athletes, coaches, and journalists via the website www.swissregard.ch. After a short registration an email is sent with a link to the protected website. A short questionnaire concerning the circumstances of sudden death has to be filled out. Sports journalists have the option to send a report of the event by E-mail.

Normal Heart

Morphologically normal hearts
(Ion Channelopathies like long QT syndrome, short QT syndrome, Brugada syndrome or catecholamine sensitive polymorphic ventricular tachycardia)

Cardiomyopathies

Hypertrophic Cardiomyopathy
Idiopathic LV Hypertrophy Arrhythmogenic Right Ventricular Cardiomyopathy
Dilatative Cardiomyopathy

Coronary Artery Pathologies

Atherosclerosis/ Ischemic Heart Disease
Anomalous origin of coronary arteries
Tunneled coronary arteries

Other cardiac pathologies

Myocarditis
Mitral valve prolapse
Aortic valve stenosis
Dissecting aortic aneurysm
Pulmonary embolism

Table 1: Common findings in Sudden Cardiac Death

Estimation of cases and data analysis

In Switzerland, the population aged 10 to 39 years averages 2 970 950, consisting of 1 487 150 females and 1 483 800 males [10]. Approximately 75% of this population is engaged in regular physical activities, and 20% of the latter are engaged in official athletic competitions [9]. Estimating an annual SCD rate of 2.3:100 000 athletes there will be 51 cases per year in all athletes, including 10 cases per year in competitive athletes.

Two comparisons are reported: 1) Incidence of sudden death/SCD in competitive athletes in comparison to leisure-time athletes and non-athletes. 2) Incidence of sudden death/SCD in competitive athletes and leisure-time athletes in comparison to non-athletes.

Numerators used to calculate annual incidence rate consist of the number of sudden deaths in the corresponding population. The denominator consists of person-years, namely the average size of the corresponding population multiplied with the duration of the observation period. The estimates of numbers of overall person-years at risk are calculated based on the annual data of the Swiss Federal Statistical Office [10]. Estimates of competitive and leisure-time athlete-years at risk are based on the data of the Swiss Federal Office of Sports [9].

Perspective

We aim to examine the incidence and etiology of sport-related sudden death in Switzerland to estimate the national magnitude of the problem. The determination of the proportion of cases that could have been prevented by an appropriate screening will be valuable in the debate of a tailored national pre-participation screening program. Furthermore, the data may help to develop preventive measures for sudden death in sports.

From an international perspective, more studies are needed to clarify the role of ethnicity and the risk of different diseases that cause SCD in young athletes [3]. The Sports Cardiology Section of the European Association of Cardiovascular Prevention and Rehabilitation intends to publish recommendations on national registries on sudden death in athletes. In this regard, our experience with *Swissregard.ch* may help to establish a protocol for an unified European registry on sudden death in athletes.

When SCD affects a supposedly healthy athlete, an autopsy is usually requested for legal reasons. However, on average in 20% of cases the cause of death remains undetermined [20]. Forensic pathologists may face diagnostic difficulties and leave clinicians and family members, without an explanation for the death. There is clear evidence suggesting that genetics play a role in some cases of SCD (Ion channelopathies, see *Table 1*) and that genetic testing can identify individuals at risk for a disease [4]. Moreover, genetic testing may be important in cases with a morphological substrate, like hypertrophic cardiomyopathy or arrhythmogenic rightventricular cardiomyopathy [12, 13].

We need to improve awareness of these diseases in the medical community. We need to improve communication pathways between specialists, primary care and sports physicians and families for genetic testing and counseling of relatives. We need thorough diagnostic evaluation, including genotyping, to improve the comprehensive clinical examination in SCD syndromes for rapid detection of individuals at risk [4]. While the problem of sudden death in sports needs thorough scientific examination, the fact that the overall risk for the athlete is extremely low has to be considered by clinicians when advising athletes, their families and coaches.

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