

10. Sports and sudden death

Authors

Mats Börjesson, MD, PhD, Associate Professor, Department of Medicine, Sahlgrenska University Hospital, Gothenburg, Sweden

Eva Nylander, MD, PhD, Professor, Division of Cardiovascular Medicine, Linköping University Hospital, Linköping, Sweden

Erik Ekker Solberg, MD, PhD, Diakonhemmet Hospital, Oslo, Norway

Summary

Cases of sudden death (SCD) among young athletes are usually due to rare previously undiagnosed hereditary and/or congenital cardiovascular diseases. The Swedish National Board of Health and Welfare recommends cardiac screening of risk groups, such as those with known cardiovascular diseases, or those with a family history of sudden cardiac death, or people who have alarming symptoms in connection with physical exertion. However, tracing individuals with hidden cardiovascular disease through cardiac screening of all adolescents or athletes or those who are routinely physically active is not recommended. This is because the diseases are rare and the diagnostics are not sufficiently accurate to identify all of those with the diseases, which leads to problems with both false negatives and false positives.

However, the Swedish Sports Confederation as well as the Swedish National Board of Health and Welfare recommend targeted heart check-ups of elite athletes.

Definition

Sudden cardiac death is commonly defined as “death occurring within one hour of the onset of symptoms in a person with previously known or unknown heart disease”, if death was witnessed (1), or “death within 24 hours of the person having been seen alive and well with no other known cause” if death is not witnessed.

Causes

The cause of sudden death related to sports or physical activity, not related to trauma or accident, is almost always heart disease. In this context, athletes are divided into younger

(< 35 years-old) and older (> 35 years-old), which is of importance regarding the specific disease that causes death (2).

In athletes over the age of 35, the cause is almost exclusively coronary artery disease/myocardial infarction. In persons with an underlying coronary disease, there is an elevated risk of sudden death in connection with intense physical activity. However, this risk decreases in persons who pursue regular physical activity. Regular, individually tailored physical activity is also of considerable health benefit for those with established coronary disease, in part due to the positive effects on classical risk factors and endothelial function. Sudden death resulting from coronary artery disease among people over the age of 35 will not be discussed in more detail in this chapter.

In those under the age of 35, virtually all cases of SCD involve people who have an undiagnosed heart condition that predisposes them to a life-threatening arrhythmia triggered by strenuous physical activity. There are several relatively uncommon diseases that can cause sudden cardiac death among young people who have lived unaware of the underlying condition. The most common is a heart muscle disease (so-called cardiomyopathy, the most common of which is hypertrophic cardiomyopathy, which has a prevalence of approximately 1/500), malformations of the coronary arteries and diseases that affect the heart rhythm and the conducting system, so-called ion channel diseases.

Prevalence/Incidence

The prevalence of sudden cardiac death in persons under the age of 35 is approximated to 1–2/100,000 individuals per year. In total, around 0.3 per cent of young people suffer from congenital/hereditary heart defects with an elevated risk of sudden death during intense physical activity. The causes of sudden cardiac death in this group may be population specific, but hypertrophic cardiomyopathy is most common, followed by coronary artery anomaly, arrhythmogenic right ventricular cardiomyopathy, which causes malignant arrhythmia, followed by ion channel diseases, Wolff-Parkinson-White (WPW) syndrome, valvular heart disease, Marfan syndrome and myocarditis.

Diagnostics

Examinations intended to find individuals with the diseases associated with an elevated risk of sudden cardiac death in connection with sports have been frequently discussed in recent years. This identification is possible, since a large part of the diseases are hereditary and many are also accompanied by findings on a 12-lead-resting electrocardiogram (ECG). Moreover, the bearer of the disease can often have symptoms that may incite suspicion of illness, but that sometimes could be ignored by the athlete himself and by those in their surroundings.

These facts constitute the background of the recommendations from the Swedish National Board of Health and Welfare (3) that people who have:

1. relatives that have died suddenly and unexpectedly before the age of 40,
2. first-degree relatives with diagnosed hypertrophic cardiomyopathy or another cardiovascular disease associated with an elevated risk,
3. “alarming symptoms” during sports activities, such as chest pain, dizziness, a feeling of fainting (pre-syncope), distinct tachycardia or abnormal breathlessness (dyspnoea),
4. demonstrated abnormal findings on ECG, shall be examined with regard to the diseases in question (see below).

Targeted cardiac screening of athletes

The European Society of Cardiology (ESC) recommendations

Should all athletes have a health exam to identify persons who have a hidden heart disease? This has been discussed and has been handled differently in different countries. In the U.S., recommendations for “preparticipation screening” of athletes have existed for many years, consisting of questions about family history, symptoms and a physical examination. No ECGs are recommended routinely in the U.S. at the present time (4). Since the beginning of the 1980s, Italy has introduced mandatory screening of all “competitive athletes” consisting of medical history, physical examination and resting ECG (5).

In 2005, the European Society of Cardiology (ESC) presented a proposal for a common European protocol concerning cardiac screening of individuals who participate in competitive sports (6). The objective is to prevent sudden cardiac death by finding potential cardiovascular abnormalities that could convey an elevated risk of sudden death during intense physical exertion.

The European expert group recommends a systematic cardiovascular evaluation of everyone who is to participate in organised competitive sports. The recommended protocol comprises the following:

1. personal and family medical history (symptoms and heredity),
2. clinical examination,
3. 12-lead resting ECG.

If no relevant findings are made on this screening, the person is judged to be eligible for competitive sports. Upon abnormal findings, further examinations are needed by a physician experienced in sports cardiology. Examinations such as echocardiography, maximum exercise tests, 24-hour ECG, MRI, angiography/heart muscle biopsy and electrophysiological examinations may be advocated.

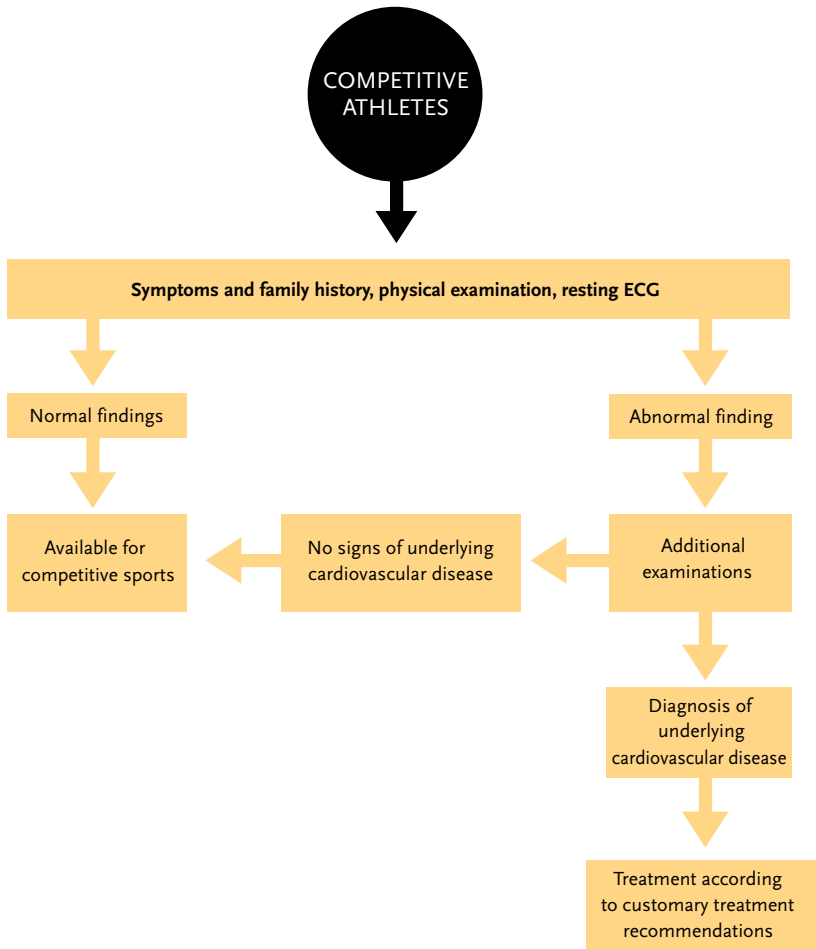


Figure 1. European Society of Cardiology's screening recommendations. Adapted from: Corrado et al. *Eur Heart J* 2005.

Nordic recommendations

Previously, no screening was recommended in the Nordic countries. They were historically even able to compete internationally without having been subjected to targeted cardiac screening, although regular pre-season examinations were conducted, focusing on the musculoskeletal system.

The ESC recommends that every country adapt the existing recommendations to their specific healthcare systems, including making practical adjustments with regard to access to sports medicine expertise.

The discussion in the Nordic countries in recent years has focused on the problems versus the benefits of screening. Screening as the term is commonly used, meaning looking for disease in large population groups without elevated risk, does not meet the WHO criteria. In addition, the diseases are too rare and the diagnostic methods do not have sufficient sensitivity or specificity. The expression screening may therefore be somewhat inappropriate, since most advocate a more targeted examination limited to specified risk groups. Consequently, the expression “targeted cardiovascular examination” may be more relevant.

In Sweden, a review of existing procedures has been conducted and, since 2005, the Swedish Sports Confederation has recommended that the examinations including medical history, physical examination and resting ECG should be confined to elite athletes (7). “Elite” is defined by the respective sporting associations and includes students at national sports gymnasiums. Since 2006, the Swedish National Board of Health and Welfare recommends (3) screening of risk groups as well as elite athletes.

The debate continues in the other Nordic countries, but no formal recommendations from the authorities have yet been issued (8). The Danish Society of Cardiology emphasizes the importance of targeted screening of certain risk groups with symptoms and a positive family history, but does not propose any general screening. Authors from Finland and Sweden have recently proposed a “Nordic approach” where cardiac screening is limited to elite athletes in sports with a considerable strain on the cardiovascular system (most). Education is also proposed to provide better knowledge about risk groups in both sports and among non-athletes (9).

Does screening save lives?

In Italy, where cardiac screening of competitive athletes has been conducted since the 1980s, a decrease in the incidence of sudden cardiac death from 4.2/100,000 per year to 0.4/100,000 per year (10) during the period 1979–2004 has been shown. However, this was not a controlled study and factors other than screening may have played a role.

A number of international sports associations also currently recommend, or plan to introduce, mandatory cardiac screening for participating athletes in international elite contexts (UEFA, FIFA).

Recommendation summary

- Greater preparedness in healthcare and the school system is proposed to find first-degree relatives of individuals with heart disease that is associated with an elevated risk of sudden death, regardless of whether they participate in sports or not. Similarly, greater awareness of these problems in the sports community is of value.
- Awareness of exertion-related symptoms, in the form of chest pains, dizziness, syncope, tachycardia attacks or abnormal breathlessness (dyspnea) is recommended, and athletes with such symptoms should be examined at the appropriate level of the healthcare system.
- Elite athletes are recommended targeted cardiac screening with medical history, physical examination and resting ECG. This is the responsibility of organised sports (clubs, sports schools), primarily through their respective team physicians/school doctors. However, abnormalities found should lead to a referral for further examination in a healthcare facility with good knowledge in sports cardiology, sports physiology and diagnostics.

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