Periodical cardiovascular screening is mandatory for elite athletes

Pro screening

H. Inklaar, N.M. Panhuyzen-Goedkoop

Sudden cardiac death (SCD) of an athlete is a very tragic event. When it concerns a well-known elite sportsman, it creates much publicity. Inevitably questions are raised about the health-promoting value of sports and the necessity of a mandatory periodical cardiovascular screening to prevent sports-related cardiac events. In order to answer the final question many aspects of SCD and screening in general have to be addressed.

SCD is defined as nontraumatic, nonviolent, unexpected death due to cardiac causes within one hour of the onset of symptoms. Sports-related SCD are defined as those with symptoms occurring within one hour of sports participation.¹

Cardiac causes of SCD in the young population (≥ 35 years) are inherited (long-QT syndrome, short-QT syndrome, Brugada syndrome, Wolff-Parkinson-White syndrome, arrhythmogenic right ventricular cardiomyopathy, hypertrophic cardiomyopathy, anomalous coronary artery) or congenital diseases, and myocarditis. In the older population (> 35 years) cardiac causes of SCD are often acquired diseases, such as coronary artery disease.

As with any other health screening procedures, a systemic preventive cardiovascular screening of young athletes should meet a number of conditions to be meaningful (table 1).²

The incidence of SCD in young athletes (aged 12 to 35 years) is 0.5 to 2/100,000 per year and is ap-

proximately 2.5 times higher than in nonathletes, with a male-female ratio 10:1.³⁻⁵ Based on information from case series, the number of sports-related SCD in the Netherlands is estimated at 150 to 200 persons a year.⁶ About 40% of SCD in sports concerns the age group 12 to 35 years. The male-female ratio is 10:1. These numbers are probably underestimated due to the lack of an official registry for SCD in sports in the Netherlands, the lack of a mandatory postmortem analysis in case of SCD in sports, the lack of sufficient experience with the cardiac conditions on the part of pathologists performing postmortem analysis in young athletes and some conditions leading to fatal arrhythmias without microscopic structural heart disease at postmortem analysis.

In a systematic literature review congenital anatomical heart diseases (33%), cardiomyopathies (31%) and atherosclerosis (11%) represent 75% of the cases of SCD.⁴

Sports with the highest cardiovascular demands and the highest isotonic work involve more risk for SCD.⁴ However the sport itself is not per se the cause of the increased mortality; it rather acts as a trigger for underlying cardiovascular diseases predisposing to life-threatening ventricular arrhythmias during physical exercise.^{4,5} It is currently not known if elite athletes run a higher risk of SCD than athletes at lower levels of competition and recreational athletes. Doping as a main cause of SCD seems unlikely, as underlying cardiac diseases account for approximately 90% of exercise-related sudden deaths. However, use of prohibited substances may trigger lethal arrhythmia in athletes with underlying (silent) inherited cardiac disorder.⁷

In the recommendations for determining eligibility for competition in athletes with cardiovascular abnormalities issued by the American Heart Association it was stated that preparticipation screening for young athletes is justifiable and compelling on ethical, legal and medical grounds.⁸

A nationwide systemic preparticipation screening to obtain eligibility for engagement in competitive sports activity was introduced by law in Italy in 1982.^{5,9} The cardiovascular screening part consists of the

H. Inklaar

University Centre for Sports Medicine, location KNVB Sport Medical Centre, Zeist, the Netherlands; Working Group on Cardiovascular Screening and Sports

N.M. Panhuyzen-Goedkoop

Heart Centre, Radboud University Medical Centre, Nijmegen and Sint Maartenskliniek, location Sport Medical Centre Papendal, Arnhem, the Netherlands; Working Group on Cardiovascular Screening and Sports

Correspondence to: H. Inklaar
KNVB Sport Medical Centre, PO Box 515, 3700 AM Zeist, the Netherlands
E-mail: han.inklaar@knvb.nl

Table 1. Criteria for screening procedures.²

- Important community health problem
- Detectable asymptomatic phase
- Valid, safe and generally accepted test procedure
- Treatment with proven favourable effects in the asymptomatic phase
- Cost-effectiveness
- Do no harm principle

combination of history, physical examination and a 12-lead resting ECG. There is also an official registry of SCD in athletes and nonathletes and an obligation to perform postmortem analysis in case of SCD that does not require the consent of relatives.

Corrado et al. showed that this screening programme was valid with respect to the detection of hypertrophic cardiomyopathy (HCM) with a high specificity (91%).⁸ As only 9% are false-positive, the low cost of the ECG justifies the small percentage of unnecessary echocardiographies.

The validity with respect to the detection of other causes of SCD is limited, although in time the validity with respect to the detection of arrhythmogenic right ventricular cardiomyopathy greatly improved.⁹

A trend analysis over a 26-year period of screening in the Veneto region showed that 2.0% of the athletes were disqualified from competition due to cardiovascular causes.⁹ The proportion of athletes disqualified due to cardiomyopathies increased from 4.4% in the early screening period to 9.4% in the late screening period. The incidence of SCD in young competitive athletes has substantially declined in the Veneto region since the introduction of the nationwide systematic screening and has even declined below the incidence of SCD in nonathletes. Mortality reduction was predominantly due to the lower incidence of SCD from cardiomyopathies that parallel the increasing identification of athletes with cardiomyopathies at preparticipation screening. The question remains if all athletes with detected HCM should be expelled from competition, since it has been shown that the prognosis of HCM varies with the genotype and the phenotype.¹¹

And one can wonder if not all young individuals should be screened for HCM. Due to the low prevalence of SCD the costs of this screening represent an important investment. Corrado et al. calculated that it costs € 28,450 in Italy for each correct diagnosis of HCM.¹⁰ However, the cost efficiency of their screening programme was far better than the cost efficiency of mass screening programmes conducted at the colleges and high schools in the USA using only a combination of history and physical examination. In a Minneapolis registry in the United States, HCM was a common cause of SCD in young unidentified African-American athletes, and would justify 12-lead resting ECG in the

preparticipation cardiovascular screening programme in young athletes.¹²

Bille et al. concluded from a systematic review of the literature that the large variety of preparticipation screening protocols and the variability of persons using them prevent serious evaluation of their efficacy.⁴ Mainly based on the Veneto study, the consensus meeting on SCD in athletes, organised by the Medical Commission of the IOC, launched the Lausanne Recommendations for the prevention of SCD in 2004.¹³ The European Society of Cardiology study group of Sports Cardiology proposed that all young competitive athletes should be screened according to a common European protocol.¹³ The purpose of the Lausanne Recommendations and the European protocol is to present a uniform and generally acceptable screening procedure. The Lausanne protocol uses a stepwise approach. Step 1 is a basic screening, with a questionnaire (yes/no answers) concerning personal history and family history, followed by physical examination and a 12-lead ECG at rest.

The screening physician should be experienced in possible cardiac problems of intense sports activity and in reading the ECG. Step 2 follows step 1 in case of positive findings. These further evaluations and investigations are the concern of a (sports)cardiologist.

Although the questionnaire has not been validated, the Lausanne Recommendations are now widely adopted by international sports organisations. In professional sports, a more extensive cardiovascular screening has often already been introduced, by adding echocardiography, stress ECG and sometimes even MRI to step 1 of the Lausanne Recommendations.

In its yearly report on health screening in the Dutch population the Health Council concludes that there is no scientific basis for recommendations for periodical screening of competitive athletes for SCD.¹⁴ Screening is of unproven value for the prevention of SCD, and carries a risk for false-positive results, overdiagnosis and stigmatisation.

With respect to mass screening of competitive athletes, this conclusion is agreed upon. However, this report did not have the data of the trend analysis of the incidence of SCD in the Veneto region where the incidence of SCD in screened athletes has declined below the incidence of SCD in nonathletes.

The working group on cardiovascular screening and sports (participants: Netherlands Association of Cardiology, Association of Sports Medicine, National Olympic Committee, Royal Netherlands Football Association) suggests increasing the awareness of the sporting population for the risk of SCD through mass media education.¹⁵ Also an abbreviated version of the questionnaire from the Lausanne Recommendations may be downloaded (www.sportzorg.nl). Athletes with positive answers to these questions are advised to consult their family physician or a sports physician for

further screening. The responsibility for their health therefore lies with the athletes.

Because of the lack of reliable data on the incidence of SCD in the Netherlands and in many other countries a mandatory official nationwide registry of SCD in sports is advocated.^{15,16} The same goes for an obligation towards postmortem analysis of cases of SCD in sports. Because sudden unexplained deaths (SUD) may have heritable causes, a nationwide registry offers the opportunity for a large-scale cardiological and genetic assessment of surviving relatives of SUD victims and may reveal the underlying disease and unmask pre-symptomatic carriers.¹⁶ By examining relatives of young SUD victims, Tan et al. were able to identify the disease in 40% of families and 8.9% presymptomatic carriers.¹⁷ Contrary to the conclusion of the Health Council a periodical cardiovascular screening of elite athletes seems mandatory. In many sports organisations and professional sport clubs elite athletes in different age categories are already subjected to periodical health screening. Because of the high investments in elite athletes, these organisations demand to know if there are any substantial intrinsic health risks and if their investments are worthwhile.

This provides the opportunity to include the Lausanne Recommendations in the already existing periodical health screening of elite athletes and to collect data for further analysis.

The Working Group on Cardiovascular Screening and Sports advocates the institution of a database on sports cardiology (SportCor). The aim of this database is to develop a good organisation structure for collection, analysis and distribution of sports cardiological data in the Netherlands. Through this database many still unresolved questions may be addressed, such as:

- What developments do ECGs of elite athletes show in the course of their sports career?
- Which ECG features of elite athletes should be considered as abnormal or as normal variants following adaptation of the heart to high intensive sports activities?
- Will it be possible to increase the diagnostic value of the ECG in elite athletes through better understanding of ECG features in this subgroup?¹⁸
- What is the predictive value of the Lausanne Recommendations?
- Are certain ECG features linked to states of overtraining in elite athletes?

Finally, the information gathered in this database may be used to increase the knowledge of cardiologists and (sports)physicians in the field of sports cardiology.

Conclusively, a periodical cardiovascular screening is mandatory for elite athletes. ■

References

- 1 26th Bethesda Conference. Recommendations for determining eligibility for competition in athletes with cardiovascular abnormalities. *Am J Coll Cardiol* 1994;24:845-99 and *Med Sci Sports Exerc* 1994;26:S223-S283.
- 2 Whithby LG. Screening for disease, Definitions and criteria. *Lancet* 1974;333:819-21.
- 3 Maron BJ, Gohman TE, Aeppli D. Prevalence of sudden cardiac death during competitive sports activities in Minnesota high school athletes. *J Am Coll Cardiol* 1998;32:1881-4.
- 4 Bille K, Figueiras D, Schamash P, Kappenberger, Brenner JJ, Meijboom FJ, et al. Sudden cardiac death in athletes: the Lausanne Recommendations. *Eur J Cardiovasc Prev Rehab* 2006;13:1-17.
- 5 Corrado D, Pelliccia A, Bjornstad HH, Vanhees L, Biffi A, Borjesson M, et al. Cardiovascular pre-participation screening of young competitive athletes for prevention of sudden death: proposal for a common European protocol. *Eur Heart J* 2005;26:16-24.
- 6 Dolmans AJ, Pool J, Erdman-Trip JF, Smit B, Lubsen J. Het risico van overlijden bij sport. *Ned Tijdschr Geneesk* 1984;128:595-8.
- 7 Deligiannis A, Bjornstad H, Carre F, Heidbuchel H, Kouidi E, Panhuyzen-Goedkoop NM, et al. ESC study group of sports cardiology position paper on adverse cardiovascular effects of doping in athletes. *Eur J Cardiovasc Prev Rehab* 2006;13:687-94.
- 8 Maron BJ, Thompson PD, Puffer JC, McGrew CA, Strong WB, Douglas PS, et al. Cardiovascular preparticipation screening of competitive athletes. A statement for health professionals from the sudden death committee (clinical cardiology) and congenital cardiac defects committee (cardiovascular disease in young), American Heart Association. *Circulation* 1996;94:850-6.
- 9 Corrado D, Basso C, Pavei A, Michieli P, Schiavon M, Thiene G. Trends in Sudden Cardiovascular Death in Young Competitive Athletes After Implementation of a Preparticipation Screening Program. *JAMA* 2006;296:1593-601.
- 10 Corrado D, Basso C, Schiavon, Thiene G. Identification of athletes with hypertrophic cardiomyopathy at risk of sudden death: cost-effectiveness analysis of screening strategies. *Circulation* 2002;106 (abstract): II-701.
- 11 Elliott P, McKenna WJ. Hypertrophic cardiomyopathy. *Lancet* 2004;363:1881-91.
- 12 Maron BJ, Carney KP, Lever HM, Lewis JF, Barac I, Casey SA, et al. Relationship of race to sudden cardiac death in competitive athletes with hypertrophic cardiomyopathy. *J Am Coll Cardiol* 2003;41:974-80.
- 13 Corrado D, Pelliccia A, Bjornstad HH, Vanhees L, Biffi A, Borjesson M, et al. Cardiovascular pre-participation screening of young competitive athletes for prevention of sudden death: proposal for a common European protocol. *Eur Heart J* 2005;26: 516-24.
- 14 Gezondheidsraad. Jaarbericht bevolkingsonderzoek Den Haag: *Gezondheidsraad*, 2006; publicatie nr 2006/10.
- 15 Panhuyzen-Goedkoop NM, Pluim BM, Senden PJ, Hoogsteen J, Bennekens H, Inklaar H, et al. Preventie van plotse dood in de sport bij jonge atleten in Nederland (consensusdocument). *Geneesk en Sport* 2005;38:107-12.
- 16 Tanaka Y, Yoshinaga M, Anan R, Tanaka Y, Nomura Y, Oku S, et al. Usefulness and cost effectiveness of cardiovascular screening of young athletes. *Med Sci Sports Exerc* 2006;38:2-6.
- 17 Tan HL, Hofmans N, Langen van IM, Wal van der AC, Wilde AA. Sudden unexplained death: Heritability and Diagnostic Yield of Cardiological and Genetic Examination in Surviving Relatives. *Circulation* 2005;112:207-13.
- 18 Pelliccia A, Maron BJ, Culasso F, Di Paolo FM, Spataro A, Biffi A, et al. Clinical significance of abnormal electrocardiographic patterns in trained athletes. *Circulation* 2000;102:278-84.