UNIVERSITE DE LAUSANNE

FACULTE DE BIOLOGIE ET DE MEDECINE

DEPARTEMENT MEDICO-CHIRURGICAL DE PEDIATRIE

Service de Pédiatrie

Chef de service : Professeur Sergio Fanconi

Sudden Cardiac Death in Athletes: the Lausanne Recommendations

THESE

préparée sous la direction du
Professeur Erik J. Meijboom

et présentée à la Faculté de Biologie et de
Médecine de l'Université de Lausanne
pour l'obtention du grade de

DOCTEUR EN MEDECINE

par

Karin Bille Lemaire

Médecin diplômée de la Confédération Suisse
Originaire de Boudevilliers (NE)

Lausanne
2006

La mort subite d'origine cardiaque chez les sportifs : les Recommandations de Lausanne

Résumé

Objectifs :
Cette étude collecte les données de la littérature scientifique concernant la mort subite d'origine cardiaque chez les sportifs et a pour but d'aboutir à un protocole d'investigation de préparticipation globalement acceptable, approuvé par la conférence de consensus du Comité International Olympique(CIO), et recommandé par ce dernier.
Données cliniques :
La mort subite chez les athlètes de moins de 35 ans, engagés dans des sports de compétition, est un évènement bien connu, dont l'incidence est plus élevée (~2/100000/an) que chez les non-athlètes (2,5 :1). La cause est cardiovasculaire dans plus de 90% des cas.
Méthodes :
Une revue systématique de la littérature a mis en évidence les causes de mort subite d'origine cardiaque, le sexe, l'âge, les maladies cardiaques sous-jacentes et le type de sport, ainsi que les protocoles d'investigation de préparticipation utilisés. Les méthodes nécessaires pour détecter des anomalies cardiaques préexistantes sont discutées pour formuler un protocole d'investigation de préparticipation pour la commission médicale du CIO.
(http://www.olympic.org/uk/organisation/commissions/medical/full_story_uk.asp?id=1182)
Résultats :
1101 cas de mort subite d'origine cardiaque ont été rapportés (1966-2004) chez des athlètes de moins de 35 ans, 50% présentant des anomalies cardiaques congénitales et des cardiomyopathies et 10% une athérosclérose à début précoce. 40% des athlètes avaient moins de 18 ans, 33% moins de 16 ans ; le rapport femme/homme était de 1/9. La mort subite d'origine cardiaque était reportée dans presque tous les sports ; ceux impliqués le plus fréquemment étaient le football(30%), le basketball(25%), et la course à pied(15%). Les tests de préparticipation étaient de qualité et de contenu variables.
La conférence de consensus du CIO a accepté les « Recommandations de Lausanne » proposées, basées sur cette recherche et des opinions d'experts.
(http://multimedia.olympic.org/pdf/en_report_886.pdf)
Conclusions :
La mort subite d'origine cardiaque touche plus souvent qu'attendu les jeunes athlètes et est principalement due à des anomalies cardiaques congénitales préexistantes. Les atteintes athérosclérotiques précoces forment une autre cause importante de décès chez les jeunes adultes. L'acceptation par le CIO de « Recommandations de Lausanne » a permis d'aboutir à un protocole d'investigation de préparticipation globalement acceptable.

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Position Paper

Sudden cardiac death in athletes: the Lausanne Recommendations

Karin Bille[a], David Figueiras[b], Patrick Schamasch[c], Lukas Kappenberger[c], Joel I. Brenner[c], Folkert J. Meijboom[c] and Erik J. Meijboom[a]

[a]Divisions of Pediatric Cardiology and Cardiology, University Hospital of Lausanne, Lausanne, [b]Institute of Sport Sciences, University of Lausanne, Lausanne and [c]Members of the ad hoc Working Group on Sudden Death in Athletes of the Medical Committee of the International Olympic Committee, Lausanne, Switzerland.

Received 23 March 2006 Accepted 15 June 2006

Objectives This study reports on sudden cardiac death (SCD) in sport in the literature and aims at achieving a generally acceptable preparticipation screening protocol (PPSP) endorsed by the consensus meeting of the International Olympic Committee (IOC).

Background The sudden death of athletes under 35 years engaged in competitive sports is a well-known occurrence; the incidence is higher in athletes (~2/100 000 per year) than in non-athletes (2.5 : 1), and the cause is cardiovascular in over 90%.

Methods A systematic review of the literature identified causes of SCD, sex, age, underlying cardiac disease and the type of sport and PPSP in use. Methods necessary to detect pre-existing cardiac abnormalities are discussed to formulate a PPSP for the Medical Commission of the IOC.

Results SCD occurred in 1101 (1966–2004) reported cases in athletes under 35 years, 50% had congenital anatomical heart disease and cardiomyopathies and 10% had early-onset atherosclerotic heart disease. Forty percent occurred in athletes under 18 years, 33% under 16 years; the female/male ratio was 1/9. SCD was reported in almost all sports; most frequently involved were soccer (30%), basketball (25%) and running (15%). The PPSP were of varying quality and content. The IOC consensus meeting accepted the proposed Lausanne Recommendations based on this research and expert opinions (http://multimedia.olympic.org/pdf/en_report_886.pdf).

Conclusion SCD occurs more frequently in young athletes, even those under the age of 18 years, than expected and is predominantly caused by pre-existing congenital cardiac abnormalities. Premature atherosclerotic disease forms another important cause in these young adults. A generally acceptable PPSP has been achieved by the IOC's acceptance of the Lausanne Recommendations. *Eur J Cardiovasc Prev Rehabil* 13:000–000 © 2006 The European Society of Cardiology

European Journal of Cardiovascular Prevention and Rehabilitation 2006, 13:000–000

Keywords: cardiovascular, electrocardiogram, hypertrophic cardiomyopathy, preparticipation screening protocol, sudden cardiac death

Introduction

Active participation in sports is generally considered to be beneficial for the health and well being of the individual [1]. The increased risk of sudden death in adolescents and young adults engaged in competitive sports, a well-known and much feared occurrence, seems to contradict this adagium [2–4]. The leading cause of this mortality is underlying cardiac disease (~90%) [5–7], triggering sudden death during intensive physical exercise. The non-cardiac causes of sudden death include asthma (or other pulmonary conditions), heat stroke, drug abuse, cerebral embolism, ruptured cerebral artery and some unexplained causes.

The incidence of sudden cardiac death (SCD) in young athletes (aged 12–35 years) is 0.5–2/100 000 per year [4,6,8–10], and is approximately 2.5 times higher than in non-athletes [11]. This increased mortality has led to the

Correspondence and requests for reprints to Erik Jan Meijboom, MD, PhD, Division of Pediatric Cardiology, Bugnon 46, CHUV, BH-11.608, CH-1011, Lausanne, Switzerland.
Tel: +41 21 314 3553; fax: +41 21 314 3665;
e-mail: erik.meijboom@chuv.ch

implementation of various preparticipation screening protocols (PPSP), showing a wide difference depending on the mandating organizations or countries. Some are quite specific and involve serious physical examinations, whereas others are merely questionnaires. The quality of each of these PPSP depends greatly on the way the examinations are carried out, and varies from inadequate to thorough.

A generally accepted and implemented PPSP would be preferable, but the difficulty in achieving a consensus is located in the vast number of governing bodies involved. Several governments have by law implemented different regulations, various sports organizations have started to use their own PPSP, and a diversity of medical organizations have issued, sometimes very different, guidelines.

The aim of our review of the literature was to analyse the reported cases of SCD among young athletes, to identify the sports with the highest risk of SCD, and to compare the PPSP in use.

This was to allow an evaluation of the effect of the different methods and to assemble the parameters required to propose to the International Olympic Committee (IOC) a generally acceptable and safe screening.

Methods

For a meta-analysis of the existing literature on sudden death in sports and the effectiveness of PPSP, the following databases were searched: Medline (OVID Web, 1966–2004), PubMed (1966–2004), Cochrane Database of Systematic Reviews, EBM Reviews – ACP Journal Club, Cinahl 1982–2004, Heracles, Web of Science, Scopus < 1960–2004. We used 'sudden cardiac death' and 'preparticipation' as keywords, combined with 'sports', 'sports medicine', 'athletic injuries', 'exercise' and 'athletes'. Restrictions were English language and human subjects. To be overinclusive, there were no restrictions concerning age, sex, or study methodology. The keyword search yielded MeSH headings, which were exploded before they were combined. For study and data selection, we reviewed the results of the search and selected and grouped all studies reporting on SCD (reported cases) and the use of PPSP. Sudden death in athletes by other causes such as neurovascular, heat and doping were excluded and will not be discussed.

The initial search identified 2866 articles (SCD 1493 plus PPSP 1532) after duplicates were removed.

Sudden cardiac death

A total of 1493 articles were identified by the initial search; 124 were relevant to SCD in athletes, and were available, after review of all the titles and abstracts. We focused on athletes under 35 years of age, as the causes of SCD change past 35 years, atherosclerotic coronary artery diseases becoming much more frequent (84 versus 2% before 35 years) [5,12–18]. Content experts within the medical profession were contacted as well as the relevant sports organizations to identify studies missed by electronic searches.

We used the following criteria to select the articles relevant to our study: articles reporting cases of SCD in athletes, aged up to 35 years, exercise-related, and in the absence of evidence of drug abuse.

SCD is defined as a death occurring within 1 h of the onset of symptoms in a person without a previously recognized cardiovascular condition that would appear fatal. An athlete is defined as a person participating in an organized team or individual sport that requires systematic training and regular competition against others, and that places a high premium on athletic excellence and achievement [19,20].

There were 47 articles left after this final selection (Table 1), which we analysed.

Preparticipation screening protocols

A total of 1532 articles were identified by the initial search; 108 were relevant to PPSP in athletes, and were available, after review of all the titles and abstracts. Content experts within the medical profession were contacted as well as the relevant sports organizations to identify studies missed by electronic searches.

We used the following criteria to select and group the articles relevant to our study: (i) articles describing the PPSP procedure (cardiovascular part) and using it (± reported cases; Table 2); (ii) articles describing the PPSP procedure (cardiovascular part) but not using it (no reported cases; Table 3); (iii) articles about PPSP, specifically electrocardiography (ECG) or echocardiography [41,43,45,46,48,50–53]. Thirty-one articles were selected, with 11 in (i), 11 in (ii), and nine in (iii), and were analysed.

These groupings were reviewed and approved by an ad hoc consensus meeting of the IOC working group on Sudden Death in Athletes including several authors of the primary studies included and the Medical and Scientific Director of the IOC Medical Commission.

Outcomes of the studies were summarized for strength of evidence. The definitions of outcome of the authors of the studies included were accepted.

Results

Sudden cardiac death

Forty-seven articles were analysed and are represented in Table 1. The references of the articles were noted

Table 1 **Selected articles on sudden cardiac death (SCD)**

Authors/journal	Type of article/study	No. of cases of SCD	Sex (m, male; f, female)	Previous symptoms	Clinical data	Autopsy	Cardiovascular diagnosis
[21] Basso, JACC 2000	Review	27	22 m/5 f	Yes in 10	Yes in 12	27	27 AOCA
[22] Bharati, JACC 1983	Case reports	3	2 m/1f	?	Yes	3	1 MVP 3 Sclerosis of ventricular septum with involvement of conduction system
[23] Biffi, JACC 2002	Prospective	1	1 m	?	Yes	?	1 ARVD
[24] Burke, JACC 1993		27	17 m/10 f	Yes in 4	?	27	27 Presumed cardiac arrhythmias by non-atherosclerotic narrowing of the AV node artery
[25] Burke, Am Heart J 1991		34	31 m/3 f	?	?	30	9 Severe atherosclerosis 8 HCM 3 Idiopathic LV hypertrophy 4 Anomalous coronary artery 2 Myocarditis 1 RV dysplasia 1 Kawasaki 6 Unknown (2 tunnel arteries)
[26] Cheitlin, Circulation 1974		9+1	9 m+1 m	?+yes	?	9 (+1 aborted SCD)	9+1 AOCA
[27] Corrado, Circulation 2001	Prospective	31	24 m/7 f	Yes in some	?	31	31 ARVD
[28] Corrado, Am J Med 1990	Post-mortem	22	19 m/3 f	Yes in 9	Yes in some	22	6 ARVD 4 Atherosclerotic CAD 3 Conduction system pathology 2 AOCA 2 MVP 2 Mechanical causes 3 Cerebral causes
[11] Corrado, JACC 2003	Prospective	55	50 m/5 f	Yes in 18	Yes in some	55	12 ARVD 10 Atherosclerotic CAD 7 AOCA 6 MVP 5 Myocarditis 4 Conduction system pathology 2 Myocardial bridge 1 HCM 1 DCM 1 LQTS 2 Mechanical causes 7 Non-cardiovascular causes
[29] Corrado, Br Heart J 1992	Postmortem	12	8 m/4 f	Yes in 6	Yes in some	12	3 AOCA 5 Intramyocardial course of LADCA 1 Intramyocardial course of LMCA 1 Slit-like lumen of left coronary ostium 2 Valve-like ridge of right coronary ostium
[30] *Corrado, NEJM 1998* *Cf. study 9*	*Prospective*	*49*	*44 m/5 f*	*Yes in 14*	*Yes in some*	*49*	*11 ARVD* *9 Coronary atherosclerosis* *6 AOCA* *5 MVP* *4 Disease of conduction system* *3 Myocarditis* *2 Myocardial bridge* *1 DCM* *1 HCM* *2 Mechanical causes* *5 Other*

Table 1 *(continued)*

Authors/journal	Type of article/study	No. of cases of SCD	Sex (m, male; f, female)	Previous symptoms	Clinical data	Autopsy	Cardiovascular diagnosis
[31] Deady, J Emerg Med 1999	Case report	1	1 m	No	?	1	1 Commotio cordis
[32] Fornes, Am J Forens Med Pathol 2003		19	18 m/1 f	No	?	19	4 ARVD 4 HCM 3 CAD (1 + thrombosis) 2 Bridging of LADCA 2 Myocarditis (1 chronic + 1 acute) 1 Anatomical anomaly of CA 3 Mechanical cause (rupture of aortic aneurysm)
[33] Goldschmidt, Lancet 1996	Case report	1	1 m	No	Yes	1	1 CAD
[34] Hoogsteen, Netherlands Heart J 2004	Case reports	2	2 m	Yes in 1	Yes	2	2 ARVD
[35] Iskandar, Med Sci Sports Exerc 2004	Case report	1	1 m	Yes	Yes	1	1 Coronary artery anomaly
[36] Jokl, JAMA 1970	Case reports	2	2 m	?	?	2	1 AOCA with underdevelopment of the entire arterial system 1 Subacute myocarditis
[37] Khoury, Eur Heart J 1994	Case report	1	1 m	?	?	No (aborted SCD)	
[38] Kramer, Chest 1988	Retrospective	24	?	Yes in some	Yes in some	24	7 Myocarditis 6 HCM 3 Ischaemic heart disease 3 MVP 2 Marfan's syndrome 1 Congestive cardiomyopathy 1 Conduction abnormality 1 AOCA
[39] *Larsson, APMIS 1999 Cf study by Wesslen*	*Retrospective*	*16*	*15 m/1 f*	*Yes in 5*	*Yes in most*	*16*	*7 Myocarditis* *4 ARVD or ARVD-like* *3 Fibrosis + hypertrophy* *1 Fibrosis, fatty infiltration* *1 WPW syndrome*
[40] *Lesauskaite, Am J Forensic Med Pathol 1998*	*Case report*	*2 cases*	*2 male 20 and 22 years*	*Yes*	*Yes*	*Yes*	*2 Hypoplastic CAD myocardial hypoperfusion*
[42] Link, Chest 1998	Case report	1	1 m	No	Yes after event	No (aborted SCD)	1 commotio cordis
[44] Maron, JACC 2003	18 Competitive sports 192 cases basketball or football	286	256 m/30 f	Yes in 55	Yes in 252	286	102 HCM 37 AOCA 29 Indeterminant, possibly HCM 20 Myocarditis 12 Ruptured aortic aneurysm 11 ARVC 11 Tunnelled coronary artery 10 Aortic valve stenosis 10 Atherosclerotic CAD 9 Idiopathic DCM 9 MVP 8 Coronary artery hypoplasia 8 Other congenital anomalies 3 Cardiac sarcoidosis

							3 LQTS 3 Congenital heart disease 1 Myocardial infarction
[8] Maron, JACC 1998	Retrospective	3	3 m	No	Yes	3	1 AOCA 1 Congenital aortic valve stenosis (bicuspid) 1 Myocarditis
[47] Maron, JAMA 2002	Retrospective	128 cases ↓ 107 fatal	122 m/6 f	?	?	Yes in 82	79 Commotio cordis
[49] Maron, NEJM 1995	Retrospective	25	24 m/1 f	?	?	22	16 Commotio cordis
[12] Maron, JACC 1996	Retrospective	2	1 m/1 f	No	Yes	2	1 Anomalous LMCA 1 Atherosclerotic CAD
[2] Maron, Circulation 1980	Prospective and retrospective	29	26 m/3 f	Yes in 8	Yes in 7	29	14 HCM
							4 AOCA 5 Idiopathic concentric LV hypertrophy 3 Coronary heart disease 2 Ruptured aorta 1 Unknown
[3] Maron, JAMA 1996	Retrospective	134 SCD (+7 commotio cordis)	120 m/14 f	Yes in 24	Yes in 115	Yes	48 HCM 14 Unexplained increase in cardiac mass (HCM?) 17 Aberrant coronary arteries 8 Other coronary anomalies 6 Ruptured aortic aneurysm 6 Tunnelled LADCA 5 Aortic valve stenosis 4 Lesion consistent with myocarditis 4 Idiopathic dilated cardiomyopathy 4 ARVD 4 Idiopathic myocardial scarring 3 MVP 3 Atherosclerotic CAD 2 Other congenital heart syndrome 1 Long QT syndrome 1 Sarcoidosis 1 Sickle cell trait 3 'Normal' heart
[54] Menke, Chest 1985	Case report	1	1 m	No	Yes	1	1 Hyoplastic coronary arteries and high takeoff position of the right coronary ostium
[56] Morentin, Arch Dis Child 2000	Population-based observational study	10	9 m/1 f	Yes in some	Yes in some	10	3 Arrhythmogenic cardiomyopathy
							2 HCM 1 DCM 1 Myocarditis 1 AOCA 1 Tetralogy of Fallot 1 WPW syndrome
[58] Nilsson, Lancet 1999	Case studies	2	2 m	Yes	Yes	2	2 Myocarditis
[9] Phillips, JAMA 1986	Retrospective	19	19 m	?	?	19	8 Myocarditis 3 Coronary anomalies 2 HCM 1 Floppy mitral valve 1 Shone's syndrome (parachute mitral valve and subaortic stenosis) + sickle cell trait 1 Focal subendocardial fibrosis and calcification with normal coronary arteries 3 No aetiological DX

Table 1 (*continued*)

Authors/journal	Type of article/study	No. of cases of SCD	Sex (m, male; f, female)	Previous symptoms	Clinical data	Autopsy	Cardiovascular diagnosis
[15] Quigley, JAMA 1986	Retrospective study	51 ↓ 11< 35 y	50 m/1 f	Yes in some	?	11	3 Atherosclerotic CAD 2 Aortic stenosis 1 HCM 1 Congenital coronary artery anomaly 1 Cardiac arrhythmia 1 Multiple areas of fibrosis on myocardium 2 Non-cardiac
[61] Rozin, Am J Forens Med Pathol 2003	Retrospective review	1	1 m	No	?	1	1 Kawasaki disease
[62] Schiønning, Am J Forens Med Pathol 1997	Retrospective	3	2 m/1 f	Yes in one	Yes in 2	3	3 ARVD
[63] Scoville, Am J Prev Med 2004	Population-based review	59	?	?	?	59	27 Coronary artery abnormality 12 Myocarditis 5 Cardiomyopathy 6 Atherosclerotic cardiovascular disease 3 Conduction system abnormality 3 Cardiac valvular disease 2 Myocardial fibrosis 1 Ephedrine-induced arrhythmia
[64] Tabib, Circulation 2003	Retrospective	7	3 m/4 f	?	?	7	7 ARVD
[66] Tabib, Eur Heart J 1999	Retrospective	80	77 m/3 f	No	?	80	27 Atherosclerotic CAD (1<30 y) 19 HCM 9 DCM 8 ARVD 5 His bundle structural anomalies 2 Scarred myocardial bruising 2 Muscular bridging in LADCA 2 Congenital aortic bicuspid stenosis 1 Aneurysm (Kawasaki disease) 1 Asymmetric HCM 1 Tawarien mesothelioma 1 Marfan disease 1 MVP+IAC+fibrosis of His bundle trunk
[55] Taylor, J Am Coll Cardiol 1992	Retrospective	242 autopsies ↓ 34 cardiac and exercise-related	201 m/41 f	Yes in some	?	34	18 LMCA and RCA from R Ao sinus 6 RCA and LMCA from L Ao sinus 3 Hypoplastic CA 2 LMCA or LAD from pulmonary trunk 2 RCA and/or LMCA from posterior Ao sinus 2 Single RCA ostium from aorta 1 Single LCA ostium from aorta
[69] Tecce, Catheterization and Cardiovasc Diagnosis 1994	Case report	1	1 m	No	?	No (aborted SCD)	1 Spasm of left anterior descending artery
[71] Thakore, J Accid Emerg Med 2000	Case reports	2	2 m	No	?	2	1 Commotio cordis
[73] Trusty, AACN Clinical issues 2004	Case report	1	1 m	No	Yes	No (aborted SCD)	1 HCM
[75] Warren, West J Med 1979	Case report	1	1 f	Yes	Yes	1	1 Familial cardiomyopathy (HCM?)

[77] Wesslen, Eur Heart J 1996	Retrospective for 14/16 cases	16	15 m/1 f	Yes in 5	Yes in 16	16	5 Active myocarditis 4 ARVD-like alterations 1 HCM + healed myocarditis 1 Fibrosis, fatty infiltration + hypertrophy 1 Anatomical basis for pre-excitation 1 Early healing myocarditis + HCM? 1 Earlier myocarditis? 1? (Autopsy 6 days after death)
[17] Yanai, J Clin Forens Med 2000	Retrospective	36	35 m/1 f	?	Yes in 19	36	20 Atherosclerotic cardiovascular disease (4 < 35 y) 7 Cardiomyopathy (5 < 35 y) 4 Myocarditis (< 35 y) 1 Marfan syndrome (< 35 y) 1 MVP (< 35 y) 1 Fibrosis of bundle of His (< 35 y) 1 Commotio cordis (< 35 y) 1 Undetermined (< 35 y)
[80] Young, MJA 1999	Retrospective case series	8	8 m	Yes in 2	?	8	8 Ischaemic heart disease (CAD)

Ao, Aorta; AOCA, anomalous origin of coronary artery; ARVD, arrhythmogenic right ventricular dysplasia; AV, atrioventricular; CA, coronary artery; CAD, coronary artery disease; DCM, dilated cardiomyopathy; DX, diagnosis; HCM, hypertrophic cardiomyopathy; IAC, intra-atrial communication; LADCA, left anterior descending coronary artery; LMCA, left main coronary artery; LQTS, long QT syndrome; LV, left ventricle; MVP, mitral valve prolapse; RCA, right coronary artery; RV, right ventricle; WPW, Wolff–Parkinson–White; Y, years old; In italics, articles about cases already reported (the most recent or complete was considered). Underlined, cases of commotio cordis.

(classified in alphabetical order according to the last name of the first author), the type of article, the number of reported cases, the age and sex of the cases, the activity when SCD occurred, the eventual presence of previous symptoms and clinical data, whether an autopsy was carried out, and finally the cardiovascular diagnosis.

The articles were either case reports, relating the death of one or several athletes, or larger retrospective, sometimes prospective, studies. They were published between 1970 and 2004. The total of reported cases of SCD in athletes is 1101 cases (articles reporting the same cases are excluded, and appear in italics). This total includes the 103 cases of commotio cordis (blunt trauma to the chest, not particularly violent, that causes arrhythmias potentially followed by death) found in the articles (underlined).

The 1101 cases were grouped after pathology, according to the cardiovascular diagnosis, into 10 groups and 31 subgroups, as shown in Table 4. Coronary artery anomalies, half of them being anomalies of the origin of the coronary artery, the other half anomalies of the course of the coronary artery, and hypertrophic cardiomyopathy (HCM) are the two largest groups, with 262 and 261 cases, respectively. Traumatic cases of SCD being excluded (103 cases), there are 998 remaining cases, as shown in Fig. 1.

Focusing on the youngest athletes, 80 out of 199 were under 18 years of age, which represents as much as 40% of the cases, and 67 were under 16 years, 20% of the cases, with all types of underlying cardiac pathologies.

The cases were also grouped according to the sports the athletes were engaged in when sudden death occurred (Fig. 2). In 388 cases the sport practised when SCD occurred was mentioned. Deaths by commotio cordis (103) are not included in this figure.

Preparticipation screening protocols

A total of 108 articles were considered, and 31 articles were examined more closely, separated into three groups. (i) Articles describing the PPSP procedure (cardiovascular part) and using it (± reported cases): 11 articles. The articles were classified in Table 2, by alphabetical order of the first author. We looked at the population screened, where the PPSP was produced and when, who was performing the PPSP, at what frequency, what was the content of the PPSP (medical history, physical examination, non-invasive tests), the number of athletes screened, the number of athletes disqualified by the PPSP and the number of SCDs. (ii) Articles defining the PPSP procedure (cardiovascular part) but not using it; no reported cases: 11 articles. The articles were classified in Table 3, by alphabetical order of the first author. We

Table 2 Articles describing the preparticipation screening protocols procedure (PSPP) and using it

Authors	Population screened	Origin of protocol	Persons performing PPSP	Frequency of screening	Content	No. of athletes screened	No. of athletes disqualified	No. of SCD
[68] Bader, 2004	School grades 7–12	USA 2001	Healthcare professionals	Every 2 years	MH PhE NITa	–	–	–
[81] Brukner, 2004	Athletes <35 years	Australia	Doctors	On admission + regular basis	MH PhE NIT[a]	–	–	–
[82] Cantwell, 1998	Athletes	USA	Doctor	?	MH PhE NIT[a]	–	–	–
[30] Corrado, 1998	Athletes <35 years (prospective study)	Italy	?	?	MH PhE NIT	33 735 3016 Echo	1058: CV cause: 621 1 SD	269 SD, 49 in compet. athletes
[70] Fuller, 1997	High school: 13–19 years (prospective study)	USA	MH, BP, ECG by cardiac technician Cardiologists	?	MH PhE NIT	5615 582 Echo	22	1 Aborted SCD
[72] Glover, 1998	High school, grades 9–12	USA	21/51 Others than physicians OK	?	MH PhE	–	–	–
[74] Koester, 2003	High school	USA	72/154 OK by nurse practitioner 39/154 OK by physician assistant 22/154 OK by chiropractors 12/154 OK by naturopathic clinicians	?	MH PhE	–	–	–
[83] Maron, 1987	College: 17–30 years (prospective screening)	USA	3 Clinicians member of Health Center staff	Annual	MH PhE NIT	501 Athletes 90 Echo	0	–
[76] Pfister, 2000	College	USA	Team physician 603/713 (451 orthoped surg, 149 internal med, 32 pediatr) 135 OK by nurse practitioners 244/713 OK by athletic trainers	Annual in 446/879 On college entry in 433/879	MH PhE NIT in 58	–	–	–
[78] Smith, 1998	High school	USA	Physicians and residents + therapists, dietitians, secretaries, nurse counsellor in sports psychology Final decision by physician	Every 3 years PhE	MH +17 cases with CV anomalies where follow-up was recommended	2739 Athletes	53 (10 for cardiac reasons)	–
UCI, 2003	Cyclists	Switzerland	Team doctor	6 Months–2 years + in case of symptoms	MH PhE NIT (echo and stress ECG every other year)	14 Cyclists with life-threatening CV problems (21–35 years)	14	1 SCD (ARVD) before disqual. 7 SCD after disqual.

ARVD, Arrhythmogenic right ventricular dysplasia; BP, blood pressure; CV, cardiovascular; ECG, electrocardiogram; MH, medical history; NIT, non-invasive tests; PhE, physical examination; SCD, sudden cardiac death; SD, sudden death. [a]Not routine.

Table 3 Articles describing a preparticipation screening protocols procedure, without using it

Authors	Origin of protocol, year when introduced	Persons performing PPSP	Frequency of screening	Content
[79] Armsey, 2004	Athletes high school and college (USA)	Sports medicine team (PPSP stations) Athletic training staff Team orthopaedists Team medical staff (medical exam, orthopaedic assessment and checkout by physician)	Every 3–4 years (some places every year): full PPSP screening Yearly: medical history	MH PhE Further testing not discussed
[84] Beckermann, 2004	Athletes high school and college Stanford CA (USA)	?	AHA 1996 recommends: every 2 years for young athletes + annual BP for college athletes	MH PhE NIT
[85] Drezner, 2000	Athletes high school and college (USA)	?	AHA 1996 recommends: for high school athletes every 2 years + interim history in intervening years. For college athletes, history and BP every year	MH PhE NIT[a]
[86] Glorioso, 2002	AHA recommendations + Marfan screening	?	?	MH PhE NIT
[87] Gomez, 1999	Survey of 500 US high schools → 254 responses. Recommendations from the American Academy of Pediatrics.	?	?	MH Further testing not discussed
[88] Kurowski, 2000	Preparticipation athletic evaluation, Illinois (USA)	?	?	MH PhE NIT[a]
[89] Lyznicki, 2000	CV screening of student athletes. AHA recommendations (18) PPSP Task Force (18)	Trained healthcare worker, preferably a physician (AMA recommends only licensed physicians)	For high school athletes, every 2 years with an interim history in intervening years	MH PhE NIT[a]
[90] Maron, 1996 (+1998 addendum)	AHA scientific statement (USA) 1996. High school and collegiate athletes	Healthcare worker with requisite training. Preferably a licensed physician	Before participation then every 2 years. Interim history in intervening years + BP	MH PhE NIT[a]
[91] Metzl, 2001	PPSP of the adolescent (USA) (NB: not focused on CV system)	Physician	?	MH PhE Refer to cardiologist for further investigations
[92] O'Connor, 1998	AHA recommendations (American Academy of Family Physicians)	?	Before participation then every 2 years. Interim history in intervening years	MH PhE Refer to cardiologist for further investigations
[93] Soni, 1997	Children	Physician	To be reviewed on an ongoing basis	MH PhE NIT ([a]by specialist)

AHA, American Heart Association; AMA, American Medical Association; BP, blood pressure; MH, medical history; NIT, non-invasive tests; PPSP, preparticipation screening protocol; PhE, physical examination. [a]Not routine.

looked at the origin of the protocol, the persons performing the PPSP, the frequency of screening and the content of the protocol. (iii) Articles about PPSP, specifically ECG or echocardiography: nine articles [41,43,45,46,48,50–53]. We focused on the utility of the test for diagnosing underlying cardiovascular diseases, the indications for using the test, the cost-effectiveness, and the opinion of the author on the test.

Discussion

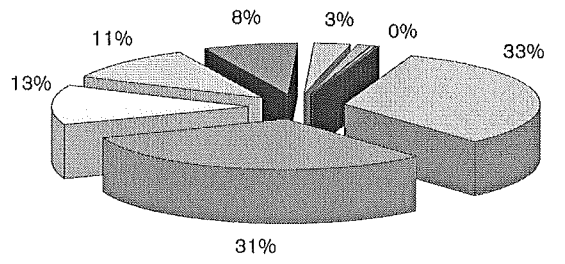
Physical activity is promoted and encouraged in society. It is considered healthy, with positive effects on the body and the mind. Healthcare and educational systems incite the population to be active in sports. The increased risk of exercise-related sudden death [10,55,57] might pose an irrelevant question mark on this statement, which is not justified because the positive effects of regular

Table 4 Causes of sudden cardiac death

Pathology groups	No. of cases	Pathology subgroups	No. of cases
Congenital	326	Coronary artery anomalies:	262
		Origin (L>R)	125
		Course (L>R)	124
		Spasm (LCA)	1
		Hypoplasia	12
		Cardiac valve diseases: 50	
		Mitral valve prolapse	27
		Aortic valve stenosis	20
		Unspecified	3
		Shone's syndrome	1
Cardiomyopathies	309	Hypertrophic cardiomyopathy	261
		Dilated cardiomyopathy	24
		Unspecified	12
		Myocardial scarring	6
		Sickle cell trait	1
		Fibrosis + fatty infiltration	5
		Unspecified	13
Arrhythmia	126	Arrhythmogenic right ventricular cardiopathy/dysplasia	97
		Long QT syndrome	5
		Conduction system pathology	20
		Wolf–Parkinson–White syndrome	1
		Anatomical basis for preexcitation	1
		Ephedrine-induced arrhythmia	1
		Unspecified	1
Atherosclerotic	112	Atherosclerotic coronary artery disease	112
Trauma	103	Commotio cordis	103
Infectious	78	Myocarditis	78
Degenerative	31	Cardiac sarcoidosis	4
		Marfan's syndrome	4
		Ruptured aorta (aneurysm)	23
Undetermined	10		10
Acquired	3	Kawasaki	3
'Normal heart'	3		3
Total	1101		1101

L, Left; LCA, left coronary artery; R, right.

Fig. 1



Causes of sudden cardiac death. ▨, Congenital anatomical; ▩, cardiomyopathies; □, arrhythmias; ▤, atherosclerotic; ▩, infectious; ▨, degenerative; ▩, undetermined; □, acquired; ▩, 'normal heart'.

physical activity far outweigh the negative effects. Exercise-related sudden death can have many causes, such as cardiac, asthma or other pulmonary conditions, heat stroke (hyperthermia and dehydration), cerebral embolism or ruptured cerebral artery, trauma of the spine or head, drug abuse and doping. Doping is often considered to be the main cause of sudden death by the media and lay people [59], which seems unlikely, as underlying cardiac diseases account for approximately 90% of exercise-related sudden deaths [5–7].

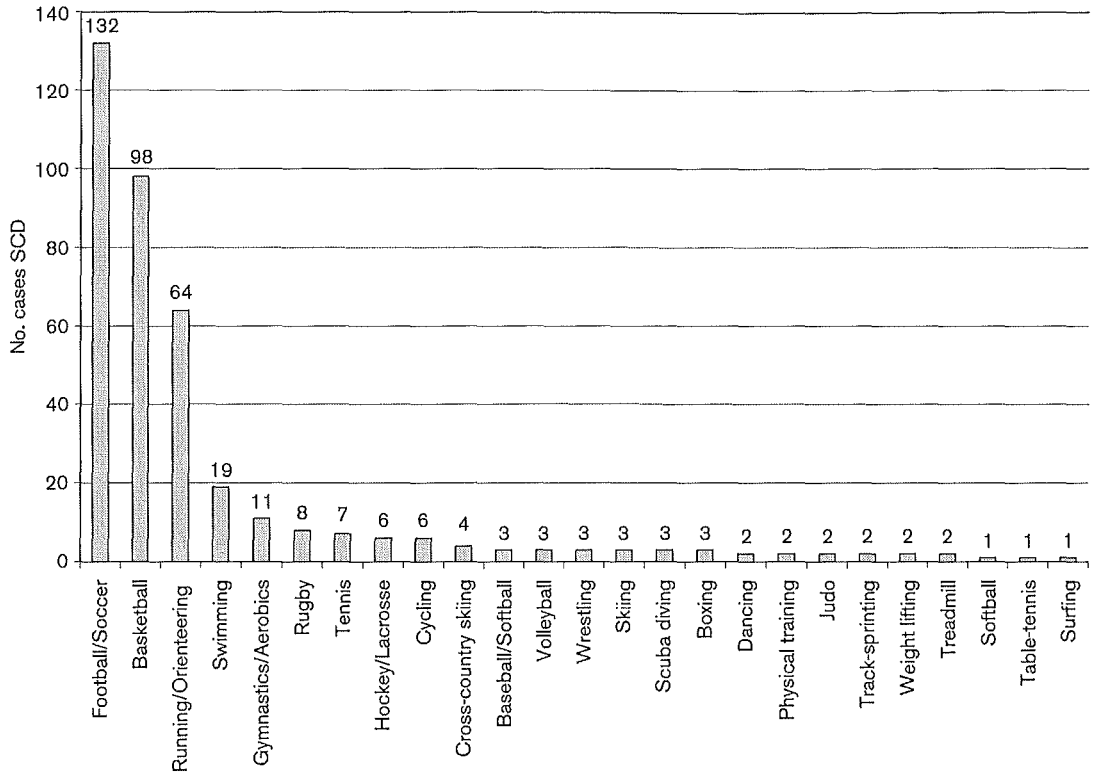
The incidence of SCD in young athletes is 0.5–2/100 000 per year [6,8–10]. Although the underlying forms of cardiac pathology are infrequent, they are represented in the athletic population as they are in the general population. The intense physical training and competition, with the accompanying higher cardiovascular demands, enhances the risk of athletes suffering serious consequences from their underlying cardiovascular disease [2,3]. The identification of the pre-existing pathology suggests that sport itself is not *per se* the cause of the increased mortality; it rather acts as a trigger upon underlying cardiovascular diseases, predisposing to life-threatening ventricular arrhythmias during physical exercise.

The unfortunate absence of national or international registers, which could provide precise data, suggests that SCD, although the present numbers are already impressive, is probably under-reported. The absence of precise data makes our/any analyses of reasons for SCD fragile and probably biased. For example, coronary artery diseases and HCM are the leading causes of SCD but are also the easiest to find; more difficult to find causes such as arrhythmias and arrhythmogenic right ventricular dysplasia may be under-represented. Cardiac causes of sudden death in the cases reported are listed in Table 4. Congenital coronary anomalies, such as abnormal origin and abnormal course of the coronary arteries and HCM are the leading causes of SCD [3,4,6,7,17–19,60]. The reported leading causes of SCD, however, vary greatly from author to author. Corrado *et al.* [11] reported coronary artery anomalies (atherosclerotic and non-atherosclerotic) as by far the leading group with 30% of cases, whereas HCM accounts for only 7% [27,29,30]. Hoogsteen *et al.* [34] reported 38% of cases caused by HCM, with 26% of coronary artery anomalies.

The low proportion of SCD with a normal heart at pathological examination is another point of interest, which probably has to do with the interest of the pathologist or cardiologist to report on clearcut pathology rather than normal conditions, together with the fact that in our study we excluded all potential doping-related events.

A remarkable point is that atherosclerotic coronary artery disease already plays an important role in SCD in the young athlete; this premature form of atherosclerosis accounts for 2–20% of the cases of SCD depending on the study, and has long been overlooked.

Fig. 2



Number of cases of sudden cardiac death (SCD) by sport.

Table 5 Classification of sports

Examples	Low isotonic	Moderate isotonic	High isotonic
Low isometric	Golf	Volley-ball	Soccer
Moderate isometric	Equestrian	Running sprint	Basketball
High isometric	Gymnastics	Downhill skiing	Cycling

Congenital anatomical heart diseases, cardiomyopathies and atherosclerosis represent 75% of the cases of SCD reported, if traumatic causes (commotio cordis) are excluded (Fig. 1). The various percentages of cardiopathic causes can be explained by the different incidences of the cardiopathies, HCM having an incidence of only 1/500 [4,6,65,67], whereas anatomical anomalies of the coronary arteries have an incidence of 0.3–1/100 [5,55].

A strikingly high percentage of very young athletes suffer from SCD. We found that approximately 40% of the total of SCD, in which the age was specified, occurred in athletes under 18 years of age, 80 of 199 athletes (out of the total 998 cases), 67 of these were even younger than 16 years. In the remaining 799 (998–80) the age was not specified.

To assess the risk of SCD, sports can be classified depending on the dynamic (isotonic) and static (isometric) work (Table 5). We grouped the reported cases by sport in Fig. 2, which shows that football/soccer, basketball and running/orienteering are the largest but not necessarily the most dangerous groups. They probably encompass the largest population of sports participants, because of their popularity. This does not mean that although these sports provide the largest number of SCD they necessarily are the sports with

Table 6 Lausanne Recommendations: sudden cardiovascular death in sport. Preparticipation cardiovascular screening. Under the umbrella of the International Olympic Committee Medical Commission, 10 December 2004

Step 1: For all participants	Potentially detectable cardiovascular conditions
Personal history: Questionnaire by examining physician	Any cardiovascular condition
Have you ever fainted or passed out when exercising?	
Do you ever have chest tightness?	
Does running ever cause chest tightness?	
Have you ever had chest tightness, cough, wheezing, which made it difficult for you to perform in sports?	
Have you ever been treated/hospitalized for asthma?	
Have you ever had a seizure?	
Have you ever been told that you have epilepsy?	
Have you ever been told to give up sports because of health problems?	
Have you ever been told you have high blood pressure?	
Have you ever been told you have high cholesterol?	
Do you have trouble breathing or do you cough during or after activity?	
Have you ever been dizzy during or after exercise?	
Have you ever had chest pain during or after exercise?	
Do you have or have you ever had racing of your heart or skipped heartbeats?	
Do you get tired more quickly than your friends do during exercise?	
Have you ever been told you have a heart murmur?	
Have you ever been told you have a heart arrhythmia?	
Do you have any other history of heart problems?	
Have you had a severe viral infection (for example myocarditis or mononucleosis) within the past month?	
Have you ever been told you had rheumatic fever?	
Do you have any allergies?	
Are you taking any medications at the present time?	
Have you routinely taken any medication in the past two years?	
Family history: Questionnaire by examining physician	
Has anyone in your family less than 50 years old:	Inherited cardiomyopathy (hypertrophic, arrhythmogenic RV, dilated)
Died suddenly and unexpectedly?	
Been treated for recurrent fainting?	
Had unexplained seizure problems?	Inherited heart rhythm problem/cardiac ion channel diseases (long and short QT syndrome, Brugada syndrome, Lenègre disease, catecholaminergic polymorphic VT)
Had unexplained drowning when swimming?	
Had unexplained car accident?	
Had heart transplantation?	
Had a pacemaker or defibrillator implanted?	
Been treated for irregular heart beat?	
Had heart surgery?	
Has anyone in your family experienced sudden infant death (cot death)?	
Has anyone in your family been told they have Marfan syndrome?	Connective tissue disorders
Physical examination:	
General:	Coarctation of the aorta
Peripheral pulses	Aortic dilatation, mitral valve prolapse
Marfan stigmata	
Cardiac auscultation:	Ventricular ectopic beats
Rate/rhythm	Structural heart disease, outflow tract obstruction
Murmur: systolic/diastolic	Aortic valve disease
Systolic click	Hypertension
Blood pressure	Abnormal heart rate for age (unexplained sinus tachycardia or bradycardia)
Diagnostic tests:	Abnormal heart rhythm
12-lead rest ECG:	PR/QRS: WPW
(after the onset of puberty)	QTc/T wave: long or short QT syndrome
	T wave inversion/flattening: myopathy
Step 2: Selected cases with positive personal history, family history of potentially inherited cardiac disease, or positive physical or ECG findings in Step 1 require further evaluation by an age-appropriate cardiac specialist to qualify the athlete for sports participation	
Further evaluation may include transthoracic echocardiography, maximal exercise testing, and 24-h ECG monitoring. In addition, non-invasive screening of family members may provide valuable information about inherited cardiovascular disease	

ECG, electrocardiogram; IOC, International Olympic Committee; RV, right ventricle; VT, ventricular tachycardia; WPW, Wolff–Parkinson–White syndrome.

the highest risks. The sports with the highest cardiovascular demands and the highest isotonic work remain therefore to be more at risk of SCD, but because of a smaller number of participants have a smaller absolute number of SCD. This does not mean that the mentioned examples of sports *per se* represent a risk of SCD.

After reviewing the causes of SCD and the sports with the highest risk of a cardiac event, the PPSP was

considered. The studies reviewed reveal that the protocols are often considered as recommendations, and are only partly used. Some are considered inadequate for cardiovascular evaluation and contain four or fewer items recommended by the American Heart Association. The existing protocols all include a medical history and a physical examination, in some cases diagnostic tests, of varying content. The person performing the screening is also variable, some places allowing non-physicians [19,68,70,72,74,76,78,79,89,90] to conduct the examination, which decreases the chances of positive findings. One wonders what the results will be when technicians, nurses, chiropractors, or athletic trainers perform the cardiovascular screening.

Even if a general consensus concerning the need for screening did exist, cost-effectiveness and the fact that it is not possible to prevent all deaths constitute a major problem. The Italian screening programme, consisting of history, physical examination and ECG seems to be cost-effective and efficient. Abnormal ECG lead to further investigations (often echocardiography) to detect cardiovascular diseases at risk of sudden death in athletes. As only 9% are false positives, the low cost of the ECG justifies this small percentage of unnecessary echocardiographies. As sport is a voluntary activity, death should not occur or at least be prevented as much as possible. Knowing the amount of money spent on top sports, the budget should also allow for the most efficient screening, to be performed by professionals.

The articles studied in the PPSP parts (i) and (ii) show a striking lack of uniformity, considering both quantity, the number of questions and tests, and quality. Although the protocols studied are aimed at detecting underlying cardiovascular diseases, in some the application of the recommendations is so limited that the usefulness of such screenings is questionable. The quality of the screening will not change its cost, as far as medical history and physical examination are concerned.

When it comes to the diagnostic tests, there are varying approaches, depending mostly on the population screened. Mass screenings in high schools and colleges most often do not comprise routine diagnostic tests, but in some places an ECG is included [30,70,83,84,93]. Suspect cases are referred to specialists for further investigations. In screening protocols for elite athletes, ECG and echocardiography at least are often routine [94]. This is the case for example in the programme of obligatory examinations of the International Cycling Union.

ECG and echocardiography each have their strong and weak points, and uniformity is not reached here either. ECG can often be abnormal in well-trained athletes [4,41,95–97], with patterns resembling those found in cardiovascular diseases such as HCM, which is the origin of many false positives. Despite this disadvantage, it is still considered a valuable screening tool because of its high negative predictive value and its low cost. Echocardiography on the contrary is expensive, but can reveal the pathologies most frequently encountered in SCD.

Our meta-analysis aims to provide a well furnished literature basis and form a platform for a generally accepted PPSP. The cooperation with the IOC and the consensus meeting on SCD in athletes organized by the Medical Commission of the IOC provided a tool to come to a generally acceptable screening procedure adopted by the IOC as the Lausanne Recommendations.

The Lausanne Recommendations (Table 6) are aimed at preventing SCD. We have assembled elements from the various existing protocols and studies that will help us best detect underlying cardiovascualr diseases that could cause SCD in athletes. It is a stepwise approach.

Step 1 is the basic screening that should be offered to every athlete. It is focused on detecting underlying cardiovascular diseases and includes a detailed personal history, a family history, a physical examination and a 12-lead ECG. The questionnaire consists of preconceived closed questions, in which a 'yes–no' answer has to be given. Any 'yes' requires further enquiry. The family history is focused on cardiovascular diseases, either congenital or acquired, and premature sudden death. It is important that the physician performing the screening is trained to focus on the possible cardiac problems of intense sport, and that he/she can read the 12-lead ECG.

Step 2 follows step 1 in case of positive findings in the medical history (personal or family history) or in the diagnostic tests of the first step. These further evaluations and investigations are the concern of a cardiologist. The tests in step 2 are chosen according to the type of positive finding.

These Recommendations should be offered to the athletes with an explanation of the tests performed and the results that can be expected, and it should be clear that it is a voluntary participation for the athlete. Therefore his/her participation in the screening should be preceded by the signing of a form stating the written and informed consent and his/her right not to know should be respected in the case of refusal. Most recently, shortly after the acceptance of the Lausanne Recommendations, both the European Society of Cardiology [98] and the American Heart Association [99,100] published their recommendations. The European Society of Cardiology report recommendations coincide with the Lausanne recommendations (including the use of

a 12-lead ECG), whereas the Bethesda Conference discussed the use of an ECG, but felt it was currently inappropriate to advise for use in the United States.

Limitations of the study

Several limitations in this study need to be specified. The definition of an athlete is very vague, without any quantitative values. Competition does not either have a quantitative definition.

The most important limitation however is that SCD in young athletes as reported in the published and studied papers is certainly underestimated. Most of the events occur in youth potentially involved in sports activities, which are not reported in the literature. SCD is therefore likely to have comprised more than the 1101 athletes in the 38-year period across the world, which we reported. The lack of national or international registers and the uncertainty of the number of athletes involved forms the basis of this problem. An effort to inventory all cases of SCD in the future seems mandatory.

A bias resides in the fact that some articles do not give the precise age of every case reported, but sometimes only an age range for the whole group considered.

Certain sports are under-reported in the articles studied, such as cycling and rowing/canoeing. There are newer articles on cycling [101], which show significant numbers as reported to the smaller groups concerned. The International Cycling Union Cardiology Subcommission reports 14 competitive cyclists with major life-threatening cardiovascular problems. The athletes had trained and participated in competition for at least 5 years. They all seem to have developed an acquired form of right ventricular electrical instability.

Despite these limitations, this article tries to report facts and draw conclusions and cannot provide a final answer, but may provide a uniform format to help diminish this mortality. Further studies of efficacy are needed.

Conclusion

Sudden death in athletes is a serious problem that requires serious attention. The vast majority of these sudden deaths are caused by underlying cardiovascular diseases. Therefore, the general idea that most sudden deaths in sports are related to doping is contradicted by the large numbers of sudden deaths related to underlying cardiovascular diseases.

The importance of the group of athletes suffering SCD under 18 or even 16 years of age is to be noted, and a serious screening should be offered to teenagers training and competing at a high level.

Premature atherosclerosis in young athletes leads to death in a surprisingly large group of competitive athletes, when it was long thought to be a cause of death almost exclusively concerning older athletes.

The large variety of PPSP and the variability of persons using them prevent a serious evaluation of their efficacy. This evaluation would benefit from unified preparticipation recommendations, for which we propose the Lausanne Recommendations.

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Appendix

Members of the IOC ad hoc working group on Sudden Cardiac Death in Athletes:

Dr P. Schamasch, IOC Medical and Scientific Director, Lausanne, Switzerland

Dr. E.J. Meijboom, Professor of Congenital Cardiology, CHUV, Lausanne, Switzerland

K. Bille, Resident Pediatrics, Thesis on SCD in Athletes, CHUV, Lausanne, Switzerland

Dr J. Brenner, Professor of Pediatric Cardiology, Johns Hopkins, Baltimore, USA

Dr D. Corrado, Director of Cardiology Laboratory, Padua, Italy

Dr J. Dvorak, FIFA Chief Medical Officer, Zurich, Switzerland

D. Figueiras, Department of Sports Sciences, Lausanne, Switzerland

S. Greinig, IOC Manager of Medical Program, Lausanne, Switzerland

Dr J. Hoogsteen, Head of Training Centre Sports Cardiology, Veldhoven, the Netherlands

Dr L. Kappenberger, Professor of Cardiology, Head of Department, Lausanne, Switzerland

Dr W. McKenna, Professor, Director of Heart Hospital, London, UK

Dr F.J. Meijboom, Cardiologist, Thoraxcentre, Rotterdam, the Netherlands

D. Oswald, Lawyer, Director of International Centre for Sports Studies, Neuchâtel, Switzerland

Dr L. Rivier, Toxicologist, Lausanne, Switzerland

Dr P. Sangenis, Cardiologist, IOC MC Director Institute in Sports Medicine, Buenos Aires, Argentina

D. Sprumont, Lawyer, Director Institute of Health Law, Neuchâtel, Switzerland

Dr G. Theine, Professor of Pathology, Director Institute of Pathological Anatomy, Padua, Italy

Dr M. Zorzoli, International Cycling Union Health Manager, Aigle, Switzerland.