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J. Am. Coll. Cardiol. 2005;45;1318-1321
doi:10.1016/j.jacc.2005.02.006

This information is current as of March 27, 2009

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<http://content.onlinejacc.org/cgi/content/full/45/8/1318>



36TH BETHESDA CONFERENCE

Introduction: Eligibility Recommendations for Competitive Athletes With Cardiovascular Abnormalities—General Considerations

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The focus of this 36th Bethesda Conference is the trained athlete with an identified cardiovascular abnormality. The goal is to formally develop prudent consensus recommendations regarding the eligibility of such individuals for competition in organized sports, and to present these considerations in a readily useable format for clinicians. This document constitutes an update of the 16th (in 1985) (1) and more recent 26th (in 1994) (2) Bethesda Conferences. Once again, we have attempted to ascertain by the consensus of an expert panel which cardiovascular abnormalities (and with what degree of severity) place the competitive athlete at increased risk for sudden and unexpected death or disease progression so as to justify medical recommendations against participation in all or certain competitive sports, for the purpose of reducing that risk.

IMPETUS FOR THE REVISION

There are several reasons to offer this revision of the 26th Bethesda Conference (2). Over the past decade, substantial advances have taken place in the diagnosis and management of a variety of genetic and acquired cardiovascular diseases, in the recognition of the causes of athletic field deaths (3–5), and in ethical and legal issues that impact medical decision making (3–7). Also, sudden cardiac deaths in competitive athletes continue to be highly visible, compelling, and emotional events with significant liability considerations (3,6,7). These catastrophes are frequently subjected to intense public scrutiny largely because of their occurrence in young and otherwise healthy appearing individuals, including elite participants in collegiate and professional sports (3,6,7). Such sudden deaths are known to occur in athletes of both genders (although much more commonly in men by 9:1), minorities, at a wide range of ages, and in a broad spectrum of sports (most commonly basketball and football in the U.S. and soccer in Europe) (3–5). All of these considerations, as well as the legal decision in Knapp vs. Northwestern, which placed the Bethesda Conference recommendations for the eligibility of athletes with cardiovascular abnormalities as a precedent (6), have led to the necessity for this revised document.

DEFINITIONS

As before, we define the competitive athlete as one who participates in an organized team or individual sport that

requires regular competition against others as a central component, places a high premium on excellence and achievement, and requires some form of systematic (and usually intense) training. Therefore, organized competitive sports are regarded as a distinctive activity and lifestyle. An important component of a competitive sports activity concerns whether athletes are able to properly judge when it is prudent to terminate physical exertion. For example, the unique pressures of organized sports do not allow athletes to demonstrate strict control over their level of exertion or reliably discern when cardiac-related symptoms or warning signs arise. However, the panel recognizes that this definition is most easily applied to high school, college, and professional sports. Clinicians may want to use individual judgment in defining competitive forms of physical activity for participants in many youth sports activities, particularly those for children less than age 12 years.

Athletes may be regarded as competitive in many sporting disciplines—at almost any age or level of participation, including involvement in high school, college, professional, and master's sports (8). The recommendations in this document do not apply to (and are not specifically designed for) non-competitive recreational sports activities; such appropriate guidelines appear elsewhere (9). Nevertheless, we also recognize that some practitioners will choose to extrapolate or translate the recommendations for competitive athletes selectively to some recreational sports, and to non-athletes with occupations that require vigorous physical exertion (e.g., firefighters or emergency medical technicians), or to cardiac rehabilitation programs.

Furthermore, it is emphasized that these Bethesda Conference recommendations should not be regarded as an injunction against physical activity in general; indeed, the panel recognizes the well-documented health benefits of exercise. In particular, regular recreational physical activities should be encouraged. Excessive and unnecessary restrictions could potentially create physical and psychological burdens (particularly in young children).

PREVALENCE

The prevalence of cardiovascular disease in the young athletic population is low. Furthermore, although the precise risk of sudden cardiac death in athletes with underlying disease is unresolved, it is also undoubtedly low (3,10).

Indeed, the number of athletes who die of cardiovascular or related causes each year in the U.S. is probably less than 300, compared with the large number of athletes participating in a broad spectrum of organized sports (about 10 to 15 million) of all ages in the U.S.

If sudden death in athletes is a relatively uncommon event, why is it regarded as a substantive medical issue? This relates largely to the generally held perception that competitive athletes represent the healthiest and most dynamic members of society, in whom cardiovascular sudden deaths become symbolic and riveting and strike to the core of our sensibilities. Indeed, these events are counterintuitive, and the visibility of such catastrophes is often enhanced by their portrayal in the news media as public events rather than personal and family tragedies (3,6,7). For elite athletes who often achieve celebrity status, the economic stakes may be high, making medical decision making even more difficult (3,6,7). For all these reasons, the sudden deaths of athletes have had enormous impact on the public consciousness and attitudes of the medical profession.

CONFERENCE DESIGN AND FORMAT

A primary foundation for this 36th Bethesda Conference is the generally accepted tenet that young trained athletes with underlying cardiovascular abnormalities are at increased risk for sudden cardiac death (usually on the athletic field) (3-5), in comparison to non-athletes with cardiovascular disease (relative risk 2.5, in one study) (11), largely by virtue of exposure to the unique physiologic and psychologic stresses of intense training and competition. Indeed, physical activity during training or competition appears to trigger the vast majority of sudden deaths in athletes (3-5). That trained athletes may occasionally die suddenly unrelated to exercise or significant physical activity does not negate this principle.

Task Force reports 2 to 7 of this conference document offer specific recommendations for the eligibility and temporary or permanent disqualification of trained athletes with cardiovascular abnormalities and structural diseases previously implicated in sudden cardiac death (or disease progression). The most common of these conditions in young athletes (less than 35 years of age) are hypertrophic cardiomyopathy (HCM), congenital coronary artery anomalies of wrong sinus origin, myocarditis, Marfan syndrome (with aortic dissection), and arrhythmogenic right ventricular cardiomyopathy (predominantly in Italy) (3-5,11) (Table 1).

Although many of these diseases have been generally regarded as rare, it now appears that HCM is relatively common (1:500 in the general population) (12). In contrast, atherosclerotic coronary artery disease is by far the most common cause of sudden death in older athletes, and relatively uncommon in the young (3,13). We have arbitrarily excluded from consideration other particularly rare potential causes of sudden death, such as heat stroke, bronchial asthma, cerebral hemorrhage, and hematologic disorders.

Other sections of this document address related areas

Table 1. Causes of Sudden Death in 387 Young Athletes*

Cause	No. of Athletes	Percent
Hypertrophic cardiomyopathy	102	26.4
Commotio cordis	77	19.9
Coronary artery anomalies	53	13.7
Left ventricular hypertrophy of indeterminate causation†	29	7.5
Myocarditis	20	5.2
Ruptured aortic aneurysm (Marfan syndrome)	12	3.1
Arrhythmogenic right ventricular cardiomyopathy	11	2.8
Tunneled (bridged) coronary artery‡	11	2.8
Aortic valve stenosis	10	2.6
Atherosclerotic coronary artery disease	10	2.6
Dilated cardiomyopathy	9	2.3
Myxomatous mitral valve degeneration	9	2.3
Asthma (or other pulmonary condition)	8	2.1
Heat stroke	6	1.6
Drug abuse	4	1.0
Other cardiovascular cause	4	1.0
Long QT syndrome§	3	0.8
Cardiac sarcoidosis	3	0.8
Trauma causing structural cardiac injury	3	0.8
Ruptured cerebral artery	3	0.8

*Data are from the registry of the Minneapolis Heart Institute Foundation (3).
 †Findings at autopsy were suggestive of HCM but were insufficient to be diagnostic.
 ‡Tunneled coronary artery was deemed the cause of death in the absence of any other cardiac abnormality. §The long QT syndrome was documented on clinical evaluation.
 Source: Reproduced from Maron B.J. (3) with permission of the Massachusetts Medical Society.

such as preparticipation screening and diagnostic strategies, use of illicit drugs and dietary supplements, ethical and medical-legal considerations for sports disqualification, as well as sudden death due to blunt non-penetrating chest blows in the absence of heart disease (i.e., commotio cordis) (14,15).

RECOMMENDATIONS FOR DISQUALIFICATION AND ELIGIBILITY

Although recommendations of the 36th Bethesda Conference do not reflect policy of the American College of Cardiology Foundation, they are well considered views of a group of experts convened to address the medical risks imposed by competition on an athlete with a cardiovascular abnormality. The present consensus recommendations were necessarily based largely on individual and collective judgments and the experience of the panel, as well as on the available pertinent scientific data. Indeed, in the process of formulating the recommendations, the panel participants, who are cardiovascular authorities most comfortable with a high level of precision, were often required to confront areas in which there was a paucity of hard evidence and utilize the "art of medicine" in designing recommendations.

It is the premise of the expert panel that firm recommendations for temporary or permanent sports disqualification be confined to individual athletes with probable or conclusive evidence of disease rather than those with only border-

line findings or the presumption of a diagnosis. In this way, unnecessary restrictions from sports and the stigma of a cardiac diagnosis in healthy individuals may be minimized. We do recognize, however, that such an approach will inevitably permit an occasional athlete to participate who might otherwise be at some risk. Nevertheless, the level of importance the individual athlete personally attaches to continuing or resuming competitive sports is not regarded as a primary determinant in formulating eligibility recommendations.

The recognition by panel members that all competitive sports do not necessarily involve identical types or intensity of exercise is reflected in the Task Force 8: Classification of Sports. Training demands vary considerably even within the same sport and the intensity of conditioning regimens often exceed that of competition itself. However, it is often difficult to accurately grade such differences in exercise intensity owing to a variety of factors, including differing motivational attitudes and training demands. The demands of competitive sports may place athletes with certain cardiovascular abnormalities in extreme, unusual, and unpredictable environmental conditions (associated with alterations in blood volume, hydration, and electrolytes), over which they have limited control. These circumstances could enhance the risk for potentially lethal arrhythmias and sudden death and unavoidably distort the reliability of individualized and prospective risk stratification. Conversely, it is suspected that for many athletes the removal from their lifestyle of athletic training and competition will reduce risk for sudden death or disease progression.

The recommendations in this report should also be viewed in perspective. Appropriate sports disqualification is only one component for potentially reducing risk, and each relevant cardiovascular disease has its own treatment algorithms, which can include selective implantation of a cardioverter-defibrillator in high-risk patients (16).

The present recommendations formulated with respect to allowable levels of sports activity can be regarded as generally conservative. Certainly, this is a prudent posture to assume when the amount of available hard data and evidence is limited in many decision-making areas, as may be the case in portions of the 36th Bethesda Conference document. The panel acknowledges that while available data support the principle that competition in sports is associated with an increased *relative* risk for sudden death in the setting of known cardiovascular disease (11), the absolute risk cannot be determined with certainty in an individual patient/athlete, and in fact may be low in certain individuals. However, at present, additional risk-stratifying tools are not available to independently (and more precisely) guide many of these difficult medical decisions. Thus, it is possible that the recommendations of this consensus panel will occasionally cause some athletes to be withdrawn from competition unnecessarily. This is, of course, unfortunate because athletes derive considerable self-assurance, confidence, physical well-being, and, even on occasion, financial security from these activities. Nonetheless, the increased risk

of sudden death associated with intense athletic participation is a *controllable* risk factor, and the devastating impact of even infrequent sudden deaths in this young population underscores the wisdom of the conservative nature of these recommendations. Indeed, various cultures harbor differing societal views on the individual rights and prerogatives of athletes to persist in their chosen lifestyle, independent of the potential risks involved. In practice, consideration may be given in individual athletes to changing their competitive sport from a prohibited high-intensity activity to a permissible low-intensity one (i.e., usually to class IA). However, changing the position in which an athlete competes (e.g., from running-back to place-kicker in football) to accomplish the same end within high-intensity team sports may prove difficult in practical terms.

Consequently, the 36th Bethesda Conference report is presented here in the context of measured and prudent recommendations—intended neither to be overly permissive nor restrictive—and which should not be regarded as an absolutely rigid dictum. Indeed, the managing physician with particular knowledge regarding a given athlete's cardiovascular abnormality, psychological response to competition, and other medically relevant factors may choose to adopt somewhat different recommendations in selected individuals.

SPECIAL CONSIDERATIONS

Three considerations relevant to much of the document deserve special consideration. First, medications, such as beta-blockers commonly used to treat a variety of cardiac diseases including systemic hypertension, HCM, long QT syndrome, and Marfan syndrome, are likely to inhibit performance in trained competitive athletes (7). The use of such drugs in athletes cannot be regarded as either a means of affording safety and specific protection against arrhythmias, nor as a primary means for retaining eligibility in vigorous competitive sports. Furthermore, use of beta-blockers is specifically contraindicated in some sports.

Second, the availability of a free-standing automatic external defibrillator at a sporting event should not be considered either as absolute protection against a sudden death event, a prospectively designed treatment strategy for athletes with known cardiovascular disease, or justification for participation in competitive sports that otherwise would be restricted owing to underlying cardiac abnormalities and the risk of life-threatening ventricular tachyarrhythmias.

Third, with the increased employment of the implantable cardioverter-defibrillator (ICD) it is inevitable that increasing numbers of high-risk athletes with defibrillators will come to recognition. Although differences of opinion exist and little direct evidence is available, the panel asserts that the presence of an ICD (whether for primary or secondary prevention of sudden death) should disqualify athletes from most competitive sports (with the exception of low-intensity, class IA), including those that potentially involve

bodily trauma. The presence of an implantable device in high-risk patients with cardiovascular disease should not be regarded as protective therapy and therefore a justification for permitting participation in competitive sports that would otherwise be restricted. This conservative but prudent posture is justified on the basis of the uncertainties associated with ICDs during intense competitive sports, including the possibility that the device will not perform effectively at peak exercise, the likelihood of a sinus tachycardia-triggered inappropriate shock or an appropriate discharge, and the risk for physical injury to the athlete or other competitors as the result of an ICD shock. Also, pacemaker-dependent athletes should not participate in most competitive sports that potentially involve bodily trauma.

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Appendix 1. Author Relationships With Industry and Others

Name	Consultant	Research Grant	Scientific Advisory Board	Stock Holder	Expert Witness Testimony
Dr. Barry J. Maron	None	• Medtronic	None	None	• 1996, Defense, Knapp vs. Northwestern
Dr. Douglas P. Zipes	• Cardiofocus • Janssen • Medtronic	• Medtronic	• Medtronic	• MVMD	• 1996, Defense, Knapp vs. Northwestern

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