

36th Bethesda Conference

Eligibility Recommendations for Competitive Athletes With Cardiovascular Abnormalities

BETHESDA CONFERENCE REPORT

36th Bethesda Conference: Eligibility Recommendations for Competitive Athletes With Cardiovascular Abnormalities

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The recommendations set forth in this report are those of the Conference participants and do not necessarily reflect the official position of the American College of Cardiology Foundation.

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36TH BETHESDA CONFERENCE

Preamble

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This 36th Bethesda Conference report is the result of a consensus conference held on November 6, 2004, in New Orleans, Louisiana. Consensus conferences are designed to facilitate consideration of significant and timely issues regarding the practice of cardiovascular medicine and matters affecting patient care, research, and training for which absolute or hard data are incomplete. Appropriate organizations other than the American College of Cardiology Foundation (ACCF) are invited to send representatives to participate in the process, as appropriate to each topic. These may include physicians and health care specialists as well as representatives from industry, government, foundations, and the public sector. A report is published to highlight recommendations, along with dissenting opinion, if appropriate. The recommendations set forth in this report are those of the conference participants only and do not necessarily reflect the official position of the ACCF.

The Task Force on Clinical Expert Consensus recognizes the importance of maintaining high ethical standards and avoiding conflicts of interests. Because the development of this conference report depends on the knowledge and experience of its volunteers, many of whom have relationships with industry, the policy for writing committee members' relationships must be realistic, workable, and implemented in a way that protects the integrity of the process while allowing an open and honest exchange of information. Therefore, the Task Force facilitates a process for collection of disclosure statements of all relationships of conference participants that might be perceived as real or potential conflicts of interest relevant to the conference activity and publishes this information with the conference report. Please refer to the appendix of each Task Force report for author disclosure information.

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.

INTRODUCTION REFERENCES

1. Mitchell JH, Maron BJ, Epstein SE. 16th Bethesda Conference: cardiovascular abnormalities in the athlete: recommendations regarding eligibility for competition. *J Am Coll Cardiol* 1985;6:1186–232.
2. Maron BJ, Mitchell JH. 26th Bethesda Conference: recommendations for determining eligibility for competition in athletes with cardiovascular abnormalities. *J Am Coll Cardiol* 1994;24:845–99.
3. Maron BJ. Sudden death in young athletes. *N Engl J Med* 2003;349:1064–75.
4. Maron BJ, Shirani J, Poliac LC, Mathenge R, Roberts WC, Mueller FO. Sudden death in young competitive athletes: clinical, demographic, and pathological profiles. *JAMA* 1996;276:199–204.
5. Maron BJ, Carney KP, Lever HM, et al. Relationship of race to sudden cardiac death in competitive athletes with hypertrophic cardiomyopathy. *J Am Coll Cardiol* 2003;41:974–80.

6. Maron BJ, Mitten MJ, Quandt EF, Zipes DP. Competitive athletes with cardiovascular disease—the case of Nicholas Knapp. *N Engl J Med* 1998;339:1632–5.
7. Maron BJ. Sudden death in young athletes: lessons from the Hank Gathers affair. *N Engl J Med* 1993;329:55–7.
8. Maron BJ, Araújo CG, Thompson PD, et al. Recommendations for preparticipation screening and the assessment of cardiovascular disease in Masters athletes: an advisory for healthcare professionals from the working groups of the World Heart Federation, the International Federation of Sports Medicine, and the American Heart Association Committee on Exercise, Cardiac Rehabilitation, and Prevention. *Circulation* 2001;103:327–34.
9. Maron BJ, Chaitman BR, Ackerman MJ, et al. AHA scientific statement: recommendations for physical activity and recreational sports participation for young patients with genetic cardiovascular diseases. *Circulation* 2004;109:2807–16.
10. 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.
11. Corrado D, Basso C, Rizzoli G, Schiavon M, Thiene G. Does sports activity enhance the risk of sudden death in adolescents and young adults? *J Am Coll Cardiol* 2003;42:1959–63.
12. Maron BJ. Hypertrophic cardiomyopathy: an important global disease. *Am J Med* 2004;116:63–5.
13. Burke AP, Farb A, Virmani R, Goodin J, Smialek JE. Sports-related and non-sports-related sudden cardiac death in young adults. *Am Heart J* 1991;121:568–75.
14. Maron BJ, Gohman TE, Kyle SB, Estes NA III, Link MS. Clinical profile and spectrum of commotio cordis. *JAMA* 2002;287:1142–6.
15. Maron BJ, Poliac L, Kaplan JA, Mueller FO. Blunt impact to the chest leading to sudden death from cardiac arrest during sports activities. *N Engl J Med* 1995;333:337–42.
16. Maron BJ, Shen W-K, Link MS, et al. Efficacy of implantable cardioverter-defibrillators for the prevention of sudden death in patients with hypertrophic cardiomyopathy. *N Engl J Med* 2000;342:365–73.

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

TASK FORCES

Task Force 1: Preparticipation Screening and Diagnosis of Cardiovascular Disease in Athletes

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The present consensus panel recommendations of the 36th Bethesda Conference for eligibility and disqualification of competitive athletes are predicated on the prior diagnosis of cardiovascular abnormalities. However, the methodology by which these diseases are identified (including preparticipation screening) and how athletes come to evaluation for competitive eligibility, may involve several scenarios. First, athletes may be referred for assessment of clinical symptoms or signs. Second, fortuitous recognition may occur in routine clinical practice, triggered by findings on history and physical examination, such as a heart murmur. Third, young athletes may be suspected of having cardiovascular disease by virtue of formalized large population screening examinations that are customary before participation in competitive athletics (1).

PREPARTICIPATION SCREENING

Indeed, the ultimate objective of preparticipation screening carried out in general populations of trained athletes is the recognition of “silent” cardiovascular abnormalities that can progress or cause sudden cardiac death. Such screening efforts have the capability of raising the clinical suspicion of several cardiovascular diseases—usually by virtue of a heart murmur, regarded to be of potential clinical significance, cardiac symptoms (e.g., exertional chest pain, disproportionate dyspnea, or impairment in consciousness), or a family history of heart disease or sudden unexpected death. However, a major obstacle to implementation of large-scale screening in the U.S. is the substantial number of young athletes eligible for evaluation (about 10 to 12 million) and the rarity of the cardiac diseases capable of causing sudden death in this population (estimated prevalence, less than or equal to 0.3%) (2).

Customary screening strategies for U.S. high school and college athletes is confined to history-taking and physical examination, generally acknowledged to be limited in its power to consistently identify important cardiovascular abnormalities. In one retrospective study, only 3% of trained athletes who died suddenly of heart disease (and had been exposed to preparticipation screening) were suspected of harboring cardiovascular disease on the basis of history and physical examination—and none had been disqualified from competition (3). Although most trained athletes with occult cardiovascular disease are asymptomatic, the prior history of

exertional-related syncope in a young athlete unavoidably raises the consideration of a number of diseases known to cause sudden cardiac death, including hypertrophic cardiomyopathy (HCM) and ion-channel disorders, but in particular should also heighten the level of clinical suspicion for congenital coronary anomalies of wrong sinus origin (4).

Furthermore, the quality of cardiovascular screening for U.S. high school and college athletes, particularly the design of approved questionnaires, has come under scrutiny regarding inadequacies (5,6) when measured against American Heart Association (AHA) recommendations (1) (Table 1). Legislation in several states allows health care workers with vastly different levels of training and expertise (including chiropractors and naturopathic clinicians) to conduct preparticipation sports examinations, often under suboptimal conditions. Improvement in this screening process, including the training level of examiners, would undoubtedly result in a greater number of athletes identified with previously unsuspected but clinically relevant cardiovascular abnormalities. Indeed, development and dissemination of a standardized and uniform national preparticipation history and physical examination form for medical screening in all high schools and colleges (which incorporates the AHA recommendations) would be the most practical approach for achieving this goal.

Certainly, the diagnosis of genetic diseases, such as HCM, arrhythmogenic right ventricular cardiomyopathy (ARVC), long QT and Brugada syndromes, and other inherited arrhythmia syndromes in asymptomatic patients has now taken on even greater relevance. This is because individuals judged to be at sufficiently high-risk may be eligible for primary prevention of sudden cardiac death with an implantable cardioverter-defibrillator (7,8).

Conversely, in Italy, for the past 25 years, a formal national preparticipation screening and medical clearance program has been mandated for young competitive athletes in organized sports programs (9,10). The Italian system is unique by virtue of requiring annual evaluations that routinely include a 12-lead electrocardiogram (ECG) as well as a history and physical examination; the ECG itself has proven most useful in the identification of many previously undiagnosed athletes with HCM (10). However, such screening efforts may be complicated by the substantial proportion of false-positive test results that potentially represent a major burden to athletes, their families, and the

Table 1. AHA Consensus Panel Recommendations for Preparticipation Athletic Screening (1)

Family History
1. Premature sudden cardiac death
2. Heart disease in surviving relatives less than 50 years old
Personal History
3. Heart murmur
4. Systemic hypertension
5. Fatigue
6. Syncope/near-syncope
7. Excessive/unexplained exertional dyspnea
8. Exertional chest pain
Physical Examination
9. Heart murmur (supine/standing*)
10. Femoral arterial pulses (to exclude coarctation of aorta)
11. Stigmata of Marfan syndrome
12. Brachial blood pressure measurement (sitting)

*In particular, to identify heart murmur consistent with dynamic obstruction to left ventricular outflow. From Maron BJ, et al. *Circulation* 1996;94:850-6, reprinted with permission of the American Heart Association.

testing facilities. Obstacles in the U.S. to implementing obligatory government-sponsored national screening including ECGs or echocardiograms are the particularly large population of athletes to screen, major cost-benefit considerations, and the recognition that it is impossible to absolutely eliminate the risks associated with competitive sports (1,2). Nevertheless, some volunteer-based small-scale screening programs using portable echocardiograms to examine high-school athletes on the field for HCM have emerged.

Systematic preparticipation cardiovascular screening, primarily to exclude atherosclerotic coronary artery disease in older athletes, is not customary practice. Such persons are largely participants in individual athletic activities such as road and marathon racing, or in a variety of other organized master's sports (11).

DIAGNOSTIC TESTING STRATEGIES

When a cardiovascular abnormality is initially suspected (by formal screening or otherwise), the diagnostic strategy should focus on the systematic exclusion of those conditions known to cause sudden death in young athletes; these approaches include echocardiography, ECG, history, and physical examination. Additional noninvasive (and invasive) testing with cardiac magnetic resonance imaging (CMR), exercise testing, ambulatory Holter ECG recording, implanted loop recorder, tilt table examination, or electrophysiologic testing with programmed stimulation can be considered in selected patients. Diagnostic myocardial biopsies are used only selectively in athletes suspected clinically of myocarditis.

Despite considerable assembled data regarding DNA-based diagnosis over the past decade, identification of genetic cardiovascular diseases such as HCM, long QT syndrome, and other ion-channel disorders, ARVC, and Marfan syndrome continues to be made through clinical testing in the vast majority of cases, and this will remain so

in the foreseeable future. At present, genetic testing is not easily available on a routine clinical basis for most genetic heart diseases, or for application to large athletic populations given the expensive and complex methodologies involved and the genetic heterogeneity characteristic of these diseases (12).

Echocardiography. Two-dimensional echocardiography is the principal diagnostic imaging modality for clinical identification of HCM by demonstrating otherwise unexplained and usually asymmetric left ventricular (LV) wall thickening (12,13). In this regard, a maximal LV end-diastolic wall thickness of 15 mm or more (or on occasion, 13 or 14 mm) is the absolute dimension generally accepted for the clinical diagnosis of HCM in an adult athlete (two or more standard deviations from the mean relative to body surface area; z-score of two or more in children) (12,13); however, any specific LV wall thickness (including normal) is theoretically compatible with the presence of a mutant HCM gene (12,14). Echocardiography would also be expected to detect and define other specific and relevant congenital structural abnormalities associated with sudden death or disease progression in young athletes such as valvular heart disease (e.g., mitral valve prolapse and aortic valve stenosis), aortic root dilatation and mitral valve prolapse in Marfan or related syndromes, and LV dysfunction and/or enlargement (evident in myocarditis and dilated cardiomyopathy). Such diagnostic testing requires interpretation by physicians trained in echocardiography, but cannot guarantee full recognition of all relevant lesions, and some important diseases may escape detection despite expert screening methodology. For example, the HCM phenotype may not be evident when echocardiography is performed in the pre-hypertrophic phase (i.e., a patient less than 14 years of age) (12). Annual serial echocardiography is recommended in HCM family members throughout adolescence (12,14).

Electrocardiography. The 12-lead ECG may be of use in the diagnosis of cardiovascular disease in young athletes, and has been promoted as a practical and cost-effective strategic alternative to routine echocardiography for population-based preparticipation screening. For example, the ECG is abnormal in up to 75% to 95% of patients with HCM, and often before the appearance of hypertrophy (12). The ECG will also identify many individuals with the long QT, Brugada, and other inherited syndromes associated with ventricular arrhythmias. It raises the suspicion of myocarditis by premature ventricular complexes and ST-T abnormalities, or ARVC by T-wave inversion in leads V₁ through V₃ and low amplitude potentials (epsilon waves) (1,2). Of note, however, a not inconsequential proportion of genetically affected family members with long QT syndrome may not express QT interval prolongation, and ECG abnormalities are usually absent in random recordings from patients with congenital coronary artery anomalies (4).

Other tests. In those cases in which the echocardiogram is normal or borderline for LV hypertrophy, but a suspicion

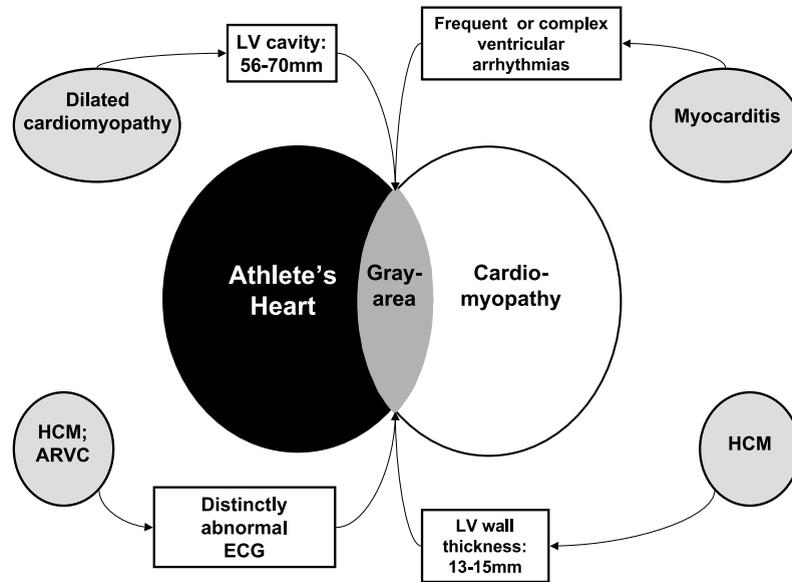


Figure 1. Gray area of overlap between athlete's heart and cardiomyopathies, including myocarditis, hypertrophic cardiomyopathy (HCM), and arrhythmogenic right ventricular cardiomyopathy (ARVC). ECG = electrocardiogram; LV = left ventricular. From Maron BJ, *N Engl J Med* 2003;349:1064-75, with permission of Massachusetts Medical Society (2).

for HCM persists (often due to an abnormal 12-lead ECG), CMR may be useful in clarifying wall thickness or detecting segmental areas of hypertrophy in selected regions of the LV chamber which may be more difficult to image reliably with conventional echocardiography, such as anterolateral free wall or apex (15,16).

Definitive identification of congenital coronary artery anomalies of wrong sinus origin usually requires sophisticated laboratory imaging, including multi-slice computed tomography or coronary arteriography. However, in young athletes it is possible to raise the suspicion of these malformations with transthoracic or transesophageal echocardiography or CMR imaging. Often, ARVC cannot be diagnosed reliably with echocardiography, and CMR is probably the most useful noninvasive test for identifying the structural abnormalities in this condition (i.e., right ventricular enlargement, wall motion abnormalities, adipose tissue replacement within the wall, and aneurysm formation); however, CMR is not an entirely sensitive or specific diagnostic modality in ARVC (17).

ATHLETE'S HEART AND CARDIOVASCULAR DISEASE

Systematic training in endurance or isometric sports may trigger physiologic adaptations and structural cardiac remodeling, including increased LV wall thickness, enlarged ventricular and atrial cavity dimensions, and calculated cardiac mass, in the presence of normal systolic and diastolic function (i.e., athlete's heart) (18). The magnitude of physiologic hypertrophy may also vary according to the particular type of sports training. Other adaptations to training include a variety of abnormal 12-lead ECG patterns in about 40% of elite athletes, which not infrequently

mimic those of cardiac disease (i.e., increased R- or S-wave voltages, Q waves, and repolarization abnormalities) (19). Frequent and/or complex ventricular tachyarrhythmias on ambulatory (Holter) ECG are not uncommonly found in athletes and can also mimic certain cardiac diseases, including myocarditis (20).

Clinical distinctions between physiologic athlete's heart and pathologic conditions (18-23) have critical implications for trained athletes, because cardiovascular abnormalities may trigger disqualification from competitive sports to reduce the risk of sudden death or disease progression. An over-diagnosis may lead to unnecessary restrictions, depriving athletes of the psychological, social, or possibly (in some elite athletes) economic benefits of sports (2).

Morphologic adaptations of athlete's heart can closely resemble certain cardiovascular diseases and lead to a differential diagnosis with HCM, dilated cardiomyopathy, and ARVC (2) (Fig. 1). Such clinical dilemmas not infrequently arise when cardiac dimensions fall outside clinically accepted partition values. For example, 2% of highly trained adult male athletes show relatively mild increases in LV wall thickness (13 to 15 mm) and 15% have LV cavity enlargement greater than or equal to 60 mm (2,21,22); both fall into a borderline and inconclusive "gray zone" for which extreme expressions of benign athlete's heart and mild morphologic forms of cardiomyopathy overlap (2,22,23). Indeed, the two most common clinical scenarios encountered that unavoidably generate ambiguous diagnoses in trained athletes are: 1) differentiating HCM from athlete's heart in athletes with an LV wall thickness of 13 to 15 mm, non-dilated and normally contractile LV, and absence of mitral valve systolic anterior motion; and 2) differentiating

Task Force 1: Preparticipation Screening and Diagnosis

CMR imaging, genotyping, and serial acquisition of clinical and morphologic evidence over time.

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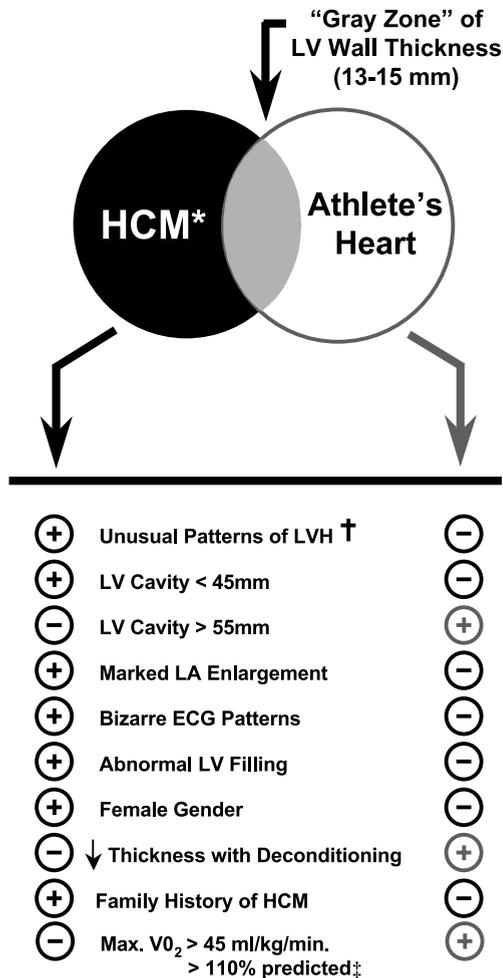


Figure 2. Chart showing criteria used to favor or distinguish hypertrophic cardiomyopathy (HCM) from athlete's heart when maximal left ventricular (LV) wall thickness is within shaded gray zone of overlap (i.e., 13 to 15 mm), consistent with both diagnoses. *Assumed to be the non-obstructive form of HCM (under resting conditions) in this discussion because the presence of substantial mitral valve systolic anterior motion would confirm the diagnosis of HCM in the athlete. †May involve a variety of abnormalities, including heterogeneous distribution of left ventricular hypertrophy (LVH) in which asymmetry is prominent and adjacent regions may show greatly different thicknesses with sharp transitions evident between segments, as well as unusual patterns in which the anterior ventricular septum is spared from the hypertrophic process and LV thickening may be in posterior portion of septum or anterolateral or posterior free wall or apex. ‡Assessed with cardiopulmonary (metabolic) exercise testing. ↓ = decreased; ECG = electrocardiogram; LA = left atrial. From Maron BJ, et al. *Circulation* 1995;91:1596-601, reprinted with permission of the American Heart Association (22).

early presentation of dilated cardiomyopathy from athlete's heart with LV end-diastolic cavity dimension 60 mm or more with low-normal LV function (i.e., ejection fraction of 50% to 55%).

Such cases with diagnostic uncertainty are not uncommon and may be resolved in many athletes by a number of independent noninvasive clinical parameters, including the response of cardiac mass to short periods of deconditioning, or assessment of diastolic filling (22) (Fig. 2). Clarification of such diagnostic ambiguities may also be achieved with

TASK FORCE 1 REFERENCES

1. Maron BJ, Thompson PD, Puffer JC, 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 the young), American Heart Association. *Circulation* 1996;94:850-6.
2. Maron BJ. Sudden death in young athletes. *N Engl J Med* 2003;349:1064-75.
3. Maron BJ, Shirani J, Poliac LC, Mathenge R, Roberts WC, Mueller FO. Sudden death in young competitive athletes: clinical, demographic, and pathological profiles. *JAMA* 1996;276:199-204.
4. Basso C, Maron BJ, Corrado D, Thiene G. Clinical profile of congenital coronary artery anomalies with origin from the wrong aortic sinus leading to sudden death in young competitive athletes. *J Am Coll Cardiol* 2000;35:1493-501.
5. Glover DW, Maron BJ. Profile of preparticipation cardiovascular screening for high school athletes. *JAMA* 1998;279:1817-9.
6. Pfister GC, Puffer JC, Maron BJ. Preparticipation cardiovascular screening for U.S. collegiate student-athletes. *JAMA* 2000;283:1597-9.
7. Maron BJ, Shen WK, Link MS, et al. Efficacy of implantable cardioverter-defibrillators for the prevention of sudden death in patients with hypertrophic cardiomyopathy. *N Engl J Med* 2000;342:365-73.
8. Corrado D, Leoni L, Link MS, et al. Implantable cardioverter-defibrillator therapy for prevention of sudden death in patients with arrhythmogenic right ventricular cardiomyopathy/dysplasia. *Circulation* 2003;108:3084-91.
9. Pelliccia A, Maron BJ. Preparticipation cardiovascular evaluation of the competitive athlete: perspectives from the 30-year Italian experience. *Am J Cardiol* 1995;75:827-9.
10. Corrado D, Basso C, Schiavon M, Thiene G. Screening for hypertrophic cardiomyopathy in young athletes. *N Engl J Med* 1998;339:364-9.
11. Maron BJ, Araújo CG, Thompson PD, et al. Recommendations for preparticipation screening and the assessment of cardiovascular disease in Master's athletes: an advisory for healthcare professionals from the working groups of the World Heart Federation, the International Federation of Sports Medicine, and the American Heart Association Committee on Exercise, Cardiac Rehabilitation, and Prevention. *Circulation* 2001;103:327-34.
12. Maron BJ, McKenna WJ, Danielson GK, et al. American College of Cardiology/European Society of Cardiology clinical expert consensus document on hypertrophic cardiomyopathy: a report of the American College of Cardiology Foundation Task Force on Clinical Expert Consensus Documents and the European Society of Cardiology Committee for Practice Guidelines. *J Am Coll Cardiol* 2003;42:1687-713.
13. Klues HG, Schiffers A, Maron BJ. Phenotypic spectrum and patterns of left ventricular hypertrophy in hypertrophic cardiomyopathy: morphologic observations and significance as assessed by two-dimensional echocardiography in 600 patients. *J Am Coll Cardiol* 1995;26:1699-708.
14. Maron BJ, Seidman JG, Seidman CE. Proposal for contemporary screening strategies in families with hypertrophic cardiomyopathy. *J Am Coll Cardiol* 2004;44:2125-32.
15. Rickers C, Wilke NM, Jerosch-Herold M, et al. Utility of cardiac magnetic resonance imaging in the diagnosis of hypertrophic cardiomyopathy. *Circulation* 2005. In press.
16. Moon JCC, Fisher NG, McKenna WJ, Pennell DJ. Detection of apical hypertrophic cardiomyopathy by cardiovascular magnetic resonance in patients with non-diagnostic echocardiography. *Heart* 2004;90:645-49.
17. Tandri H, Calkins H, Nasir K, et al. Magnetic resonance imaging findings in patients meeting task force criteria for arrhythmogenic right ventricular dysplasia. *J Cardiovasc Electrophysiol* 2003;14:476-82.

18. Pluim BM, Zwinderman AH, van der Laarse A, van der Wall EE. The athlete's heart: a meta-analysis of cardiac structure and function. *Circulation* 2000;101:336–44.
19. Pelliccia A, Maron BJ, Culasso F, et al. Clinical significance of abnormal electrocardiographic patterns in trained athletes. *Circulation* 2000;102:278–84.
20. Biffi A, Pelliccia A, Verdile L, et al. Long-term clinical significance of frequent and complex ventricular tachyarrhythmias in trained athletes. *J Am Coll Cardiol* 2002;40:446–52.
21. Pelliccia A, Maron BJ, Spataro A, Proschan MA, Spirito P. The upper limit of physiologic cardiac hypertrophy in highly trained elite athletes. *N Engl J Med* 1991;324:295–301.
22. Maron BJ, Pelliccia A, Spirito P. Cardiac disease in young trained athletes: insights into methods for distinguishing athlete's heart from structural heart disease, with particular emphasis on hypertrophic cardiomyopathy. *Circulation* 1995;91:1596–601.
23. Pelliccia A, Culasso F, Di Paolo FM, Maron BJ. Physiologic left ventricular cavity dilatation in elite athletes. *Ann Intern Med* 1999;130:23–31.

Appendix 1. Author Relationships With Industry and Others

Name	Consultant	Research Grant	Speaker's Bureau	Stock Holder	Expert Witness Testimony
Dr. Barry J. Maron	None	• Medtronic	None	None	• 1996, Defense, Northwestern vs. Knapp
Dr. Pamela S. Douglas	None	None	None	None	None
Dr. Rick A. Nishimura	None	None	None	None	None
Dr. Thomas P. Graham	None	None	None	None	None
Dr. Paul D. Thompson	<ul style="list-style-type: none"> • Astra Zeneca • Bristol Myers Squibb 	<ul style="list-style-type: none"> • Astra Zeneca • Merck • Pfizer • Schering 	<ul style="list-style-type: none"> • Astra Zeneca • Merck • Pfizer • Schering 	<ul style="list-style-type: none"> • Pfizer • Schering 	<ul style="list-style-type: none"> • 2005, Plaintiff, Sudden death in college athlete • 2004, Defense, Stress test/recreational sports • 2002, Sudden death, World Gym • 2002, Plaintiff, Sudden death, truck driver • 1998, Plaintiff, Sudden death, recreational sports

Task Force 2: Congenital Heart Disease

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GENERAL CONSIDERATIONS

The most common congenital heart lesions that have been associated with sudden death during sports participation are hypertrophic cardiomyopathy, coronary artery anomalies, Marfan syndrome, and aortic valve disease (1–3). Less common lesions include complex defects, such as transposition and single ventricle, and those with associated pulmonary vascular disease.

The recommendations presented are intended to provide broad guidelines for patients with congenital heart defects (4). When questions about the safety of sports participation arise, there is no substitute for a comprehensive evaluation by a knowledgeable and experienced physician. Exercise testing can be useful, particularly if symptoms, the electrocardiogram (ECG), and blood pressure are monitored during conditions that simulate the sport in question. Arrhythmias discussed in this Task Force are usually identified by exercise testing or some form of long-term monitoring (including ambulatory Holter and event recording). Serial evaluations may be required because of changing hemodynamic status with time.

TYPES OF CONGENITAL DEFECTS

Atrial septal defect (ASD)—untreated. Most children with ASD are asymptomatic, and closure is usually carried out before they are active in competitive sports. An ECG and echocardiogram are required for evaluation. Small atrial defects are characterized by minimal or no right ventricular volume overload, moderate or large defects have significant volume overload but pulmonary hypertension is unusual.

Recommendations:

1. **Athletes with small defects, normal right heart volume, and no pulmonary hypertension can participate in all sports.**
2. **Athletes with a large ASD and normal pulmonary artery pressure can participate in all competitive sports.**
3. **Athletes with an ASD and mild pulmonary hypertension can participate in low-intensity competitive sports (class IA). Patients with associated pulmonary vascular obstructive disease who have cyanosis and a large right-to-left shunt cannot participate in competitive sports.**

4. **Athletes with ASD and symptomatic atrial or ventricular tachyarrhythmia or moderate-to-severe mitral regurgitation should follow the recommendations in Task Force 1: Preparticipation Screening and Diagnosis of Cardiovascular Disease in Athletes, and Task Force 6: Coronary Artery Disease, respectively.**

ASD—closed at operation or by interventional catheterization. The ASDs usually are completely closed by operation or device insertion. When closure is performed in childhood there is little or no residual right ventricular enlargement. Supraventricular arrhythmias can occur after closure and are more common when the defect is repaired later in life (5–7). Evaluation should include an estimate of cardiac performance, pulmonary vascular resistance and right ventricular size, and a search for conduction or rhythm disturbances. A chest radiograph, ECG, and echocardiogram usually are needed. Patients with preoperative pulmonary hypertension, a right-to-left shunt, or both, before operation require postoperative assessment of pulmonary artery pressure with Doppler echocardiography or cardiac catheterization.

Recommendations:

1. **Three to six months after the operation or intervention, patients can participate in all sports unless the following are present: 1) evidence of pulmonary hypertension; 2) symptomatic atrial or ventricular tachyarrhythmias or second- or third-degree heart block; and 3) evidence of myocardial dysfunction.**
2. **Patients with any of these abnormalities should have an exercise evaluation and an individualized exercise prescription with respect to competitive sports. For related recommendations on competitive sports participation, see the section entitled Elevated Pulmonary Resistance and Ventricular Dysfunction After Cardiac Surgery in this Task Force report, as well as Task Force 7: Arrhythmias.**

Ventricular septal defect (VSD)—untreated. The VSDs are categorized as small, moderate, or large. If physical examination and echocardiography indicate normal heart size and normal pulmonary artery pressure in patients with a suspected small VSD, further evaluation is not required.

A patient with a VSD that does not fit into the small defect category may require further investigation. Cardiac catheterization might be required.

A moderate-sized defect with low pulmonary vascular resistance will have a pulmonary/systemic flow ratio of about 1.5 to 1.9. A large defect with low or mildly increased pulmonary vascular resistance is defined as a pulmonary/systemic flow ratio greater than or equal to 2 and pulmonary resistance less than 3 U/m². Large right-to-left shunts are discussed in the section entitled Elevated Pulmonary Resistance.

Recommendations:

1. **Athletes with a VSD and normal pulmonary artery pressure can participate in all sports.**
2. **Athletes with a large VSD who do not have marked elevation of pulmonary resistance are candidates for repair, and full participation in all sports would normally occur after a successful VSD closure.**

VSD—closed at operation or by interventional catheterization. Successful repair is characterized by the absence of symptoms, absence of significant shunt, cardiomegaly, or arrhythmias, and the presence of normal pulmonary artery pressure. Minimal diagnostic evaluation after surgery before participation in sports includes a chest radiograph, ECG, and echocardiogram. Patients with residual left or right ventricular enlargement, myocardial dysfunction, or pulmonary hypertension may require an exercise test or cardiac catheterization before a decision for participation in sports can be made.

Recommendations:

1. **At three to six months after repair, asymptomatic athletes with no defect or only a small residual defect can participate in all competitive sports if they have no evidence of pulmonary artery hypertension, ventricular or atrial tachyarrhythmia, or myocardial dysfunction.**
2. **Athletes with symptomatic atrial or ventricular tachyarrhythmias or second- or third-degree atrioventricular (AV) block should follow the recommendations in Task Force 7: Arrhythmias. Athletes with mild-to-moderate pulmonary hypertension or ventricular dysfunction should follow the recommendations in the section entitled Elevated Pulmonary Resistance or Ventricular Dysfunction After Cardiac Surgery.**
3. **Athletes with persistent, severe pulmonary hypertension cannot participate in competitive sports (see section entitled Elevated Pulmonary Resistance).**

Patent ductus arteriosus (PDA)—untreated. The patient with a small PDA has a characteristic murmur, absence of symptoms, and normal left heart chamber dimensions. Those with a larger patent ductus have cardiomegaly and widened pulse pressure. There may be evidence of pulmonary hypertension. Minimal diagnostic studies generally include echocardiography.

Recommendations:

1. **Athletes with a small PDA and normal left heart chamber dimension can participate in all competitive sports.**
2. **Athletes with a moderate or large PDA, causing left ventricular (LV) enlargement, should undergo surgical or interventional catheterization closure before unrestricted competition.**

Table 1. Definitions of Mild, Moderate, and Severe Aortic Stenosis

	Peak-to-Peak Systolic Gradient (Catherization)	Mean Echo Doppler Gradient (CW)	Peak Instantaneous Echo Doppler Gradient (CW)*
Mild AS	less than 30 mm Hg	less than 25 mm Hg	less than 40 mm Hg
Moderate AS	30 to 50 mm Hg	25 to 40 mm Hg	40 to 70 mm Hg
Severe AS	greater than 50 mm Hg	greater than 40 mm Hg	greater than 70 mm Hg

*Gradients obtained from apical window usually most predictive of catheter gradient.

3. For athletes with a moderate or large PDA, severe pulmonary hypertension, and cyanosis, see the section entitled Elevated Pulmonary Resistance.

PDA, closed at operation or by interventional catheterization. A successful result is characterized by the absence of symptoms, a normal cardiac examination, and normal echo/Doppler study.

Recommendations:

1. Three months after PDA closure, patients with no symptoms, with normal cardiac examination, and with no evidence of pulmonary hypertension or LV enlargement can participate in all competitive sports.
2. For athletes with residual pulmonary artery hypertension see the section entitled Elevated Pulmonary Resistance.

Pulmonary valve stenosis (PS)—untreated. Mild stenosis is characterized by a systolic ejection murmur, variable ejection click, and a normal ECG. A Doppler peak instantaneous gradient less than 40 mm Hg usually indicates mild stenosis, 40 to 60 mm Hg moderate stenosis, and greater than 60 mm Hg severe stenosis. Most patients with a gradient 50 mm Hg or greater undergo balloon valvuloplasty.

Recommendations:

1. Athletes with a peak systolic gradient less than 40 mm Hg and normal right ventricular function can participate in all competitive sports if no symptoms are present. Annual re-evaluation is recommended.
2. Athletes with a peak systolic gradient greater than 40 mm Hg can participate in low-intensity competitive sports (classes IA and IB). Patients in this category usually are referred for balloon valvuloplasty or operative valvotomy before sports participation.

PS—treated by operation or balloon valvuloplasty. Adequate relief of PS is characterized by the absence or improvement in symptoms, an improved physical examination, and Doppler study showing mild or no residual gradient and mild or no pulmonary valvular regurgitation.

Recommendations:

1. Athletes with no or only residual mild PS and normal ventricular function without symptoms can participate in all competitive sports. Participation in sports can begin two to four weeks after balloon valvulo-

plasty. After operation, an interval of approximately three months is suggested before resuming sports participation.

2. Athletes with a persistent peak systolic gradient greater than 40 mm Hg should follow the same recommendations as those for patients before treatment.
3. Athletes with severe pulmonary incompetence characterized by a marked right ventricular enlargement can participate in class IA and IB competitive sports.

Aortic valve stenosis (AS)—untreated. This section discusses congenital valvular AS in young patients; AS in adults is addressed in Task Force 3. Differentiation between mild and either moderate or severe AS is accomplished by physical examination, ECG, and Doppler echocardiography. The distinction between moderate or severe stenosis is more difficult and may require cardiac catheterization in rare situations when clinical examination, ECG, echocardiography, and/or other data are discrepant. Patients with a history of fatigue, light-headedness, dizziness, syncope, chest pain, or pallor on exercise deserve a full evaluation, which may include cardiac catheterization and exercise testing. Because AS may progress, periodic re-evaluation is needed. Sudden death is more likely to occur in patients with severe LV hypertrophy, exertional syncope, chest pain or dyspnea and a LV strain pattern on the ECG. Between approximately 20% and 80% of sudden deaths in patients with severe AS have been found to occur on physical exertion (2,8).

For the purpose of these recommendations, Table 1 summarizes the definitions of mild moderate and severe AS. It should be emphasized that virtually all patients classified as moderate or severe AS would be expected to have left ventricular hypertrophy by echocardiography.

Recommendations:

1. Athletes with mild AS can participate in all competitive sports if they have a normal ECG, normal exercise tolerance, and no history of exercise-related chest pain, syncope, or atrial or ventricular tachyarrhythmia associated with symptoms.
2. Athletes with moderate AS can participate in low static/low-to-moderate dynamic, and moderate static/low-to-moderate dynamic (classes IA, IB, and IIA) competitive sports if the following conditions are met:
 - Mild or no LV hypertrophy by echocardiography and the absence of LV strain pattern on the ECG.

- Normal exercise test without evidence of myocardial ischemia or atrial or ventricular tachyarrhythmia and with normal exercise duration and blood pressure response. Those athletes with supraventricular tachycardia or multiple or complex ventricular tachyarrhythmias at rest or with exercise can participate only in low-intensity competitive sports, classes IA and IB.
 - Absence of symptoms, as defined in the preceding text.
3. Athletes with severe AS should not participate in competitive sports.

The criteria in this section also apply to athletes with *discrete (membranous) subaortic stenosis* and *supravalvular aortic stenosis*.

Aortic stenosis—treated by operation or balloon valvuloplasty. After operation, a variable degree of residual stenosis or regurgitation, or both, can be present. Re-evaluation by physical examination, ECG, and echocardiography is necessary for reassessment. In addition, exercise stress testing or catheterization, or both, can be required for patients whose physiologic and anatomic severity cannot otherwise be determined.

Recommendations:

1. Athletes with residual mild, moderate, or severe stenosis should follow the same recommendations as previously defined for untreated patients.
2. Athletes with moderate to severe aortic regurgitation should follow the recommendations in Task Force 3: Valvular Heart Disease.
3. Because of the propensity for recurrence of LV outflow obstruction in discrete subaortic stenosis postoperatively, these patients require continued follow-up and re-evaluation annually for exercise recommendations. This recommendation also applies to all other forms of fixed AS.

Coarctation of the aorta—untreated. This abnormality is characterized by obstruction usually present in the juxtaductal or juxtaligamentary area. Severity is assessed by the arm and leg pressure gradient, physical examination, exercise testing, echocardiographic/Doppler studies, and cardiac magnetic resonance (CMR) studies. Virtually all patients, except those with mild coarctation, will undergo either surgical repair or balloon dilation/stenting.

Recommendations:

1. Athletes with mild coarctation and the absence of large collateral vessels or significant aortic root dilation (z -score 3.0 or less) (score 3.0 = 3 standard deviations from the mean for patient size), with a normal exercise test and a small pressure gradient at rest (usually 20 mm Hg or less between upper and lower limbs), and a peak systolic blood pressure 230

mm Hg or less with exercise can engage in all competitive sports.

2. Athletes with a systolic arm/leg gradient more than 20 mm Hg or exercise-induced hypertension with a systolic blood pressure more than 230 mm Hg can engage in only low-intensity competitive sports (class IA) until treated.

Coarctation of the aorta—treated by surgery or balloon angioplasty. The majority of patients will have coarctation repair or balloon arterioplasty performed during childhood. After repair, abnormalities can persist, such as a mild residual gradient, ventricular hypertrophy, systemic hypertension, and residual obstruction evident on exercise (9,10). Before a decision for participation, minimal diagnostic studies, including a chest radiograph, ECG, exercise testing, and echocardiographic evaluation of LV function and aortic anatomy, should be done. Magnetic resonance imaging can be useful to document residual anatomical abnormalities, aortic dilation, or aneurysm formation.

Recommendations:

1. Participation in sports, three or more months after surgical or balloon angioplasty for coarctation of the aorta, is permitted for athletes with a 20 mm Hg or less arm/leg blood pressure gradient at rest and a normal peak systolic blood pressure during rest and exercise.
2. During the first postoperative year, athletes should refrain from high-intensity static exercise (classes IIIA, IIIB, and IIIC) and sports that pose the danger of bodily collision.
3. After three months, if patients continue to be asymptomatic, with normal blood pressure at rest and exercise, all sports are permissible except those with a large static component (particularly, classes IIIA, IIIB, and IIIC).
4. For athletes with evidence of significant aortic dilation, wall thinning, or aneurysm formation, participation should be restricted to low-intensity competitive sports (classes IA and IB).

Elevated pulmonary resistance with congenital heart disease. Patients who have pulmonary vascular disease and congenital heart disease are at risk for sudden death during sports activity. As pulmonary vascular obstruction progresses, these patients develop cyanosis at rest and intense cyanosis with exercise. Although most of these patients self-limit their activity, they should not participate in competitive sports.

Patients who have suspected elevated pulmonary artery pressure after operation or interventional catheterization for shunt lesions should be evaluated by echocardiography and/or cardiac catheterization before engaging in competitive athletics.

Recommendations:

1. If pulmonary artery peak systolic pressure is 30 mm Hg or less, athletes can participate in all sports.
2. If pulmonary artery pressure is more than 30 mm Hg, a full evaluation and individual exercise prescription are required for athletic participation.

Ventricular dysfunction after cardiac surgery. Left and/or right ventricular dysfunction can occur after surgical treatment of both simple and complex congenital heart diseases and affect exercise performance. Periodic assessment of ventricular function is required for participation in sports because ventricular function may deteriorate over time.

Recommendations:

1. For full participation, normal or near-normal ventricular function is required (ejection fraction 50% or more).
2. Athletes with mildly depressed ventricular function (ejection fraction 40% to 50%) should participate in low-intensity static competitive sports only (classes IA, IB, and IC).
3. Athletes with moderately to severely depressed ventricular function (ejection fraction less than 40%) should not participate in competitive sports.

Cyanotic congenital cardiac disease—unoperated. Cyanotic congenital heart disease is associated with exercise intolerance and progressive hypoxemia with increasing effort. Patients are unlikely to engage in competitive sports because of their own self-limiting activity. There are rare patients with cyanotic congenital heart disease (such as pulmonary stenosis or mildly elevated pulmonary vascular resistance plus atrial or ventricular defects) who reach adolescence or even adult life with mild cyanosis at rest and shortness of breath only with exercise. These patients may experience a profound increase in cyanosis during sports participation.

Recommendation:

1. Patients with untreated cyanotic heart disease can usually participate in low-intensity competitive sports of only class IA.

Postoperative palliated cyanotic congenital heart disease. Palliative surgical intervention can be performed to increase pulmonary blood flow in patients with decreased flow or to limit blood flow in those with excessive flow. Often these patients have significant relief of symptoms at rest, but arterial desaturation during exercise frequently persists.

Recommendation:

1. Patients can participate in low-intensity competitive sports (class IA), provided that the following criteria are met:

- Arterial saturation remains above approximately 80%
- Tachyarrhythmias associated with symptoms of impaired consciousness not present
- There is not moderate/severe ventricular dysfunction.

Postoperative tetralogy of Fallot (T/F). The current treatment for T/F is early repair, and patients have varying degrees of pulmonary stenosis and mild/moderate pulmonary insufficiency. Diagnostic evaluation usually includes physical examination, chest radiograph, echocardiography, CMR, ambulatory ECG monitoring, and exercise testing. Cardiac catheterization and/or exercise testing may be required for complete evaluation in selected patients, particularly those with significant cardiomegaly and/or symptoms. Patients with important residual abnormalities, such as a significant left-to-right shunt, right ventricular hypertension, moderate-to-severe pulmonary regurgitation, or right ventricular dysfunction, who also have a history of syncope and/or arrhythmia, may be at risk for sudden death (11).

Recommendations:

1. Athletes with an excellent repair should be allowed to participate in all sports, providing that the following criteria are met:
 - Normal or near-normal right heart pressure
 - No or only mild right ventricular volume overload
 - No evidence of a significant residual shunt
 - No atrial or ventricular tachyarrhythmia abnormality on ambulatory ECG monitoring or exercise testing
2. Patients with marked pulmonary regurgitation and right ventricular volume overload, residual right ventricular hypertension (peak systolic right ventricular pressure greater than or equal to 50% systemic pressure), or atrial or ventricular tachyarrhythmias, should participate in low-intensity competitive sports only (class IA).

Transposition of the great arteries (TGA)—postoperative Mustard or Senning operation. Patients who have had atrial repair of TGA can have significant hemodynamic abnormalities including impaired systemic venous return, abnormal right ventricular (systemic ventricular) function, pulmonary stenosis or pulmonary hypertension, abnormalities of pulmonary venous return, tricuspid insufficiency, and significant atrial or ventricular arrhythmias (12). After atrial repair, the right ventricle is subjected to systemic pressure, and because of the intrinsic properties of the right ventricle its reserve is believed to be less than that of the left ventricle. The consequences of hypertrophy and dilation in a trained athlete after an otherwise excellent atrial repair of transposition are unknown. Evaluation before training and competition in moderate- and low-intensity sports should include history and examination, chest radiograph, ECG, echocardiogram, CMR, ambulatory ECG

monitoring, and exercise testing. For patients in whom data are unclear with regard to hemodynamic abnormalities or ventricular function, cardiac catheterization may be necessary.

Recommendations:

1. **Selected patients can engage in low and moderate static/low dynamic competitive sports (classes IA and IIA) provided there is:**
 - Mild or no cardiac chamber enlargement on chest radiograph, echocardiography, or CMR
 - No history of atrial flutter, supraventricular tachycardia, or ventricular tachyarrhythmia
 - No history of syncope or other cardiac symptoms
 - A normal exercise test defined as normal duration, workload, heart rate, ECG, and blood pressure response for age and gender.
2. **Patients not in this category require an individualized exercise prescription.**

Postoperative arterial switch for TGA. A significant cohort of patients have had arterial switch repair of TGA and are now old enough to engage in competitive sports. These patients have a low prevalence of ventricular dysfunction, arrhythmia with symptoms, and hemodynamic sequelae (with the possible exception of pulmonary artery or anastomotic stenosis and neo-aortic root dilation). Only limited exercise data are available.

Recommendations:

1. **Athletes with normal ventricular function, normal exercise test, and no atrial or ventricular tachyarrhythmias can participate in all sports.**
2. **Athletes with more than mild hemodynamic abnormalities or ventricular dysfunction can participate in low and moderate static/low dynamic competitive sports (classes IA, IB, IC, and IIA), provided that their exercise test is normal.**

Congenitally corrected transposition of the great arteries (CCTGA). Usually, CCTGA is associated with other congenital malformations of the heart, such as ventricular septal defect, pulmonary stenosis, and systemic AV valve abnormalities, which may dictate the level of participation in competitive sports. These patients are at risk for development of supraventricular tachycardia and spontaneous AV block.

Recommendations:

1. **Asymptomatic patients with CCTGA without other cardiac abnormalities may be eligible for participation in class IA and IIA sports if there is no systemic ventricle enlargement, no evidence of atrial or ventricular tachyarrhythmia on ambulatory ECG monitoring or exercise testing or normal exercise tests (including normal maximum oxygen consumption for age and gender).**

2. **Periodic re-evaluation is important to detect development of arrhythmias and deterioration of systemic (right) ventricular function and systemic (tricuspid) AV valve regurgitation. In particular, sports with a large static component (classes IIIA, IIIB, IIIC) such as power weightlifting are not recommended.**

Postoperative Fontan operation. The Fontan operation is characterized by systemic venous return bypassing the right ventricle. The operation is used for the long-term palliation of patients with tricuspid atresia or other complex types of single ventricle. Although many patients improve clinically after the Fontan operation, they usually have limited exercise capacity and reduced cardiac output at rest and during exercise (13). Postoperative arrhythmias have been associated with significant morbidity and mortality. Diagnostic evaluation before sports participation should include a chest radiograph, ECG, echocardiography or CMR, and exercise testing with oxygen saturations.

Recommendations:

1. **Athletes can participate in low-intensity competitive sports (class IA).**
2. **Athletes can engage in class IB sports if they have normal ventricular function and oxygen saturation.**

Ebstein's anomaly. A great deal of variability exists in the severity of this malformation, which is characterized by variable degrees of tricuspid regurgitation and right-heart enlargement caused by a malformed and displaced tricuspid valve. Cyanosis may be present due to atrial right-to-left shunting. Even mild cases may be associated with important arrhythmias. Severe cases can be associated with physical disability and increased risk for sudden death with exercise.

Recommendations:

1. **Athletes with a mild expression of Ebstein's anomaly without cyanosis, with normal right ventricular size, and with no evidence of atrial or ventricular tachyarrhythmias can participate in all sports.**
2. **Athletes with tricuspid regurgitation of moderate severity can participate in low-intensity competitive sports (class IA) if there is no evidence of arrhythmia on ambulatory ECG Holter monitoring other than isolated premature contractions.**
3. **Athletes with severe Ebstein's anomaly are precluded from all sports participation. However, after surgical repair, low-intensity competitive sports (class IA) can be permitted if tricuspid regurgitation is absent or mild, cardiac chamber size on chest radiograph or by echocardiography is not substantially increased, and symptomatic atrial or ventricular tachyarrhythmias are not present on ambulatory ECG monitoring and exercise test. Selected athletes with an excellent hemodynamic result after repair may be permitted additional participation on an individual basis.**

Congenital coronary artery anomalies. Congenital coronary anomalies of wrong sinus origin are the second most common cardiovascular cause of sudden death in young athletes (1). The most common of these malformations is anomalous origin of the left main coronary artery from the anterior (right) sinus of Valsalva, with an acute angled bend coursing between the pulmonary trunk and the anterior aspect of the aorta (14,15). Rare cases of anomalous origin of the right coronary artery from the left coronary sinus, congenitally hypoplastic coronary arteries, and anomalous origin of the left main coronary artery from the pulmonary trunk have also been associated with sudden cardiac death during exercise (1). Identification of these anomalies during life can be difficult because patients often do not experience warning symptoms, and rest and exercise ECGs are usually normal. Coronary anomalies should be considered in athletes with exertional syncope or symptomatic ventricular arrhythmia and should be investigated using appropriate studies such as echocardiography, CMR, or ultrafast computed tomography imaging. Coronary arteriography is indicated if other studies are not diagnostic. Surgery is usually performed when the diagnosis is made (16).

Recommendations:

1. **Detection of coronary anomalies of wrong sinus origin in which a coronary artery passes between great arteries should result in exclusion from all participation in competitive sports.**
2. **Participation in all sports three months after successful operation would be permitted for an athlete without ischemia, ventricular or tachyarrhythmia, or dysfunction during maximal exercise testing.**
3. **Athletes with previous myocardial infarction (MI) should follow the appropriate recommendations in Task Force 6: Coronary Artery Disease.**

A discussion of coronary artery myocardial bridging appears in Task Force 6: Coronary Artery Disease.

Kawasaki disease. Kawasaki disease, an acute, self-limited vasculitis of unknown etiology, is now the most common cause of acquired heart disease in children in the U.S. (17). Coronary artery aneurysms develop in approximately 20% of untreated children and in 4% of those treated with high-dose intravenous gammaglobulin in the acute phase (18). Coronary aneurysms, together with progressive coronary artery stenosis, can lead to ischemic heart disease, MI, or sudden death (19). Because coronary artery morphology evolves over time, the risk of exercise for the individual patient may change. Patients without coronary artery changes on echocardiography at any stage of the illness appear to have risk for ventricular tachyarrhythmias and sudden death similar to that of the normal population in the first 20 years after illness onset (19). When aneurysms regress to normal lumen diameter, structural and functional coronary abnormalities persist (20). Arteries with persistent

aneurysmal morphology may develop stenoses or occlusion, increasing the risk of myocardial ischemia (21). The risk associated with competitive sports in individuals who have had Kawasaki disease depends upon the degree of coronary artery involvement. Of note, because of the general cardiovascular benefits of exercise, all patients with Kawasaki disease should avoid a sedentary lifestyle.

Recommendations:

1. **Patients with no coronary artery abnormalities or transient coronary artery ectasia resolving during the convalescent phase of the disease are encouraged to participate in all sports after six to eight weeks.**
2. **Patients with regressed aneurysms can participate in all competitive sports if they have no evidence of exercise-induced ischemia by stress testing with myocardial perfusion imaging.**
3. **Patients with isolated small- to medium-sized aneurysms in one or more coronary arteries and judged to be at low risk for ischemic complications (normal left ventricular function, absence of exercise-induced ischemia or arrhythmia) may participate in low to moderate static and dynamic competitive sports (classes IA, IB, IIA, and IIB). Stress testing with evaluation of myocardial perfusion should be repeated at one- to two-year intervals to monitor ischemia and guide further recommendations about sports competition.**
4. **Patients with one or more large coronary aneurysms or multiple (segmented) or complex aneurysms with or without obstruction to coronary flow may participate in class IA and IIA sports if they have no evidence of reversible ischemia on stress testing, normal LV function, and absence of exercise-induced arrhythmia. Stress testing with evaluation of myocardial perfusion should be repeated at one-year intervals to monitor ischemia and guide further recommendations about sports competition.**
5. **Athletes with recent MI or revascularization should avoid competitive sports until their recovery is complete—usually six to eight weeks. Those with normal LV ejection fraction, exercise tolerance, absence of reversible ischemia or myocardial perfusion testing, and absence of exercise-induced arrhythmias can participate in class IA and IB sports. Those with left ventricular ejection fraction less than 40%, exercise intolerance, or exercise-induced ventricular tachyarrhythmias should not participate in competitive sports.**
6. **Patients with coronary lesions who are taking anti-coagulants and/or antiplatelet drugs (aspirin, clopidogrel) should not participate in sports that pose a danger of high speed collision.**

TASK FORCE 2 REFERENCES

1. Maron BJ. Sudden death in young athletes. *N Engl J Med* 2003;349:1064–75.
2. Driscoll DJ, Edwards WD. Sudden unexpected death in children and adolescents. *J Am Coll Cardiol* 1985;5:118B–21B.
3. Garson AJ, McNamara DG. Sudden death in a pediatric cardiology population, 1958 to 1983: relation to prior arrhythmias. *J Am Coll Cardiol* 1985;5:134B–7B.
4. Mitchell JH, Maron BJ, Epstein SE. 16th Bethesda conference: cardiovascular abnormalities in the athlete: recommendations regarding eligibility for competition: October 3–5, 1984. *J Am Coll Cardiol* 1985;6:1186–232.
5. Sealy WC, Fanner JC, Young WG J, Brown IW J. Atrial dysrhythmia and atrial secundum defects. *J Thorac Cardiovasc Surg* 1969;57:245–50.
6. Vetter VL, Horowitz LN. Electrophysiologic residua and sequelae of surgery for congenital heart defects. *Am J Cardiol* 1982;50:588–604.
7. Bink-Boelkens MT, Velvis H, van der Heide JJ, Eygelaar A, Hardjowijono RA. Dysrhythmias after atrial surgery in children. *Am Heart J* 1983;106:125–30.
8. Doyle EF, Arumugham P, Lara E, Rutkowski MR, Kiely B. Sudden death in young patients with congenital aortic stenosis. *Pediatrics* 1974;53:481–9.
9. Pelech AN, Kartodihardjo W, Balfe JA, Balfe JW, Olley PM, Leenen FH. Exercise in children before and after coarctectomy: hemodynamic, echocardiographic, and biochemical assessment. *Am Heart J* 1986; 112:1263–70.
10. Freed MD, Rocchini A, Rosenthal A, Nadas AS, Castaneda AR. Exercise-induced hypertension after surgical repair of coarctation of the aorta. *Am J Cardiol* 1979;43:253–8.
11. Garson AJ, Gillette PC, Gutgesell HP, McNamara DG. Stress-induced ventricular arrhythmia after repair of tetralogy of Fallot. *Am J Cardiol* 1980;46:1006–12.
12. Graham TP Jr. Hemodynamic residua and sequelae following intra-atrial repair of transposition of the great arteries: a review. *Pediatr Cardiol* 1982;2:203–13.
13. Driscoll DJ. Exercise responses in functional single ventricle before and after Fontan operation. *Prog Pediatr Cardiol* 1993;2:44–9.
14. Basso C, Maron BJ, Corrado D, Thiene G. Clinical profile of congenital coronary artery anomalies with origin from the wrong aortic sinus leading to sudden death in young competitive athletes. *J Am Coll Cardiol* 2000;35:1493–501.
15. Davis JA, Cecchin F, Jones TK, Portman MA. Major coronary artery anomalies in a pediatric population: incidence and clinical importance. *J Am Coll Cardiol* 2001;37:593–7.
16. Romp RL, Herlong JR, Landolfo CK, et al. Outcome of unroofing procedure for repair of anomalous aortic origin of left or right coronary artery. *Ann Thorac Surg* 2003;76:589–95.
17. Newburger JW, Takahashi M, Gerber MA, et al. Diagnosis, treatment, and long-term management of Kawasaki disease: a statement for health professionals from the Committee on Rheumatic Fever, Endocarditis and Kawasaki Disease, Council on Cardiovascular Disease in the Young. *Circulation* 2004;110:2747–71.
18. Terai M, Shulman ST. Prevalence of coronary artery abnormalities in Kawasaki disease is highly dependent on gamma globulin dose but independent of salicylate dose. *J Pediatr* 1997;131:888–93.
19. Kato H, Sugimura T, Akagi T, et al. Long-term consequences of Kawasaki disease: a 10- to 21-year follow-up study of 594 patients. *Circulation* 1996;94:1379–85.
20. Tsuda E, Kamiya T, Kimura K, Ono Y, Echigo S. Coronary artery dilatation exceeding 4.0 mm during acute Kawasaki disease predicts a high probability of subsequent late intima-medial thickening. *Pediatr Cardiol* 2002;23:9–14.
21. Tsuda E, Kamiya T, Ono Y, Kimura K, Kurosaki K, Echigo S. Incidence of stenotic lesions predicted by acute phase changes in coronary arterial diameter during Kawasaki disease (epub ahead of print). *Pediatr Cardiol* 2004;May 12.

Appendix 1. Author Relationships With Industry and Others

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Task Force 3: Valvular Heart Disease

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GENERAL CONSIDERATIONS

The following valvular disorders are considered here in the context of competitive athletes: mitral stenosis (MS), mitral regurgitation (MR), aortic stenosis (AS), aortic regurgitation (AR), tricuspid regurgitation (TR), tricuspid stenosis, and multivalvular disease. Recommendations are also provided for patients who have undergone valve repair or replacement.

The *diagnosis* of valvular heart disease usually can be made on the basis of the characteristic murmurs and associated findings on physical examination. Estimation of the *severity* of valvular disease is more difficult. In athletes, where secondary gain in denying symptoms is likely, the history may be unreliable. In truly asymptomatic patients, severity can usually be determined from the physical examination and Doppler echocardiography. Additional testing is often not necessary, but the electrocardiogram (ECG) and chest radiograph can also aid in assessing severity, and radionuclide angiography or cardiac magnetic resonance are helpful in assessing left ventricular (LV) function when the echocardiogram is of suboptimal quality.

Doppler echocardiography is the most important diagnostic test for valvular heart disease and is generally reliable in distinguishing the severity of the lesion and assessing hemodynamics and ventricular compensation. Although very sensitive in identifying regurgitant jets, quantitation can be more difficult. Recent guidelines have provided recommendations for quantifying valvular regurgitation (1). Only rarely are cardiac catheterization and angiography necessary to determine the hemodynamic severity of valvular lesions.

Valvular regurgitation is frequently detected in normal subjects, including those without murmurs. With pulsed Doppler echocardiography, TR has been reported in 24% to 96% of normal subjects, MR in 10% to 40%, pulmonic regurgitation in 18% to 92%, and AR in 0% to 33% (2,3). In athletes, the prevalence of valvular regurgitation detected by Doppler is even higher, with at least one regurgitant jet found in greater than 90% of subjects and triple-valve regurgitation in 20% (4). The vast majority of these jets represents a trivial regurgitant volume and is of no clinical significance.

Although considerable information exists concerning the natural history, development of symptoms, importance of LV function, and indications for surgical or catheter-based intervention in patients with valvular heart disease (5), there are few data with regard to the rate of progression of the valvular disease (especially in those exercising vigorously) or factors that influence the rate of progression. The rate of

progression of aortic stenosis is highly variable among individuals and difficult to predict (6). Chronic MR may increase in severity with time but is more likely to do so in patients who experience a new chordal rupture and flail mitral leaflet (7). Little is known about the influence of strenuous exertion on the progression of ventricular dysfunction, especially when that exertion is periodic in nature. When valvular disease coexists with another cardiovascular abnormality, such as arrhythmias or coronary artery disease (CAD), recommendations with regard to eligibility for sports should be based on the most restrictive of these guidelines.

The present recommendations with regard to the permitted level of athletic activity are offered only as guidelines. Physicians with knowledge of an individual athlete, including the severity of the lesion and the physiological and psychological response to competition, may liberalize these recommendations in selected instances. The recommendations are for those athletes who are asymptomatic, as symptoms will prevent vigorous exercise in most subjects and, in most circumstances, symptoms represent an indication for valve replacement or repair (5). In situations in which the history may be unreliable (e.g., when secondary gain in denying symptoms is likely), exercise tolerance testing is useful in confirming normal effort tolerance and suitability for the proposed athletic activity.

MITRAL STENOSIS

The etiology of MS is almost always rheumatic. Most patients with significant MS will be sufficiently symptomatic that participation in competitive sports is not an issue, but patients with mild-to-moderate MS may be asymptomatic even with strenuous exercise. Although MS rarely causes sudden death, exercise (with an increase in heart rate and cardiac output) can cause sudden marked increases in pulmonary capillary and pulmonary artery pressures, at times resulting in sudden acute pulmonary edema (8). Furthermore, the long-term effect of repeated exertion-related increases in pulmonary artery wedge and pulmonary artery pressures on the lungs or right ventricle is unknown. The effect of even periodic strenuous exercise on the likelihood of developing atrial fibrillation is also not known. When atrial fibrillation occurs, even patients with mild MS must be anticoagulated. The aforementioned considerations must be understood by the patient and the family in considering participation in strenuous competitive activity. Another problem associated with MS is systemic embolization, occurring most commonly in the presence of atrial fibrillation, but there is no evidence that this potential complication is provoked by strenuous exercise.

Evaluation. Clues regarding the hemodynamic severity of MS may often be obtained from the history, physical examination, ECG, and chest radiograph, but accurate noninvasive assessment of severity requires two-dimensional and Doppler echocardiography (9). In patients with MS and minimal or no symptoms who wish to engage in competitive sports, exercise stress testing should be performed to at least the level of activity approximating the exercise demands of the sport, particularly when there is a question as to the severity of the MS. In addition, pulmonary artery systolic pressure during exercise can be estimated noninvasively by Doppler echocardiography and may be helpful in making a decision as to how much activity is safe, even if the severity of MS in an individual patient is estimated to be only mild (5). If this is not practical or possible, then right heart catheterization with exercise can be performed to measure the pulmonary capillary wedge pressure and pulmonary artery pressure.

Hemodynamic severity of MS can be categorized as follows: *mild* = mitral valve area greater than 1.5 cm², exercise pulmonary artery wedge pressure less than or equal to 20 mm Hg, or rest pulmonary artery systolic pressure less than 35 mm Hg; *moderate* = mitral valve area 1.0 to 1.5 cm², exercise pulmonary artery wedge pressure less than or equal to 25 mm Hg, or rest pulmonary artery systolic pressure less than or equal to 50 mm Hg; *severe* = mitral valve area less than 1.0 cm², exercise pulmonary artery wedge pressure greater than 25 mm Hg, or rest pulmonary artery systolic pressure greater than 50 mm Hg.

Recommendations:

- 1. Athletes with mild MS (as previously defined) in sinus rhythm with peak pulmonary artery systolic pressure during exercise less than 50 mm Hg can participate in all competitive sports. Athletes with mild MS in atrial fibrillation, or a history of atrial fibrillation, are discussed in recommendation 4 below.**
- 2. Athletes with moderate MS (as previously defined), either in sinus rhythm or in atrial fibrillation, with peak pulmonary artery systolic pressure during exercise less than 50 mm Hg can participate in low and moderate static and low and moderate dynamic competitive sports (classes IA, IB, IIA, and IIB [see Table 1 in Task Force Report 8: Classification of Sports]).**
- 3. Athletes with severe MS in either sinus rhythm or atrial fibrillation or those with peak pulmonary artery pressure greater than 50 mm Hg during exercise should not participate in any competitive sports.**
- 4. Patients with MS of any severity who are in atrial fibrillation or have a history of atrial fibrillation, and who must receive anticoagulation therapy, should not engage in any competitive sports involving the risk for bodily contact (see Task Force Report 8: Classification of Sports) or possible trauma.**

MITRAL REGURGITATION

Mitral regurgitation, unlike MS, has a variety of etiologies, the most common of which is mitral valve prolapse (myxomatous mitral valve). Other common causes are rheumatic heart disease, infective endocarditis, CAD, connective tissue diseases (such as Marfan syndrome), and dilated cardiomyopathy. The recommendations outlined in this section are for patients with primary valvular MR rather than MR secondary to CAD or other conditions causing LV dilation (see Task Force 4: HCM and Other Cardiomyopathies, Mitral Valve Prolapse, Myocarditis, and Marfan Syndrome, and Task Force 6: Coronary Artery Disease).

Evaluation. Mitral regurgitation can be detected by the characteristic physical findings and confirmed by Doppler echocardiography. The severity of the MR is related to the magnitude of the regurgitant volume, which results in LV dilation and increased left atrial pressure. The increased LV diastolic volume enhances total LV stroke volume enough to accommodate the regurgitant volume and to maintain the forward stroke volume within normal limits. The low impedance presented by regurgitation into the left atrium unloads the left ventricle during ventricular systole, such that measures of LV pump function (e.g., ejection fraction) tend to overestimate true myocardial performance (10).

The severity of chronic MR can be adequately judged by noninvasive techniques, principally two-dimensional and Doppler echocardiography. The larger the jet area, and the wider the jet at its origin above the valve, the more severe the regurgitation. The entry of the jet into the atrial appendage or pulmonary vein or systolic reversal of the flow in pulmonary veins are all indicators of severity. Various measures of the severity of MR have been described (1,11). In some patients with eccentric jets or those impinging on the atrial wall, the assessment may be difficult (1). Generally, the LV diastolic volume reflects the severity of chronic MR. *However, it should be noted that the upper limit of normal LV size is increased in the healthy, highly trained athlete. In a series of elite athletes, echocardiographic LV end-diastolic dimensions as high as 66 mm were recorded in women (mean, 48 mm) and up to 70 mm in men (mean, 55 mm) (12). LV end-diastolic dimensions greater than or equal to 55 mm were observed in 45% of participants and greater than 60 mm in 14% of participants. Therefore, assessment of LV enlargement in a highly trained athlete with known or suspected valvular heart disease must take this issue into consideration.* Hence, for purposes of this discussion, an LV end-diastolic dimension greater than 60 mm is considered likely to represent the effects of LV volume overload due to valvular disease and not per se to physiologically based exercise training.

Patients with chronic MR should be followed longitudinally with serial echocardiograms (5). A decrease in ejection fraction and/or increase in end-systolic volume with time is a helpful marker of declining LV function and an indication of having reached the limits of cardiac compensation.

Effects of exercise. In general, exercise produces no significant change or a mild decrease in the regurgitant fraction because of reduced systemic vascular resistance. However, patients with elevation of heart rate (increased systolic ejection time per min) or blood pressure with exercise may manifest marked increases in regurgitant volume and pulmonary capillary pressures. Hence, static exercise that increases arterial pressure is potentially deleterious. Ejection fraction usually does not change or decreases slightly with exercise, although the ejection fraction response may be completely normal in young, asymptomatic subjects.

The etiology of MR may be important in making recommendations concerning heavy physical activity. In patients with MR secondary to previous infective endocarditis or ruptured chordae, the valve tissues theoretically could be further damaged or torn by marked sustained increases in LV systolic pressure.

Recommendations:

- 1. Athletes with mild to moderate MR who are in sinus rhythm with normal LV size and function and with normal pulmonary artery pressures can participate in all competitive sports.**
- 2. Athletes with mild to moderate MR in sinus rhythm with normal LV systolic function at rest and mild LV enlargement (compatible with that which may result solely from athletic training [less than 60 mm {12}]) can participate in some low and moderate static and low, moderate, and high dynamic competitive sports (classes IA, IB, 1C, IIA, IIB, and IIC).**
- 3. Athletes with severe MR and definite LV enlargement (greater than or equal to 60 mm [12]), pulmonary hypertension, or any degree of LV systolic dysfunction at rest should not participate in any competitive sports.**
- 4. Patients in atrial fibrillation or a history of atrial fibrillation who are receiving long-term anticoagulation should not engage in sports involving any risk for bodily contact (see Task Force 8: Classification of Sports) or danger of trauma.**

AORTIC STENOSIS

The diagnosis of AS is established by the characteristic physical findings and two-dimensional and Doppler echocardiography. The three most common etiologies are: 1) rheumatic, 2) congenital, and 3) calcific or degenerative. The majority of young adults with AS participating in competitive athletics have congenital lesions.

Evaluation. Continuous-wave Doppler echocardiography can reliably estimate the severity of AS, especially in the presence of normal cardiac output, which is the case in the great majority of those engaging in competitive sports (13).

Symptoms of dyspnea, syncope, or angina pectoris occur late in the course of AS (14), and the likelihood of sudden death increases significantly with the onset of symptoms.

Because even transient symptoms are so important in marking the onset of increased risk of sudden death, the physician must be aware that dyspnea, near-syncope, and even syncope are likely to be unreported in competitive athletes. Although sudden death is more frequent in symptomatic patients with severe AS, it may also occur in completely asymptomatic patients (15). When doubt persists with regard to the severity of AS after Doppler study, or if a patient with mild-to-moderate AS has symptoms, cardiac catheterization should be performed. Sudden death is rare with mild AS.

Athletes with a history of syncope, even with mild AS, should be carefully evaluated by a cardiologist. This should include assessment of arrhythmias with exercise. Syncope should be regarded as a possible surrogate for spontaneously aborted sudden death and should be thoroughly investigated (see Task Force 7: Arrhythmias).

Severity of AS measured by continuous-wave Doppler echocardiography (or in those instances previously noted by cardiac catheterization) is categorized as follows with respect to the calculated aortic valve area: *mild* = greater than 1.5 cm²; *moderate* = 1.0 to 1.5 cm²; and *severe* = less than or equal to 1.0 cm² (5). This translates roughly (assuming that athletes have normal cardiac output) to the estimated mean aortic valve pressure gradient as follows: *mild* = less than 25 mm Hg; *moderate* = 25 to 40 mm Hg; and *severe* = greater than 40 mm Hg (5).

Because AS is often progressive, periodic re-evaluation at least yearly is necessary and should be performed by a physician with expertise in cardiology. This reassessment includes physical examination and Doppler echocardiography, but may require cardiac catheterization in selected patients as previously noted. In addition, Holter monitoring with intense exercise resembling competition is recommended to detect ventricular arrhythmias in patients with AS who wish to participate in competitive athletics.

In patients with AS, a markedly elevated cardiac output or peripheral vascular resistance for sustained periods of time could result in an exaggerated valvular gradient and a marked increase in LV systolic pressure. Given these precautions, the following recommendations can be made.

Recommendations:

- 1. Athletes with mild AS can participate in all competitive sports, but should undergo serial evaluations of AS severity on at least an annual basis.**
- 2. Athletes with moderate AS can engage in low-intensity competitive sports (class IA). Selected athletes may participate in low and moderate static or low and moderate dynamic competitive sports (classes IA, IB, and IIA) if exercise tolerance testing to at least the level of activity achieved in competition demonstrates satisfactory exercise capacity without symptoms, ST-segment depression or ventricular tachyarrhythmias, and with a normal blood pressure response. Those**

athletes with supraventricular tachycardia or multiple or complex ventricular tachyarrhythmias at rest or with exercise can participate only in low-intensity competitive sports (class IA).

- 3. Patients with severe AS or symptomatic patients with moderate AS should not engage in any competitive sports.**

AORTIC REGURGITATION

Aortic regurgitation has multiple etiologies, as any disease affecting the aortic valve, annulus, or proximal ascending aorta can result in AR. The common etiologies are: 1) congenital bicuspid aortic valve; 2) rheumatic heart disease; 3) infective endocarditis; and 4) aortic root diseases, including Marfan syndrome, ascending aortic aneurysm, aortic dissection, systemic hypertension, and rheumatoid spondylitis. Aortic regurgitation increases LV diastolic volume and stroke volume, which may ultimately lead to LV systolic dysfunction (10). In addition, myocardial oxygen supply/demand imbalance may develop because of the increased wall stress, LV hypertrophy, and reduced diastolic blood pressure (reduced coronary perfusion pressure).

Patients with severe AR may remain asymptomatic and athletic for many years, but angina pectoris, syncope, and ventricular arrhythmias ultimately may appear. Sudden death is rare among asymptomatic patients (less than 0.2% per year [5]) but can occur.

Evaluation. The hemodynamic severity of AR can be assessed noninvasively by physical examination (the severity being reflected by the degree of LV dilation and the peripheral signs of AR), chest radiography, and echocardiography. As noted previously, the upper limit of normal LV end-diastolic size is increased in the healthy, highly trained athlete (12), and this may well affect assessment of LV enlargement in the setting of AR.

Because of the importance of assessing LV function and the size of the aortic root and proximal ascending aorta in determining the etiology of AR, with resulting implications for athletic participation, evaluation by echocardiography is essential. Doppler echocardiography is very sensitive in detecting any degree of AR. Similar to MR, the greatest difficulty arises in differentiating moderate and severe AR. Qualitatively, the width of the regurgitant jet and the proportion of the LV outflow tract occupied by the jet are related to the severity of the AR, as is the slope velocity profile of the diastolic jet. The regurgitant volume can also be measured quantitatively by Doppler methods (1).

The LV function should be assessed serially by two-dimensional echocardiography (5). Radionuclide angiography or cardiac magnetic resonance may be helpful if echocardiograms are of suboptimal quality. Exercise testing can be useful in assessing exercise capacity, especially in those patients having nonspecific or mild symptoms, and it is recommended that testing be performed to at least the level of exertion required by the proposed competitive sport.

Holter monitoring with intense exercise resembling competition is recommended to detect ventricular arrhythmias in patients who wish to participate in competitive athletics.

Effects of exercise. With exercise, regurgitant volume decreases because of the decrease in peripheral vascular resistance that reduces diastolic blood pressure and the decrease in diastolic filling period that accompanies the increase in heart rate (16). Because of these changes in preload and afterload, the failure of the ejection fraction to increase with exercise is of uncertain significance, and there are insufficient data with which to use this finding in formulating recommendations regarding participation in competitive athletics. There are also no data to define whether severe increases in physical activity permanently affect the function of the left ventricle.

For purposes of the following recommendations, hemodynamic severity of AR is graded as follows: *mild* = absent to slight peripheral signs of AR, normal LV size; *moderate* = peripheral signs of AR with mild-to-moderate increases in LV size with normal systolic function; and *severe* = peripheral signs of AR with severe LV enlargement and/or LV systolic dysfunction.

Recommendations:

- 1. Athletes with mild or moderate AR, but with LV end-diastolic size that is normal or only mildly increased, consistent with that which may result solely from athletic training (12), can participate in all competitive sports. In selected instances, athletes with AR and moderate LV enlargement (60 to 65 mm) can engage in low and moderate static and low, moderate, and high dynamic competitive sports (classes IA, IB, 1C, IIA, IIB, and IIC) if exercise tolerance testing to at least the level of activity achieved in competition demonstrates no symptoms or ventricular arrhythmias. Those with asymptomatic nonsustained ventricular tachycardia at rest or with exertion should participate in low-intensity competitive sports only (class IA) (see also Task Force 7: Arrhythmias).**
- 2. Athletes with severe AR and LV diastolic diameter greater than 65 mm as well as those with mild or moderate AR and symptoms (regardless of LV dimension) should not participate in any competitive sports.**
- 3. Those with AR and significant dilation of the proximal ascending aorta (greater than 45 mm) can engage only in low-intensity competitive sports (class IA). These criteria do not apply to athletes with Marfan syndrome and AR, in whom the risks of aortic dissection and rupture are high, and any degree of aortic dilatation would be sufficient to prohibit competitive athletics, as discussed in Task Force 4: HCM and Other Cardiomyopathies, Mitral Valve Prolapse, Myocarditis, and Marfan Syndrome.**

BICUSPID AORTIC VALVES WITH AORTIC ROOT DILATATION

There is growing awareness that many patients with bicuspid aortic valves have disorders of vascular connective tissue, involving loss of elastic tissue, that may result in aortic root dilatation even in the absence of hemodynamically significant AS or AR (17,18). These patients have a risk of aortic dissection; surgery to repair the aorta has been recommended for those patients with greatly enlarged aortic roots (5,18). Recommendations for athletic participation in patients with bicuspid valve disease and associated aortic root dilatation are based on limited data, but with the understanding that aortic dissection can occur in some patients with aortic root diameters less than 50 mm. The recommendations that follow are for patients with bicuspid valves and associated aortic root enlargement. If such patients also have significant AS or AR, these recommendations should be considered in concert with those discussed in the present Task Force for patients with AS or AR. The following recommendations do not pertain to patients with Marfan syndrome, which are discussed in Task Force 4.

Recommendations:

1. Patients with bicuspid aortic valves with no aortic root dilatation (less than 40 mm or the equivalent according to body surface area in children and adolescents) and no significant AS or AR may participate in all competitive sports.
2. Patients with bicuspid aortic valves and dilated aortic roots between 40 and 45 mm may participate in low and moderate static or low and moderate dynamic competitive sports (classes IA, IB, IIA, and IIB), but should avoid any sports in these categories that involve the potential for bodily collision or trauma.
3. Patients with bicuspid aortic valves and dilated aortic roots greater than 45 mm can participate in only low-intensity competitive sports (class IA).

TRICUSPID REGURGITATION

Tricuspid regurgitation is most often secondary to right ventricular (RV) dilation and failure due to pulmonary or RV hypertension. Other etiologies that may affect athletes include rheumatic heart disease, tricuspid valve prolapse, infective endocarditis, congenital anomalies such as Ebstein's anomaly, and the sequelae of surgery for congenital heart disease leaving the patient with RV dilatation. Recommendations here with respect to athletes are for those with primary TR.

Primary TR leads to RV volume overload with the risk of subsequent RV failure as well as increased systemic venous pressure and its consequences. Severity of TR and estimation of right atrial and RV pressures can be determined noninvasively from physical examination, chest radiograph,

and Doppler echocardiography. Occasionally, right-heart catheterization is necessary when these measures cannot be determined noninvasively. There is no evidence that the athlete with isolated primary TR is placed in jeopardy by engaging in heavy physical exertion.

Recommendation:

1. Athletes with primary TR, regardless of severity, with normal RV function in the absence of right atrial pressure greater than 20 mm Hg or elevation of RV systolic pressure can engage in all competitive sports.

TRICUSPID STENOSIS

Tricuspid stenosis is for the most part caused by rheumatic heart disease and is nearly always associated with MS. In that instance, the patient should be judged according to the severity of the MS. Isolated tricuspid stenosis is rare. Such patients should undergo exercise testing at least to the level anticipated in their sport. If asymptomatic, athletes may compete in all competitive activities.

MULTIVALVULAR DISEASE

Multivalvular disease occurs in the context of rheumatic heart disease, myxomatous valvular disease, and infective endocarditis. The lesions can be diagnosed by physical examination and Doppler echocardiography. The relative contribution of each of the lesions may be difficult to assess noninvasively, and cardiac catheterization and angiocardiology are frequently necessary to resolve these distinctions.

Recommendation:

1. Multiple lesions of moderate severity may have additive physiologic effects. In general, athletes with significant multiple valvular disease should not participate in any competitive sports.

PROSTHETIC HEART VALVES

Several general comments apply to all patients who have undergone valve replacement. First, although most patients improve after valve replacement and many become asymptomatic, the long-term mortality after operation is greater than that of a normal population of similar age. Second, a transvalvular gradient of varying severity is present in most patients after valve replacement (5,19). Third, although hemodynamic variables at rest may be essentially normal after valve replacement, many patients have an abnormal response to exercise (20). Finally, after valve replacement (with few exceptions) patients with mechanical prostheses require anticoagulant agents, and those with bioprosthetic valves in sinus rhythm usually do not. In assessing the athlete's capacity for physical activity, exercise stress testing to at least the level of activity performed in the competitive sport is valuable.

Effects of exercise. There are insufficient data to determine whether vigorous repetitive exercise after valve replacement has any long-lasting effect on ventricular or prosthetic valve function. The patient should be made aware of these deficiencies in our knowledge before deciding whether to participate in competitive athletics. Because mechanical and most tissue valves have reduced effective valve areas, they perform best at normal heart rates. Therefore, a sustained heart rate greater than 120 beats/min might result in elevated valve gradients and cardiac outputs that are less than normally expected.

Recommendations:

1. Athletes with a bioprosthetic *mitral valve* not taking anticoagulant agents and who have normal valvular function and normal or near-normal LV function can participate in low and moderate static and low and moderate dynamic competitive sports (classes IA, IB, IIA, and IIB).
2. Athletes with a mechanical or bioprosthetic *aortic valve*, with normal valve function and with normal LV function, can engage in low and moderate static and low and moderate dynamic competitive sports (classes IA, IB, and IIA). Athletes participating in greater than low-intensity competitive sports (class IA) should undergo exercise testing to at least the level of activity achieved in competition to evaluate exercise tolerance and symptomatic and hemodynamic responses.
3. Independent of other considerations, athletes with a mechanical or bioprosthetic *mitral valve* or *aortic valve* who are taking anticoagulant agents should not engage in sports involving the risk of bodily contact (see Task Force 8: Classification of Sports) or the danger of trauma.

VALVE REPAIR OR PERCUTANEOUS MITRAL BALLOON VALVOTOMY

Several general comments apply to athletes who have undergone valve repair or percutaneous mitral balloon mitral valvotomy (PMBV). The benefits of surgical mitral valve repair for MR appear to persist long term, but the effects of strenuous exercise on repaired mitral valves have not been studied systematically. After treatment for MS with PMBV or either closed or open surgical mitral commissurotomy, the patient should have no disability if there has been no injury to the left ventricle or development of significant MR. After aortic valvuloplasty in young patients with congenital AS, there is the risk of subsequent endocarditis, AR, or recurrent AS. Athletes should be evaluated for competitive sports on the basis of their residual hemodynamic state.

Recommendations:

1. For patients with MS who have undergone successful PMBV or surgical commissurotomy, recommendations for participation in competitive sports should be based on the residual severity of the MS or MR, as in patients without operation. Capacity to engage in physical exercise should be evaluated with an exercise tolerance test at least to the level of anticipated activity. Patients with LV dysfunction should be restricted from athletic participation in the same context as those without operation.
2. Athletes who have undergone mitral valve repair for MR should not engage in sports involving the risk or likelihood of bodily contact (see Task Force 8: Classification of Sports) or possible trauma, which might disrupt the repair. They can participate in low-intensity competitive sports (class IA) and, in selected athletes, in low and moderate static and low and moderate dynamic competitive sports (classes IA, IB, and IIA).

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TASK FORCE 3 REFERENCES

1. Zoghbi WA, Enriquez-Sarano M, Foster E, et al. Recommendations for evaluation of the severity of native valvular regurgitation with two-dimensional and Doppler echocardiography. *J Am Soc Echocardiogr* 2003;16:777–802.
2. Kostucki W, Vandenbossche JL, Friart A, Englert M. Pulsed Doppler regurgitant flow patterns of normal valves. *Am J Cardiol* 1986;58:309–13.
3. Richards KL, Cannon SR, Crawford MH, Sorensen SG. Noninvasive diagnosis of aortic and mitral valve disease with pulsed-Doppler spectral analysis. *Am J Cardiol* 1983;51:1122–7.
4. Douglas PS, Berman GO, O'Toole ML, Hiller WD, Reichek N. Prevalence of multivalvular regurgitation in athletes. *Am J Cardiol* 1989;64:209–12.
5. Bonow RO, Carabello B, de Leon AC, et al. ACC/AHA guidelines for the management of patients with valvular heart disease: a report of the American College of Cardiology/American Heart Association Task Force on Practice Guidelines (Committee on Management of Patients With Valvular Heart Disease). *J Am Coll Cardiol* 1998;32:1486–582.
6. Otto CM, Burwash IG, Legget ME, et al. Prospective study of asymptomatic valvular aortic stenosis: clinical, echocardiographic, and exercise predictors of outcome. *Circulation* 1997;95:2262–70.
7. Enriquez-Sarano M, Basmadjian AJ, Rossi A, Bailey KR, Seward JB, Tajik AJ. Progression of mitral regurgitation: a prospective Doppler echocardiographic study. *J Am Coll Cardiol* 1999;34:1137–44.
8. Rahimtoola SH, Durairaj A, Mehra A, Nuno I. Current evaluation and management of patients with mitral stenosis. *Circulation* 2002;106:1183–8.
9. Faletta F, Pezzano A J, Fusco R, et al. Measurement of mitral valve area in mitral stenosis: four echocardiographic methods compared with direct measurement of anatomic orifices. *J Am Coll Cardiol* 1996;28:1190–7.
10. Carabello BA. Progress in mitral and aortic regurgitation. *Curr Probl Cardiol* 2003;28:549–84.
11. Dujardin KS, Enriquez-Sarano M, Bailey KR, Nishimura RA, Seward JB, Tajik AJ. Grading of mitral regurgitation by quantitative Doppler echocardiography: calibration by left ventricular angiography in routine clinical practice. *Circulation* 1997;96:3409–15.
12. Pelliccia A, Culasso F, Di Paolo FM, Maron BJ. Physiologic left ventricular cavity dilatation in elite athletes. *Ann Intern Med* 1999;130:23–31.
13. Galan A, Zoghbi WA, Quinones MA. Determination of severity of valvular aortic stenosis by Doppler echocardiography and relation of findings to clinical outcome and agreement with hemodynamic mea-

- surements determined at cardiac catheterization. *Am J Cardiol* 1991;67:1007-12.
14. Carabello BA. Evaluation and management of patients with aortic stenosis. *Circulation* 2002;105:1746-50.
 15. Rosenhek R, Binder T, Porenta G, et al. Predictors of outcome in severe, asymptomatic aortic stenosis. *N Engl J Med* 2000;343:611-7.
 16. Dehmer GJ, Firth BG, Hillis LD, et al. Alterations in left ventricular volumes and ejection fraction at rest and during exercise in patients with aortic regurgitation. *Am J Cardiol* 1981;48:17-27.
 17. Nistri S, Sorbo MD, Marin M, Scognamiglio R, Thiene G. Aortic root dilatation in young men with normally functioning bicuspid aortic valves. *Heart* 1999;82:19-22.
 18. Svensson LG, Kim KH, Lytle BW, Cosgrove DM. Relationship of aortic cross-sectional area to height ratio and the risk of aortic dissection in patients with bicuspid aortic valves. *J Thorac Cardiovasc Surg* 2003;126:892-3.
 19. Rahimtoola SH. Choice of prosthetic heart valve for adult patients. *J Am Coll Cardiol* 2003;41:893-904.
 20. Borer JS, Herrold EM, Hochreiter C, et al. Natural history of left ventricular performance at rest and during exercise after aortic valve replacement for aortic regurgitation. *Circulation* 1991;84:III133-9.

Appendix 1

The authors of this report declared they have no relationships with industry pertinent to this topic.

Task Force 4: HCM and Other Cardiomyopathies, Mitral Valve Prolapse, Myocarditis, and Marfan Syndrome

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HYPERTROPHIC CARDIOMYOPATHY

General considerations. Hypertrophic cardiomyopathy (HCM) is a relatively common form of genetic heart disease (0.2%; 1:500 in the general population) (1), and the most common cause of sudden unexpected cardiac death in young people, including competitive athletes (2). Sudden death may occur at any age, but is most common in individuals 30 years of age or less. At present, 12 mutant genes (most encoding sarcomeric proteins) and over 400 specific mutations in these genes have been implicated in the pathogenesis of clinically diagnosed HCM (3).

The disease is characterized by heterogeneous presentation and natural history in which the most consistent diagnostic feature demonstrated by echocardiography is otherwise unexplained and usually asymmetric hypertrophy associated with a non-dilated left ventricle (LV) (3-5). Clinical diagnosis of HCM is made by recognition of the disease phenotype with LV hypertrophy (3,4). In this regard, a maximal LV end-diastolic wall thickness of 15 mm or more (or on occasion, 13 or 14 mm) is the absolute dimension generally accepted for the clinical diagnosis of HCM in an adult athlete (in children, 2 or more standard deviations from the mean relative to body surface are; z-score of 2 or more); however, any LV wall thickness (including normal) is theoretically compatible with the presence of a mutant HCM gene (3,4). Of note, individuals of virtually any age (but usually less than 14 years old) harboring a HCM-causing mutant gene may not manifest LV hypertrophy (3,4).

In a disease such as HCM, extrapolation of risk level

from non-athletes to highly trained competitive athletes is tenuous. This relates to the unstable electrophysiologic substrate and propensity for potentially lethal ventricular tachyarrhythmias in HCM, interacting with the physiologic stresses inherent in athletic training and competition (i.e., alterations in blood volume, hydration, and electrolytes). Furthermore, no single clinical, morphologic, or electrophysiologic factor has emerged as the reliable predictor of risk in HCM (3,4). Therefore, because the panel could not precisely stratify sudden death risk specifically for all athletes with HCM, the present recommendations for sports eligibility remain conservative and homogeneous for those athletes within the diverse HCM clinical spectrum.

Given the inability to precisely stratify risk on clinical grounds in individual young patients with HCM, a broad recommendation to exclude such individuals from competitive sports will, by definition, deny participation to some unnecessarily. However, given the frequency with which HCM is associated with sudden death in young athletes (2), and recent data showing that athletic activity per se is associated with higher risk in those with underlying cardiovascular abnormalities (6), the present recommendations are viewed as prudent. That is, the goal is to encompass all preventable sudden deaths in young persons with HCM, while acknowledging that other athletes who may not be destined for sudden death will also be subjected to the same recommendations.

Preclinical diagnosis. With the availability of preclinical genetic diagnosis, a relatively small number of youthful family members have been identified as affected by a HCM-causing mutant gene solely on the basis of laboratory

DNA analysis, and in the absence of typical morphologic (phenotypic) features of the disease (3,4). As family genetic screening for HCM becomes more widespread, clinicians may be increasingly faced with the dilemma of making recommendations regarding sports participation for subjects who have only preclinical evidence of HCM (i.e., genotype positive-phenotype negative). Nonetheless, it is likely that most such individuals are destined to ultimately develop the HCM phenotype with the attendant possibility of a potentially unstable electrophysiologic milieu. Moreover, the HCM phenotype (i.e., LV hypertrophy) may develop over the course of several years (3,4) after initial evaluation, when competitive sports participation could still be an ongoing and important lifestyle issue.

Based on these considerations, prudent recommendations for athletes with preclinical HCM would include, on a 12- to 18-month basis, in addition to serial two-dimensional echocardiography: 12-lead ECG and ambulatory Holter electrocardiogram (ECG), and less frequently cardiac magnetic resonance (CMR) imaging and exercise stress testing to a level similar to that expected in the sport under consideration (for evaluation of exercise tolerance, blood pressure, and ventricular tachyarrhythmias). If all of these parameters continue to be normal, then based on the level of present knowledge, restriction from competitive athletic activities is not recommended. Such systematic follow-up of this subgroup is strongly recommended, particularly if there is a family history of HCM and sudden cardiac death.

Relevant to these considerations are the prior observations that abnormalities on 12-lead ECG and preload-independent measures of diastolic dysfunction with tissue Doppler ultrasonography may precede the appearance of LV hypertrophy, providing clues to impending development of wall thickening (7–9). Athletes with abnormal 12-lead ECG and absence of LV hypertrophy on two-dimensional echocardiogram (particularly if relatives in HCM families) should be afforded a high index of suspicion for HCM and undergo CMR imaging to determine whether areas of segmental hypertrophy undetected by echocardiography are present in regions of the LV chamber such as anterolateral free wall or apex (7). However, 12-lead ECG abnormalities in family members without LV hypertrophy (particularly if relatively minor or nonspecific) should not per se be regarded as evidence of HCM.

Recommendations:

- 1. Athletes with a probable or unequivocal clinical diagnosis of HCM should be excluded from most competitive sports, with the possible exception of those of low intensity (class IA). This recommendation is independent of age, gender, and phenotypic appearance, and does not differ for those athletes with or without symptoms, LV outflow obstruction, or prior treatment with drugs or major interventions with surgery, alcohol septal ablation, pacemaker, or implantable defibrillator.**

- 2. Although the clinical significance and natural history of genotype positive-phenotype negative individuals remains unresolved, no compelling data are available at present with which to preclude these patients from competitive sports, particularly in the absence of cardiac symptoms or a family history of sudden death.**

Given the effectiveness of implantable cardioverter-defibrillators (ICDs) in preventing sudden death in HCM (10), clinicians will increasingly be faced with decisions regarding athletic participation for HCM patients with ICDs. Although effective for sudden death prevention in observational studies (10), the unique physiologic milieu associated with competitive athletic activities, including intravascular volume and electrolyte disturbances, neurohormonal activity, and the potential for myocardial ischemia make the absolute reliability of ICDs in such settings unpredictable. Furthermore, there is a possibility for device malfunction and the risk for traumatic injury to the athlete-patient (or other competitors) should the ICD discharge either appropriately or inappropriately. Thus, the placement of an ICD in an HCM patient does not change the competitive sports recommendations for this disease (as previously noted), namely, that restriction from participation in contact and most non-contact sports is advisable (11); such individuals may engage only in low-intensity competitive sports (class IA).

The presence of a free-standing automated external defibrillator (AED) at sporting events should not be considered either absolute protection against sudden death, a prospectively designed treatment strategy for known cardiovascular disease, nor a justification for participation in competitive sports in athletes with previously diagnosed HCM. Athletes with HCM using drugs such as anabolic steroids or energy stimulant drinks may in fact increase their risk of arrhythmias, although definitive data are lacking.

MITRAL VALVE PROLAPSE (MVP)

Mitral valve prolapse (i.e., myxomatous degeneration) is of particular importance in the evaluation of athletes, given its relatively high prevalence in the general population (estimated, 2% to 3%) (12–14). The condition is defined by echocardiography as systolic displacement of one or both mitral leaflets into the left atrium beyond the plane of the mitral annulus in the parasternal long-axis view (13). These patients may be identified by auscultation with a mid-systolic click and/or murmur of mitral regurgitation. Additionally, MVP is characterized by a mostly favorable prognosis and low event rate (12–15). In general, the greatest risks for unfavorable clinical sequelae—which include severe progressive mitral regurgitation requiring valve surgery, infective endocarditis, embolic events, atrial and ventricular tachyarrhythmias, and sudden death appear to be associated with substantial structural abnormality of the mitral valve (i.e., “classic” MVP) with diffuse leaflet thickening, elonga-

tion, and redundancy, and in some cases ruptured chordae tendineae (12–15).

Sudden cardiac death due to isolated MVP is rare among young patients, particularly in relation to exercise and/or in trained athletes (12–16). Such events are probably not more frequent than in the general population and occur predominantly in patients older than 50 years with severe mitral regurgitation and/or systolic dysfunction (15).

Some individuals with MVP appear to be part of a connective tissue phenotypic spectrum with tall, thin habitus, thoracic cage deformity, and joint hypermobility (i.e., MASS phenotype), for which there is a risk, albeit low, for progression to aortic dilatation or sudden death (17).

Recommendations:

1. **Athletes with MVP—but without any of the following features—can engage in all competitive sports:**
 - a. **prior syncope, judged probably to be arrhythmogenic in origin**
 - b. **sustained or repetitive and nonsustained supraventricular tachycardia or frequent and/or complex ventricular tachyarrhythmias on ambulatory Holter monitoring**
 - c. **severe mitral regurgitation assessed with color-flow imaging**
 - d. **LV systolic dysfunction (ejection fraction less than 50%)**
 - e. **prior embolic event**
 - f. **family history of MVP-related sudden death**
2. **Athletes with MVP and any of the aforementioned disease features can participate in low-intensity competitive sports only (class IA).**

Recommendations related to hemodynamic burden secondary to moderate–severe mitral regurgitation (as assessed by physical examination and two-dimensional echocardiogram and Doppler study) in athletes with MVP appear in Task Force 3.

MYOCARDITIS

Myocarditis is an inflammatory disease of myocardium and a cause of sudden death in young athletes (2,18–24). It is usually of infectious etiology due to a variety of viral agents, most commonly enterovirus (e.g., Coxsackie virus), adenovirus, or parvovirus in young people, but also by drugs and toxic agents such as cocaine (22–25). Myocarditis evolves through active, healing, and healed pathologic stages—characterized progressively by inflammatory cell infiltrates leading to interstitial edema and focal myocyte necrosis and replacement fibrosis (20)—which potentially create an electrically unstable substrate for development of ventricular tachyarrhythmias (2,18,24). In some instances, viral myocarditis can culminate in dilated cardiomyopathy with LV systolic dysfunction, presumably as a consequence of viral-mediated immunologic damage to the myocardium or cytoskeletal disruption (22–25).

Myocarditis can be diagnosed by established histopathologic, histochemical, or molecular criteria (20,23–25), but is challenging to identify clinically. Suspicion may be raised by chest pain, exertional dyspnea, fatigue, syncope, palpitations, ventricular tachyarrhythmias and conduction abnormalities or by acute congestive heart failure associated with LV dilatation and/or segmental systolic dysfunction, cardiogenic shock, or ST-T changes on ECG (22,24).

When clinical judgment suggests the presence of myocarditis, an endomyocardial biopsy may clarify an otherwise ambiguous clinical profile. Because of patchy distribution of inflammatory cells, biopsies are often insensitive and frequently yield false-negative histologic results (20,22,24). However, the diagnostic yield of histology can be enhanced by molecular analysis with PCR amplification of the viral genome (23,25).

Recommendations:

1. **Athletes with probable or definite evidence of myocarditis should be withdrawn from all competitive sports and undergo a prudent convalescent period of about six months following the onset of clinical manifestations.**
2. **Athletes may return to training and competition after this period of time if:**
 - a. **LV function, wall motion, and cardiac dimensions return to normal (based on echocardiographic and/or radionuclide studies at rest and with exercise)**
 - b. **clinically relevant arrhythmias such as frequent and/or complex repetitive forms of ventricular or supraventricular ectopic activity are absent on ambulatory Holter monitoring and graded exercise testing**
 - c. **serum markers of inflammation and heart failure have normalized**
 - d. **the 12-lead ECG has normalized. Persistence of relatively minor ECG alterations such as some ST-T changes are not, per se, the basis for restriction from competition.**

MARFAN SYNDROME

Marfan syndrome (and related disorders), caused by more than 400 individual mutations in the gene encoding fibrillin-1 (FBN1), is an autosomal dominant disorder of connective tissue with estimated prevalence of 1:5,000 to 1:10,000 in the general population (26–29). It is characterized clinically by a diverse constellation of abnormalities variable in severity and involving primarily the ocular, skeletal, and cardiovascular organ systems (28–30). Diagnosis is made according to the Gent nosology if major criteria are present in two organ systems and a third is involved, or when there is a family history of Marfan syndrome (28,29). Skeletal abnormalities include arm span-to-height ratio greater than 1.05, tall stature, arachnodactyly, dolichostenomelia (long, thin limbs), hyperextensibility

and ligamentous laxity, scoliosis, and chest wall deformity (pectus excavatum or carinatum), in addition to ectopia lentis (lens dislocation).

Cardiovascular manifestations that impact on natural history are: 1) progressive dilatation of the aortic root or descending aorta, which predisposes to dissection and rupture (30-33); and 2) MVP with associated mitral regurgitation or LV systolic dysfunction, which may occasionally predispose to ventricular tachyarrhythmias and sudden death (33). The risk for aortic rupture is usually linked to marked enlargement of the aorta (transverse dimension greater than 50 mm), although dissection can occur with a normal (or near-normal) aortic root dimension (30-36) (A. DePaepe, personal communication, April 12, 2004). Weightlifting has been specifically associated with aortic dissection in athletes with cystic medial necrosis (with or without systemic hypertension or use of anabolic steroids) (35,36). The incidence of aortic dissection appears to have decreased with earlier prophylactic surgical aortic root reconstruction and beta-blocker therapy (30,32,34).

Ascending aortic root dilatation (and dissection) has also been associated with congenital bicuspid aortic valve disproportionate to that attributable to abnormal valvular function (37-41), as well as familial aortic aneurysm and dissection, independent of Marfan syndrome (41-43).

Recommendations:

1. **Athletes with Marfan syndrome can participate in low and moderate static/low dynamic competitive sports (classes IA and IIA) if they do not have one or more of the following:**
 - a. **aortic root dilatation (i.e., transverse dimension 40 mm or greater in adults, or more than 2 standard deviations from the mean for body surface area in children and adolescents; z-score of 2 or more)**
 - b. **moderate-to-severe mitral regurgitation**
 - c. **family history of dissection or sudden death in a Marfan relative**

It is recommended, however, that these athletes have an echocardiographic measurement of aortic root dimension repeated every six months, for close surveillance of aortic enlargement.
2. **Athletes with unequivocal aortic root dilatation (transverse dimension 40 mm or greater in adults or greater than 2 standard deviations beyond the mean for body surface area in children and adolescents; z-score of 2 or more) (41,43), prior surgical aortic root reconstruction, chronic dissection of aorta or other artery, moderate-to-severe mitral regurgitation, or family history of dissection or sudden death can participate only in low-intensity competitive sports (class IA).**
3. **Athletes with Marfan syndrome, familial aortic aneurysm or dissection, or congenital bicuspid aortic valve with any degree of ascending aortic enlargement (as defined in 1 and 2 above) also should not participate**

in sports that involve the potential for bodily collision.

4. **Recommendations related to aortic regurgitation are the same as those in Task Force 3.**

These recommendations are offered independent of whether beta-blockers are administered to mitigate aortic root enlargement.

EHLERS-DANLOS SYNDROME

The vascular form of Ehlers-Danlos syndrome carries a substantial risk of rupture of the aorta and its major branches (28). This is a rare autosomal dominant disorder, caused by a defect in type III collagen, encoded by the COL3A1 gene. Patients have variable joint hypermobility, susceptibility to bruising, difficult wound healing, and often prematurely aged appearance.

Recommendation:

1. **Individuals with the vascular form of Ehlers-Danlos syndrome should not engage in any competitive athletic activity.**

ARRHYTHMOGENIC RIGHT VENTRICULAR CARDIOMYOPATHY (ARVC)

Cited as a major cause of sudden death in young people and athletes (44-46), particularly in the northeastern (Veneto) region of Italy (45) but seemingly less common in the U.S (2,18), ARVC is characterized by a broad phenotypic spectrum and characteristically by loss of myocytes in the right ventricular myocardium with fatty or fibro-fatty replacement resulting in segmental or diffuse wall thinning. It is frequently associated with myocarditis (44,45,47). Clinical diagnosis is challenging, but relies largely on familial occurrence, ventricular tachyarrhythmias (particularly ventricular tachycardia of right ventricular origin elicited by exercise), T-wave inversion in precordial leads V₁ through V₃ and epsilon waves on ECG, or right ventricular dilatation and/or segmental wall motion abnormalities, aneurysm formation, and fatty deposition in the right ventricular wall identified with echocardiography, multi-slice computed tomography, or cardiac magnetic resonance imaging.

Recommendation:

1. **Athletes with probable or definite diagnosis of ARVC should be excluded from most competitive sports, with the possible exception of those of low intensity (class IA).**

OTHER MYOCARDIAL DISEASES

A number of other uncommon diseases of the myocardium deserve consideration as potential causes of sudden death in athletes. These include dilated cardiomyopathy (due to a variety of etiologies including genetic); primary non-hypertrophied restrictive cardiomyopathy, systemic infiltra-

tive diseases with secondary cardiac involvement such as sarcoidosis, and also isolated non-compaction of LV myocardium with or without systolic dysfunction (48,49). Few data are presently available regarding the relative risks of athletic training and competition in athletes with the aforementioned myocardial diseases.

Recommendation:

1. **Until more information is available in this regard, it is most prudent to exclude athletes with these diseases from most competitive sports, with the possible exception of those of low intensity (class IA) in selected cases.**

PERICARDITIS

Recommendation:

1. **Athletes with pericarditis, regardless of etiology, should not participate in competitive sports during the acute phase. Such athletes can return to full activity when there is no longer evidence of active disease, including effusion by echocardiography, and when serum markers of inflammation have normalized. For pericarditis associated with evidence of myocardial involvement, eligibility recommendations should also be based on the course of myocarditis. Chronic pericardial disease that results in constriction disqualifies one from all competitive sports.**

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TASK FORCE 4 REFERENCES

1. Maron BJ, Gardin JM, Flack JM, Gidding SS, Kurosaki TT, Bild DE. Prevalence of hypertrophic cardiomyopathy in a general population of young adults: echocardiographic analysis of 4,111 subjects in the CARDIA study: Coronary Artery Risk Development In (young) Adults. *Circulation* 1995;92:785-9.
2. Maron BJ. Sudden death in young athletes. *N Engl J Med* 2003;349:1064-75.
3. Maron BJ, McKenna WJ, Danielson GK, et al. American College of Cardiology/European Society of Cardiology clinical expert consensus document on hypertrophic cardiomyopathy: a report of the American College of Cardiology Foundation Task Force on Clinical Expert Consensus Documents and the European Society of Cardiology Committee for Practice Guidelines. *J Am Coll Cardiol* 2003;42:1687-713.
4. Maron BJ. Hypertrophic cardiomyopathy: a systematic review. *JAMA* 2002;287:1308-20.
5. Wigle ED, Rakowski H, Kimball BP, Williams WG. Hypertrophic cardiomyopathy: clinical spectrum and treatment. *Circulation* 1995; 92:1680-92.
6. Corrado D, Basso C, Rizzoli G, Schiavon M, Thiene G. Does sports activity enhance the risk of sudden death in adolescents and young adults? *J Am Coll Cardiol* 2003;42:1959-63.
7. Rosenzweig A, Watkins H, Hwang DS, et al. Preclinical diagnosis of familial hypertrophic cardiomyopathy by genetic analysis of blood lymphocytes. *N Engl J Med* 1991;325:1753-60.
8. Panza JA, Maron BJ. Relation of electrocardiographic abnormalities to evolving left ventricular hypertrophy in hypertrophic cardiomyopathy. *Am J Cardiol* 1989;63:1258-65.
9. Ho CY, Sweitzer NK, McDonough B, et al. Assessment of diastolic function with Doppler tissue imaging to predict genotype in preclinical hypertrophic cardiomyopathy. *Circulation* 2002;105:2992-7.
10. Maron BJ, Shen W-K, Link MS, et al. Efficacy of implantable cardioverter-defibrillators for the prevention of sudden death in patients with hypertrophic cardiomyopathy. *N Engl J Med* 2000;342: 365-73.
11. Maron BJ, Mitten MJ, Quandt EF, Zipes DP. Competitive athletes with cardiovascular disease—the case of Nicholas Knapp. *N Engl J Med* 1998;339:1632-5.
12. Nishimura RA, McGoon MD, Shub C, Miller FA Jr., Ilstrup DM, Tajik AJ. Echocardiographically documented mitral-valve prolapse: long-term follow-up of 237 patients. *N Engl J Med* 1985;313:1305-9.
13. Marks AR, Choong CY, Sanfilippo AJ, Ferre M, Weyman AE. Identification of high-risk and low-risk subgroups of patients with mitral-valve prolapse. *N Engl J Med* 1989;320:1031-6.
14. Freed LA, Levy D, Levine RA, et al. Prevalence and clinical outcome of mitral-valve prolapse. *N Engl J Med* 1999;341:1-7.
15. Avierinos JF, Gersh BJ, Melton LJ III, et al. Natural history of asymptomatic mitral valve prolapse in the community. *Circulation* 2002;106:1355-61.
16. Corrado D, Basso C, Nava A, Rossi L, Thiene G. Sudden death in young people with apparently isolated mitral valve prolapse. *G Ital Cardiol* 1997;27:1097-105.
17. Glesby MJ, Pyeritz RE. Association of mitral valve prolapse and systemic abnormalities of connective tissue: a phenotypic continuum. *JAMA* 1989;262:523-8.
18. Maron BJ, Shirani J, Poliac LC, Mathenge R, Roberts WC, Mueller FO. Sudden death in young competitive athletes: clinical, demographic, and pathological profiles. *JAMA* 1996;276:199-204.
19. Phillips M, Robinowitz M, Higgins JR, Boran KJ, Reed T, Virmani R. Sudden cardiac death in Air Force recruits: a 20-year review. *JAMA* 1986;256:2696-9.
20. Aretz HT, Billingham ME, Edwards WD, et al. Myocarditis: a histopathologic definition and classification. *Am J Cardiovasc Pathol* 1987;1:3-14.
21. Karjalainen J, Heikkila J. Incidence of three presentations of acute myocarditis in young men in military service: a 20-year experience. *Eur Heart J* 1999;20:1120-5.
22. Lieberman EB, Hutchins GM, Herskowitz A, Rose NR, Baughman KL. Clinicopathologic description of myocarditis. *J Am Coll Cardiol* 1991;18:1617-26.
23. Bowles NE, Ni J, Kearney DL, et al. Detection of viruses in myocardial tissues by polymerase chain reaction. Evidence of adenovirus as a common cause of myocarditis in children and adults. *J Am Coll Cardiol* 2003;42:466-72.
24. Towbin JA. Myocarditis. In: Finberg L, Kleinman R, editors. *Saunders Manual of Pediatric Practice*. Philadelphia, PA: WB Saunders, 2002:660-3.
25. Martin AB, Webber S, Fricker FJ, et al. Acute myocarditis: rapid diagnosis by PCR in children. *Circulation* 1994;90:330-9.
26. Dietz HC, Cutting GR, Pyeritz RE, et al. Marfan syndrome caused by a recurrent de novo missense mutation in the fibrillin gene. *Nature* 1991;352:337-9.
27. Robinson PN, Godfrey M. The molecular genetics of Marfan syndrome and related microfibrilopathies. *J Med Genet* 2000;37:9-25.
28. Pyeritz RE. Marfan syndrome and other disorders of fibrillin. In: Rimoin DL, Conner JM, Pyeritz RE, Korf B, editors. *Principles and Practice of Medical Genetics*. 4th edition. Edinburgh: Churchill Livingstone, 2002:3977-4020.
29. De Paepe A, Devereux RB, Dietz HC, Hennekam RC, Pyeritz RE. Revised diagnostic criteria for the Marfan syndrome. *Am J Med Genet* 1996;62:417-26.
30. Januzzi JL, Isselbacher EM, Fattori R, et al. Characterizing the young patient with aortic dissection: results from the International Registry of Aortic Dissection (IRAD). *J Am Coll Cardiol* 2004;43:665-9.
31. Marsalese DL, Moodie DS, Vacante M, et al. Marfan's syndrome: natural history and long-term follow-up of cardiovascular involvement. *J Am Coll Cardiol* 1989;14:422-8.
32. Shores J, Berger KR, Murphy EA, Pyeritz RE. Progression of aortic dilatation and the benefit of long-term beta-adrenergic blockade in Marfan's syndrome. *N Engl J Med* 1994;330:1335-41.
33. Yetman AT, Bornemeier RA, McCrindle BW. Long-term outcome in patients with Marfan syndrome: is aortic dissection the only cause of sudden death? *J Am Coll Cardiol* 2003;41:329-32.
34. Gott VL, Greene PS, Alejo DE, et al. Replacement of the aortic root in patients with Marfan's syndrome. *N Engl J Med* 1999;340:1307-13.

35. de Virgilio C, Nelson RJ, Milliken J, et al. Ascending aortic dissection in weight lifters with cystic medial degeneration. *Ann Thorac Surg* 1990;49:638-42.
36. Elefteriades JA, Hatzaras I, Tranquilli MA, et al. Weight lifting and rupture of silent aortic aneurysms. *JAMA* 2003;290:2803.
37. Ferencik M, Pape LA. Changes in size of ascending aorta and aortic valve function with time in patients with congenitally bicuspid aortic valves. *Am J Cardiol* 2003;92:43-6.
38. Hahn RT, Roman MJ, Mogtader AH, Devereux RB. Association of aortic dilation with regurgitant, stenotic and functionally normal bicuspid aortic valves. *J Am Coll Cardiol* 1992;19:283-8.
39. Roberts CS, Roberts WC. Dissection of the aorta associated with congenital malformation of the aortic valve. *J Am Coll Cardiol* 1991;17:712-6.
40. Nistri S, Sorbo MD, Marin M, Palisi M, Scognamiglio R, Thiene G. Aortic root dilation in young men with normally functioning bicuspid aortic valves. *Heart* 1999;82:19-22.
41. Cripe L, Andelfinger G, Martin LJ, Shooner K, Benson DW. Bicuspid aortic valve is heritable. *J Am Coll Cardiol* 2004;44:138-43.
42. Lewin MB, McBride KL, Pignatelli R, et al. Echocardiographic evaluation of asymptomatic parental and sibling cardiovascular anomalies associated with congenital left ventricular outflow tract lesions. *Pediatrics* 2004;114:691-96.
43. Roman MJ, Devereux RB, Kramer-Fox R, O'Loughlin J. Two-dimensional echocardiographic aortic root dimensions in normal children and adults. *Am J Cardiol* 1989;64:507-12.
44. Tabib A, Loire R, Chalabreysse L, et al. Circumstances of death and gross and microscopic observations in a series of 200 cases of sudden death associated with arrhythmogenic right ventricular cardiomyopathy and/or dysplasia. *Circulation* 2003;108:3000-5.
45. Corrado D, Basso C, Thiene G, et al. Spectrum of clinicopathologic manifestations of arrhythmogenic right ventricular cardiomyopathy/dysplasia: a multicenter study. *J Am Coll Cardiol* 1997;30:1512-20.
46. Corrado D, Leoni L, Link MS, et al. Implantable cardioverter-defibrillator therapy for prevention of sudden death in patients with arrhythmogenic right ventricular cardiomyopathy/dysplasia. *Circulation* 2003;108:3084-91.
47. Bowles NE, Ni J, Marcus F, Towbin JA. The detection of cardiotropic viruses in the myocardium of patients with arrhythmogenic right ventricular dysplasia/cardiomyopathy. *J Am Coll Cardiol* 2002;39:892-5.
48. Oechslin EN, Attenhofer Jost CH, Rojas JR, Kaufmann PA, Jenni R. Long-term follow-up of 34 adults with isolated left ventricular noncompaction: a distinct cardiomyopathy with poor prognosis. *J Am Coll Cardiol* 2000;36:493-500.
49. Pignatelli RH, McMahon CJ, Dreyer WJ, et al. Clinical characterization of left ventricular noncompaction in children: a relatively common form of cardiomyopathy. *Circulation* 2003;108:2672-8.

Appendix 1. Author Relationships With Industry and Others

Name	Consultant	Research Grant	Scientific Advisory Board	Stock Holder	Expert Witness Testimony
Dr. Michael J. Ackerman	<ul style="list-style-type: none"> • CV Therapeutics • Genaisance Pharmaceuticals • Medtronic • Pfizer 	None	<ul style="list-style-type: none"> • Genaisance Pharmaceuticals 	<ul style="list-style-type: none"> • Genaisance Pharmaceuticals 	None
Dr. Barry J. Maron	None	<ul style="list-style-type: none"> • Medtronic 	None	None	<ul style="list-style-type: none"> • 1996, Defense, Northwestern vs. Knapp
Dr. Rick A. Nishimura	None	None	None	None	None
Dr. Reed E. Pyeritz	<ul style="list-style-type: none"> • Genzyme 	<ul style="list-style-type: none"> • Genzyme 	None	None	None
Dr. Jeffrey A. Towbin	None	None	None	None	<ul style="list-style-type: none"> • 2005, Defense, Myocarditis, sudden death in child • 2004, Defense, Myocarditis, sudden death in child • 2004, Defense, Myocarditis, sudden death in child • 2002, Defense, Myocarditis, sudden death in child • 2000, Defense, Myocarditis, sudden death in child • 1996, Plaintiff, Myocarditis, sudden death in professional athlete • 1995, Plaintiff, Myocarditis, sudden death in professional athlete
Dr. James E. Udelson	None	None	None	None	None

Task Force 5: Systemic Hypertension

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Samuel S. Gidding, MD, FACC, Thomas G. Pickering, MD, DPHIL, Jackson T. Wright, JR, MD, PHD

GENERAL CONSIDERATIONS

Systemic hypertension is the most common cardiovascular condition observed in competitive athletes. The diagnosis of hypertension is based on the presence of blood pressure (BP) persistently at or above certain levels as measured by routine sphygmomanometry on at least two separate occasions. A level above 140/90 mm Hg is considered to be hypertensive in people over age 18 years (1). In children and adolescents, hypertension is defined as average systolic or diastolic levels greater than or equal to the 95th percentile for gender, age, and height (2). In determining the level of competitive athletic activity that a hypertensive person may assume, it is also important to ascertain the degree of hypertension-related target organ damage. Although hypertension may be associated with an increased risk for complex ventricular arrhythmias and sudden death, this disease by itself has not been incriminated as a cause of sudden cardiac death in young competitive athletes (3). For the general population, increased levels of noncompetitive physical activity are regarded not only as beneficial by reducing BP (4) and the incidence of hypertension (5), but also protecting against stroke (6). In view of the striking increase in obesity-induced hypertension among children and adolescents related in part to physical inactivity, all people should be encouraged to engage in regular exercise. Those who are hypertensive derive protection from all-cause and cardiovascular mortality by maintaining higher levels of cardiorespiratory fitness (7).

Assessment of blood pressure. Blood pressure should be accurately measured in all individuals who wish to participate in competitive athletics before they begin training. Blood pressure should be measured by routine sphygmomanometry, using the guidelines listed in Table 1 (8). There are often "white-coat" elevations induced by anxiety related to the examination, particularly in young people concerned about the potential consequences of the examination. Therefore, additional BP recordings outside the office should be obtained in those with elevated office readings, either with readily available and inexpensive home self-recorders or with less accessible and more expensive automatic ambulatory monitors.

Evaluation. Those individuals with any degree of persistent hypertension should have a thorough history and physical examination and limited laboratory testing to evaluate secondary causes and to ascertain target organ damage (1). If agents that may raise BP such as non-steroidal anti-inflammatory drugs (NSAIDs) are being taken, additional measurements should be obtained after they have been discontinued. Laboratory testing for most

subjects with stage 1 hypertension (140 to 159 mm Hg/90 to 99 mm Hg) should include an automated blood chemistry (glucose, creatinine, or the corresponding estimated glomerular filtration rate, electrolytes, lipid profile), hematocrit, urine analysis, and an electrocardiogram. If hypertension is stage 2 (greater than or equal to 160/100 mm Hg), if results of the initial laboratory tests are abnormal, or if features suggestive of identifiable (secondary) causes are noted by history or physical examination, the patient should be referred for additional study (including echocardiography) and therapy.

Effects of exercise on BP. As noted during stress testing, systolic BP rises during aerobic (dynamic) exercise. Both systolic and diastolic pressures rise even more acutely and to higher levels during resistance (static or isometric) exercise. Because strenuous aerobic or resistance exertion may precipitate myocardial infarction and sudden death in susceptible, untrained individuals (9), those individuals who wish to engage in competitive athletics should increase exercise levels gradually to avoid such cardiac catastrophes.

However, repetitive performance of both aerobic and resistance exercise lowers systolic and diastolic BP (4,10). After each 30-min period of aerobic exercise at 50% of maximal oxygen uptake, the blood pressure remains lower for up to 24 h, with an even greater reduction after 30 min of aerobic exercise at 75% of maximal oxygen uptake (11). As a consequence, the risks of developing elevated BP (5) and of incurring a cardiovascular consequence of hypertension (6,7) are less in those who maintain higher levels of physical activity. Some conditioned athletes (particularly young men), with a slow heart rate and compensatory increase in stroke volume, have high systolic BP that is considered "spurious" hypertension (12) but should nevertheless be carefully monitored.

In normotensive untrained subjects, an excessive rise in systolic BP to above 200 mm Hg during an exercise stress test is predictive of a greater likelihood of the development of persistent hypertension in the future and may be associated with subtle systolic dysfunction (13) and an increased risk of subsequent cardiovascular disease (14). Therefore, such subjects should be advised to increase levels of physical activity gradually to moderate such excessive rises in pressure. Such rises should not restrict activity in those who are well conditioned. Because intensive resistive training may reduce arterial compliance with potential adverse consequences (15), such training should be limited in those athletes with hypertension.

Effects of blood pressure on exercise. Untreated hypertension in athletes may be accompanied by some limitation in exercise performance (16). Before initiating drug therapy, athletes should be strongly encouraged to adopt healthy

Table 1. Guidelines for Blood Pressure Measurement

Posture

Blood pressure obtained in the seated position is recommended. The subject should sit quietly for 5 min, with the back supported in a chair, with feet on the floor, and the arm supported at the level of the heart, before recording blood pressure.

Circumstances

No caffeine during the hour preceding the reading.
No smoking during the 30 min preceding the reading.
A quiet, warm setting.

Equipment

Cuff size

The bladder should encircle and cover at least 80% of the length of the arm; if it does not, use a larger cuff. If bladder is too short, misleadingly high readings may result.

Manometer

Use a mercury, recently calibrated aneroid, or validated electronic device.

Technique

Number of readings

On each occasion, take at least two readings, separated by as much time as is practical. If readings vary by greater than 5 mm Hg, take additional readings until two consecutive readings are close. If the arm pressure is elevated, take the measurement in one leg (particularly in patients less than 30 years old). Initially, take pressures in both arms; if the pressures differ, use the arm with the higher pressure. If the initial values are elevated, obtain two other sets of readings at least 1 week apart.

Performance

Inflate the bladder quickly to a pressure 20 mm Hg above the systolic pressure, as recognized by the disappearance of the radial pulse.
Deflate the bladder 2 mm Hg per second.
Record the Korotkoff phase I (appearance) and phase V (disappearance).
If the Korotkoff sounds are weak, have the patient raise the arm, open and close the hand 5 to 10 times and then reinflate the bladder quickly.

Recordings

Blood pressure, patient position, and arm and cuff size.

From Pickering TG, et al. Hypertension 2005;45:142–61, reprinted with permission of the American Heart Association.

lifestyle behavior and avoid tobacco in any form, excess alcohol, drugs of abuse (especially sympathomimetics such as cocaine or ephedra), androgens, steroids, growth hormone, NSAIDs, and excessive sodium intake. It should be emphasized that the use of antihypertensive drugs may further limit exercise capacity, more so with beta-adrenergic receptor blocking agents than with vasodilators (alpha-adrenergic blocking agents, angiotensin-converting enzyme inhibitors, angiotensin II-receptor blockers, or calcium channel blockers). Indeed, high-intensity competitive athletes may find it very difficult to perform satisfactorily while using beta-blockers (17).

Recommendations:

1. **Before individuals commence training for competitive athletics, they should undergo careful assessment of BP and those with initially high levels (above 140/90 mm Hg) should have out-of-office measurements to exclude isolated office “white-coat” hyper-**

tension. Those with pre-hypertension (120/80 mm Hg up to 139/89 mm Hg) should be encouraged to modify lifestyle but should not be restricted from physical activity. Those with sustained hypertension should have echocardiography. Left ventricular hypertrophy (LVH) beyond that seen with “athletes’ heart” should limit participation until BP is normalized by appropriate drug therapy.

2. **The presence of stage 1 hypertension in the absence of target organ damage including LVH or concomitant heart disease should not limit the eligibility for any competitive sport. Once having begun a training program, the hypertensive athlete should have BP remeasured every two to four months (or more frequently, if indicated) to monitor the impact of exercise.**
3. **Athletes with more severe hypertension (stage 2), even without evidence of target organ damage such as LVH, should be restricted, particularly from high static sports (classes IIIA to IIIC), until their hypertension is controlled by either lifestyle modification or drug therapy.**
4. **All drugs being taken must be registered with appropriate governing bodies to obtain a therapeutic exemption.**
5. **When hypertension coexists with another cardiovascular disease, eligibility for participation in competitive athletics is usually based on the type and severity of the associated condition.**

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TASK FORCE 5 REFERENCES

1. Chobanian AV, Bakris GL, Black HR, et al. The seventh report of the Joint National Committee on Prevention, Detection, Evaluation, and Treatment of High Blood Pressure: the JNC 7 report. JAMA 2003;289:2560–72.
2. National High Blood Pressure Education Program Working Group on High Blood Pressure in Children and Adolescents. The fourth report on the diagnosis, evaluation, and treatment of high blood pressure in children and adolescents. Pediatrics 2004;114 Suppl:555–76.
3. Maron BJ. Sudden death in young athletes. N Engl J Med 2003;349:1064–75.
4. Whelton SP, Chin A, Xin X, He J. Effect of aerobic exercise on blood pressure: a meta-analysis of randomized, controlled trials. Ann Intern Med 2002;136:493–503.
5. Hu G, Barengo NC, Tuomilehto J, Lakka TA, Nissinen A, Jousilahti P. Relationship of physical activity and body mass index to the risk of hypertension: a prospective study in Finland. Hypertension 2004;43:25–30.
6. Lee CD, Folsom AR, Blair SN. Physical activity and stroke risk: a meta-analysis. Stroke 2003;34:2475–81.
7. Church TS, Kampert JB, Gibbons LW, Barlow CE, Blair SN. Usefulness of cardiorespiratory fitness as a predictor of all-cause and cardiovascular disease mortality in men with systemic hypertension. Am J Cardiol 2001;88:651–6.
8. Pickering TG, Hall JE, Appel L J, et al. Recommendations for blood pressure measurement in humans and animals: part 1: blood pressure measurement in humans: an AHA scientific statement from the Council on High Blood Pressure Research, Professional, and Publication Subcommittee. Hypertension 2005;45:142–61.
9. Albert CM, Mittleman MA, Chae CU, Lee IM, Hennekens CH, Manson JE. Triggering of sudden death from cardiac causes by vigorous exertion. N Engl J Med 2000;343:1355–61.

10. Kelley GA, Kelley KS. Progressive resistance exercise and resting blood pressure: a meta-analysis of randomized controlled trials. *Hypertension* 2000;35:838–43.
11. Quinn TJ. Twenty-four hour, ambulatory blood pressure responses following acute exercise: impact of exercise intensity. *J Hum Hypertens* 2000;14:547–53.
12. Mahmud A, Feely J. Spurious systolic hypertension of youth: fit young men with elastic arteries. *Am J Hypertens* 2003;16:229–32.
13. Mottram PM, Haluska B, Yuda S, Leano R, Marwick TH. Patients with a hypertensive response to exercise have impaired systolic function without diastolic dysfunction or left ventricular hypertrophy. *J Am Coll Cardiol* 2004;43:848–53.
14. Kjeldsen SE, Mundal R, Sandvik L, Erikssen G, Thaulow E, Erikssen J. Supine and exercise systolic blood pressure predict cardiovascular death in middle-aged men. *J Hypertens* 2001;19:1343–8.
15. Miyachi M, Kawano H, Sugawara J, et al. Unfavorable effects of resistance training on central arterial compliance: a randomized intervention study. *Circulation* 2004;110:2858–63.
16. Missault L, Duprez D, de Buyzere M, de Backer G, Clement D. Decreased exercise capacity in mild essential hypertension: non-invasive indicators of limiting factors. *J Hum Hypertens* 1992;6:151–5.
17. Vanhees L, Defoor JG, Schepers D, et al. Effect of bisoprolol and atenolol on endurance exercise capacity in healthy men. *J Hypertens* 2000;18:35–43.

Appendix 1. Author Relationships With Industry and Others

Name	Research Grant	Scientific Advisory Board	Speakers' Bureau	Steering Committee
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Dr. Thomas G. Pickering	None	None	• Boehringer Ingelheim	None
Dr. Jackson T. Wright, Jr.	• Beechham • GlaxoSmithKline • Novartis Pharma AG • Phoenix Pharmaceuticals • Solvay/Unimed	• Novartis Pharma AG • Pfizer	• Astra Zeneca • Aventis Pharmaceuticals • Bayer • BMS • Beechham • Eli Lilly & Co. • GlaxoSmithKline • Merck & Co. • Novartis Pharma AG • Pfizer • Phoenix Pharmaceuticals • Proctor & Gamble • Reliant Pharmaceutical • Solvay/Unimed	• Beechham • GlaxoSmithKline • Phoenix Pharmaceuticals • Solvay/Unimed

Task Force 6: Coronary Artery Disease

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ATHEROSCLEROTIC CORONARY ARTERY DISEASE

General considerations. Compelling evidence indicates that physical activity reduces cardiovascular events in healthy subjects and cardiac mortality in patients with diagnosed coronary artery disease (CAD) (1). Despite these beneficial exercise effects, vigorous physical activity also transiently increases the risk of both acute myocardial infarction (AMI) (2–4) and sudden cardiac death (SCD) (5–7) with the greatest exercise risk among the most habitually sedentary individuals (2,4,7).

Atherosclerotic CAD is the most frequent cause of these exercise-related cardiac events in adults (8), variously defined as

older than 30, 35, or 40 years of age. Both plaque rupture (9,10) and possibly plaque erosion (11) have been implicated as the immediate cause of exercise-related events in adults, although plaque rupture is more frequent. Several studies over the last decade document that cardiac events frequently occur in coronary arteries that were not previously critically narrowed. This appears to be particularly true for exercise-related cardiac events because angiographic studies of exercise-related AMI in the general population (4) and in sport participants (10) demonstrate less extensive CAD than in comparison subjects. This observation may reflect either selection bias for less severe atherosclerosis in those capable of exercising at high intensity

or the ability of exercise to provoke events in individuals with less severe disease.

Prognosis for diagnosed CAD patients worsens with the extent of disease, left ventricular (LV) systolic dysfunction, inducible ischemia, and electrical instability. Both the recognition that acute cardiac events often occur at the site of previously mild coronary stenoses in the general population and the observation that victims of exercise-related CAD events have less extensive disease than do individuals suffering non-exercise related events reduce the utility of standard CAD risk assessment in evaluating older competitive athletes. Adolescent and young adult athletes also may have variants in their resting electrocardiograms (ECGs) that make the interpretation of their resting and exercise ECGs difficult. In addition, early CAD is increasingly identified by such imaging techniques as coronary artery calcification scoring, further complicating risk assessment because the presence of any coronary artery calcification indicates atherosclerotic disease. Finally, since CAD primarily occurs in adult athletes, the age of the athlete at risk for CAD events complicates the decision process. Older athletes are often engaged in individual competitive pursuits and may not require formal clearance for continued competition. Preparticipation screening guidelines specifically for participants in master's sports have been developed in association with the American Heart Association (AHA) (12).

Diagnosis. The diagnosis of atherosclerotic CAD is established in the presence of any of the following: 1) a history of a myocardial infarction (MI) confirmed by conventional diagnostic criteria; 2) a history suggestive of angina pectoris with objective evidence of inducible ischemia; and 3) coronary atherosclerosis of any degree is demonstrated by coronary imaging studies such as catheter-based coronary angiography, magnetic resonance angiography, or electron beam computerized tomography (EBCT).

CORONARY CALCIFICATION BY COMPUTED TOMOGRAPHY (CT). Since the last version of these guidelines, the widespread dissemination of noninvasive techniques such as EBCT, or even more recently, multi-slice gated CT, has markedly increased the number of individuals, including competitive athletes, who may be diagnosed with atherosclerotic CAD. Although exceedingly rare in young persons (6% of men and 3% of women 20 to 29 years of age), the presence of coronary calcium increases substantially with age, such that for master's athletes, age 40 to 49 years, approximately 41% of men and 13% of women may have measurable coronary calcium (13). Among individuals age 50 to 59 years, 68% of men and 27% of women have documented coronary calcium (13). There is compelling evidence that the presence of any coronary calcium indicates underlying atherosclerosis (14) and that increasing coronary calcium scores are associated with increased CAD risk (15). The coronary calcium score that warrants additional evaluation in asymptomatic competitive athletes is unknown, although scores of more than 100 (15) have been associated

with increased risk for coronary events (16) in the general population compared to patients with no coronary calcium. It is unknown whether the risk of coronary events during intense exercise is increased in the presence of this or lesser amounts of coronary calcium. Nevertheless, for the purpose of the present document, athletes with coronary artery calcification scores more than 100 should undergo the same evaluation as those with more clinically evident CAD.

RISK ASSESSMENT. A paucity of data exists in competitive athletes directly relating the presence and severity of CAD to the risk of athletic participation. This requires that these recommendations for athletes with CAD be based in part on observations obtained from non-athletes with CAD. Nevertheless, it is likely that risk is increased to some degree whenever coronary atherosclerosis is present. It is also likely that the risk of exercise-related events increases with the extent of disease, LV dysfunction, inducible ischemia, and electrical instability, and that the risk increases with the intensity of the competitive sport and the intensity of the participant's effort.

Evaluation.

1. Athletes with CAD diagnosed by any method including coronary artery classification scoring more than 100, coronary angiography, evidence of inducible ischemia, or prior coronary event, and who are undergoing evaluation for competitive athletics, should have their LV function assessed.
2. These athletes should undergo maximal treadmill (or bicycle) exercise testing to assess their exercise capacity and the presence or absence of provokable myocardial ischemia. Exercise testing should approximate as closely as possible the cardiovascular and metabolic demands of the planned competitive event and its training regimen. Despite such simulation, graded exercise testing cannot replicate the cardiovascular stress produced by the sudden bursts of activity, the combination of high dynamic and static exercise, such as rowing, or the sustained bouts of exercise required by athletic training and competition. Therefore, standard clinical exercise tests may not be appropriate for the evaluation of athletes with coronary heart disease.

RISK STRATIFICATION. Two levels of risk can be defined on the basis of testing.

Mildly increased risk. Athletes with CAD diagnosed by any method are judged to be at mildly increased risk if they demonstrate all of the following:

1. Preserved LV systolic function at rest (i.e., ejection fraction greater than 50%).
2. Normal exercise tolerance for age, demonstrated during treadmill or cycle ergometer exercise testing: greater than 10 metabolic equivalents (METs), or greater than 35 O₂/kg-min if less than 50 years old; greater than 9 METs, or greater than 31 ml O₂/kg-min for 50 to 59 years old; greater than 8 METs, or greater than 28 ml

- O₂/kg-min, if 60 to 69 years old; and greater than 7 METS, or greater than 24 ml O₂/kg-min, if greater than or equal to 70 years old. It should be noted that young, highly competitive endurance athletes should have maximal oxygen uptakes far in excess of ranges regarded as normal, which in fact may represent substantial functional impairment in this population.
3. Absence of exercise-induced ischemia and exercise-induced or post-exercise complex ventricular arrhythmias, including frequent premature ventricular contractions (greater than 10% of beats/min), couplets, or ventricular tachycardia.
 4. Absence of hemodynamically significant stenosis (generally regarded as 50% or more luminal diameter narrowing) in any major coronary artery by coronary angiography.
 5. Successful myocardial revascularization by surgical or percutaneous techniques if such revascularization was performed.

Substantially increased risk. Athletes with CAD identified by noninvasive or invasive testing are judged to be at substantially increased risk if they demonstrate *any* of the following:

1. Impaired LV systolic function at rest (i.e., ejection fraction less than 50%).
2. Evidence of exercise-induced myocardial ischemia or complex ventricular arrhythmias.
3. Hemodynamically significant stenosis of a major coronary artery (generally regarded as 50% or more lumen diameter narrowing) if coronary angiography was performed.

The American College of Cardiology/AHA guidelines on exercise testing note that it is not necessary to stop beta-blockers before routine exercise testing, although this practice may reduce the diagnostic and prognostic value of the test (17). The decision whether or not to stop beta-blocker therapy before exercise testing of athletes should be made on an individual basis. Stopping beta-blockers and other anti-ischemic medications before testing may be useful to more closely approximate the probable risk if the athlete either intentionally or unintentionally does not take these medications before competition, or when certain athletic regulatory bodies prohibit beta-blockers. If anti-ischemic medications are stopped, this should be done carefully to avoid a potential hemodynamic rebound effect, which could lead to accelerated angina or hypertension.

Coronary arteriography is not required to determine eligibility for competition in patients with known CAD, and no evidence of inducible ischemia, but is recommended in athletes with exercise-induced ischemia who choose to participate in sports against medical advice. Such studies may identify coronary lesions that may be better managed by percutaneous or surgical myocardial revascularization procedures to relieve exercise-induced ischemia and potentially to reduce exercise-related risk.

The panel wishes to emphasize that the following rec-

ommendations are prepared as a guidelines for permitting participation in *competitive sports*. Restrictions in the following recommendations, therefore, should not be misinterpreted as an injunction against regular physical activity as opposed to athletic competition. Indeed, regular and recreational physical activity and moderate-intensity exercise training are recommended for patients with CAD for its general cardiovascular benefits (1).

Recommendations:

1. **Athletes in the mildly increased risk group can participate in low dynamic and low/moderate static competitive sports (classes IA and IIA—see Fig. 1 in Task Force 8: Classification of Sports) but should avoid intensely competitive situations. We recognize that selected athletes with mildly increased risk may be permitted to compete in sports of higher levels of intensity when their overall clinical profile suggests very low exercise risk. This is particularly true for athletes in whom the only indication that coronary atherosclerosis is present is from an EBCT performed for screening purposes, and in which the total coronary calcium score is relatively low (i.e., less than 15). Increasing amounts of coronary calcium, suggestive of increasing burden disease, should dictate a more cautious approach, particularly if the coronary calcium score is more than 100. All athletes should understand that the risk of a cardiac event with exertion is probably increased once coronary atherosclerosis of any severity is present. Athletes with mildly increased risk engaging in competitive sports should undergo re-evaluation of their risk stratification at least annually.**
2. **Athletes in the substantially increased risk category should generally be restricted to low-intensity competitive sports (class IA).**
3. **Athletes should be informed of the nature of prodromal symptoms (such as chest, arm, jaw and shoulder discomfort, unusual dyspnea) and should be instructed to cease their sports activity promptly and to contact their physician if symptoms appear. Physicians should be aware that competitive athletes may minimize symptoms that occur during exertion.**
4. **Those with a recent MI or myocardial revascularization should cease their athletic training and competition until recovery is deemed complete. This interval varies among patient groups, but depends on the severity of the cardiovascular event and the extent and success of the revascularization procedure. Such patients may benefit from cardiac rehabilitation during the recovery period. No firm guidelines exist for how long patients should avoid vigorous exercise training, but in general, patients post-stent placement for stable CAD symptoms should avoid vigorous exercise training for competition for approximately four weeks. Patients undergoing stent**

placement for unstable disease should wait at least this long. Following coronary bypass surgery, patients should avoid vigorous training until their incisions can tolerate vigorous activity. After recuperation period, the risk and activity level should be defined as in recommendations 1 and 2.

5. All athletes with atherosclerotic CAD should have their atherosclerotic risk factors aggressively treated as studies suggest that comprehensive risk reduction is likely to stabilize coronary lesions and may reduce the risk of exercise-related events.

It must be emphasized that even athletes identified as being at mildly increased risk and permitted to participate in low dynamic and low/moderate static competitive sports (classes IA and IIA) cannot be assured that such participation will not increase the risk of cardiac events because it is probable that any exercise transiently poses some increased exercise risk once CAD is established.

CORONARY ARTERY VASOSPASM

Coronary artery vasospasm classically presents as rest angina associated with ST-segment elevation, but can be provoked by physical exertion on rare occasions (18). Vasospasm is an uncommon cause of chest pain that is evident in 2% to 3% of patients presenting with chest pain undergoing coronary angiography (19). Vasospasm is most frequently observed at coronary sites damaged by atherosclerosis (20), but a substantial cohort may have angiographically normal coronary arteries or minimal angiographic luminal narrowing (18,19). A vasospastic contribution to ischemia should be suspected when there is marked variation in the exercise threshold for angina (18), and when there is evidence of myocardial ischemia with little or no coronary luminal narrowing. Presently, no widely accepted noninvasive test exists for eliciting and quantifying vasospastic angina in the setting of nonobstructive or mildly obstructive coronary arteries. The occurrence of ST-segment elevation during exercise testing appears to correlate with the degree of disease activity (i.e., those with more frequent episodes of angina will more likely have a positive test) (21). Provocative testing with ergonovine-related substances during coronary arteriography is rarely used (22), but remains the only test recommended in current practice guidelines (23). However, forced hyperventilation testing, particularly when combined with nuclear perfusion imaging, may be a useful noninvasive test not requiring the administration of ergonovine (24). The risk associated with participation in sports for athletes with coronary artery spasm is not known, but we recommend a cautious approach to patients with documented coronary vasospasm until the risk of physical exertion for these patients is better defined.

Recommendations:

1. Athletes with CAD as previously defined and clinically important coronary artery vasospasm should follow the

evaluation and risk stratification approach delineated for athletes with coronary atherosclerosis.

2. Athletes with coronary vasospasm documented at rest or with exercise and angiographically normal coronary arteries or without evidence of arterial plaquing should be restricted to low-intensity competitive sports (class IA). This restriction should be re-evaluated at least annually because some patients with coronary vasospasm may experience spontaneous remission.

CAD IN CARDIAC TRANSPLANT RECIPIENTS

Orthotopic transplanted hearts develop an accelerated form of coronary vasculopathy, usually detected by serial coronary angiography or intravascular ultrasound studies, that is a leading cause of death after the first post-transplant year (25,26). The coronary disease is different from that seen in non-transplanted hearts with coronary atherosclerosis; the disease is diffuse and characterized by pronounced intimal thickening and involvement of the entire coronary tree. Discrete stenoses of epicardial arteries can coexist in some instances. Cardiac allografts are denervated, and although some recipients may develop a degree of sympathetic reinnervation, acute coronary syndromes may present with atypical symptoms as opposed to angina (27). Noninvasive testing for CAD is less sensitive in the transplant recipient; many patients do not achieve VO_{2max} , and cardiac denervation can limit peak heart rate response and symptoms. Provocative myocardial perfusion imaging can fail to detect ischemia (25,27) although dobutamine echocardiography has been shown to predict subsequent ischemic cardiac events (26,28–31) after the first three to five years post-transplant. In many cardiac transplant centers, a normal stress echocardiogram justifies postponement of annual coronary angiography (26,28–31). Coronary angiography can also underestimate disease severity because of the diffuse nature of the CAD process; intravascular ultrasound studies increase the sensitivity (26).

Evaluation.

1. Cardiac transplant recipients participating in competitive athletics should undergo yearly maximal exercise testing with echocardiography using a protocol designed to simulate the cardiac and metabolic demands of the competitive event and its training.
2. Additional evaluation, including such procedures as coronary angiography and intravascular ultrasonography (IVUS) should be performed as directed by the transplant center and the transplant cardiologist. Coronary angiography/IVUS should also be performed if the annual exercise test is abnormal and to evaluate unexplained symptoms such as dyspnea or exertional fatigue as these may be the only symptoms of progressive vascular disease.

Recommendations:

1. Because of the special issues involved with transplant patient management, decisions as to the feasibility of athletic competition for cardiac transplant recipients should be made in conjunction with the patient's transplant cardiologist.
2. Athletes with no coronary luminal narrowing, no exercise-induced ischemia, and with normal exercise tolerance for age (as previously defined) can generally participate in all competitive sports as appropriate for their exercise capacity.
3. Athletes with coronary luminal narrowing should be risk stratified as outlined in the section entitled Evaluation with activity recommendations as indicated in the section entitled Atherosclerotic Coronary Artery Disease.

MYOCARDIAL BRIDGING

Myocardial bridging is a condition in which a segment of major epicardial coronary artery (most commonly the left anterior descending) is tunneled within and completely surrounded by LV myocardium. Myocardial bridging is found in approximately 30% of hearts examined at necropsy (32), but is visualized during angiography in less than 5% of patients probably because the thin bridges identified at necropsy cause little anatomic compression during systole (32). Consequently, most tunneled epicardial coronary arteries appear to be of little clinical significance, but this malformation has occasionally been associated with exercise-related sudden death (33,34) and exercise-induced angina pectoris (35). Clinically significant myocardial bridges have a long deeply-tunneled segment and are associated with regional ischemia. Treatment options include medical management with beta-adrenergic or calcium channel-blocking agents, coronary stenting, and surgical resection of the myocardial bridge.

However, coronary stenting has been associated with restenosis and periprocedural complications in 50% of cases (32). Surgical resection in selected symptomatic patients has been shown to reduce angina (35) and improve myocardial blood flow (36). Task Force 4: HCM and Other Cardiomyopathies, Mitral Valve Prolapse, Myocarditis, and Marfan Syndrome provides recommendations for the management of myocardial bridging in patients with hypertrophic cardiomyopathy (HCM).

Recommendations:

1. Athletes with myocardial bridging of an epicardial coronary artery and no evidence of myocardial ischemia at rest or during exercise can participate in all competitive sports as appropriate for their exercise capacity.
2. Athletes with myocardial bridging of an epicardial coronary artery and objective evidence of myocardial

- ischemia or prior MI should be restricted to low-intensity competitive sports (class IA).
3. Athletes with surgical resection of the myocardial bridge or stenting should be restricted to low-intensity sports for at least six months after the procedure. Athletes who remain asymptomatic after the procedure should undergo exercise testing. If exercise tolerance is normal for age and gender, and there is no evidence of exercise-induced ischemia, the athlete may participate in all competitive sports.

Finally, recommendations for other congenital coronary artery anomalies, including those originating from the wrong coronary sinus, are provided in Task Force 1: Preparticipation Screening and Diagnosis of Cardiovascular Disease in Athletes.

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TASK FORCE 6 REFERENCES

1. Thompson PD, Buchner D, Pina IL, et al. Exercise and physical activity in the prevention and treatment of atherosclerotic cardiovascular disease: a statement from the Council on Clinical Cardiology (Subcommittee on Exercise, Rehabilitation, and Prevention) and the Council on Nutrition, Physical Activity, and Metabolism (Subcommittee on Physical Activity). *Circulation* 2003;107:3109-16.
2. Mittleman MA, Maclure M, Toffler GH, Sherwood JB, Goldberg RJ, Muller JE. Triggering of acute myocardial infarction by heavy physical exertion: protection against triggering by regular exertion. Determinants of Myocardial Infarction Onset Study Investigators. *N Engl J Med* 1993;329:1677-83.
3. Willich SN, Lewis M, Lowel H, Arntz HR, Schubert F, Schroder R. Physical exertion as a trigger of acute myocardial infarction. Triggers and Mechanisms of Myocardial Infarction Study Group. *N Engl J Med* 1993;329:1684-90.
4. Giri S, Thompson PD, Kiernan FJ, et al. Clinical and angiographic characteristics of exertion-related acute myocardial infarction. *JAMA* 1999;282:1731-6.
5. Thompson PD, Funk EJ, Carleton RA, Sturner WQ. Incidence of death during jogging in Rhode Island from 1975 through 1980. *JAMA* 1982;247:2535-8.
6. Siscovick DS, Weiss NS, Fletcher RH, Lasky T. The incidence of primary cardiac arrest during vigorous exercise. *N Engl J Med* 1984;311:874-7.
7. Albert CM, Mittleman MA, Chae CU, Lee IM, Hennekens CH, Manson JE. Triggering of sudden death from cardiac causes by vigorous exertion. *N Engl J Med* 2000;343:1355-61.
8. Ragosta M, Crabtree J, Sturner WQ, Thompson PD. Death during recreational exercise in the state of Rhode Island. *Med Sci Sports Exerc* 1984;16:339-42.
9. Black A, Black MM, Gensini G. Exertion and acute coronary artery injury. *Angiology* 1975;26:759-83.
10. Ciampricotti R, Deckers JW, Taverne R, el Gamal M, Relik-van Wely L, Pool J. Characteristics of conditioned and sedentary men with acute coronary syndromes. *Am J Cardiol* 1994;73:219-22.
11. Burke AP, Farb A, Malcom GT, Liang Y, Smialek JE, Virmani R. Plaque rupture and sudden death related to exertion in men with coronary artery disease. *JAMA* 1999;281:921-6.
12. Maron BJ, Araujo CG, Thompson PD, et al. Recommendations for preparticipation screening and the assessment of cardiovascular disease in masters athletes: an advisory for healthcare professionals from the working groups of the World Heart Federation, the International Federation of Sports Medicine, and the American Heart Association Committee on Exercise, Cardiac Rehabilitation, and Prevention. *Circulation* 2001;103:327-34.
13. Cheng YJ, Church TS, Kimball TE, et al. Comparison of coronary artery calcium detected by electron beam tomography in patients with to those without symptomatic coronary heart disease. *Am J Cardiol* 2003;92:498-503.

14. Wexler L, Brundage B, Crouse J, et al. Coronary artery calcification: pathophysiology, epidemiology, imaging methods, and clinical implications. A statement for health professionals from the American Heart Association Writing Group. *Circulation* 1996;94:1175-92.
15. O'Rourke RA, Brundage BH, Froelicher VF, et al. American College of Cardiology/American Heart Association expert consensus document on electron-beam computed tomography for the diagnosis and prognosis of coronary artery disease. *Circulation* 2000;102:126-40.
16. Greenland P, LaBree L, Azen SP, Doherty TM, Detrano RC. Coronary artery calcium score combined with Framingham score for risk prediction in asymptomatic individuals. *JAMA* 2004;291:210-5.
17. Gibbons RJ, Balady GJ, Bricker JT, et al. ACC/AHA 2002 guideline update for exercise testing: a report of the American College of Cardiology/American Heart Association Task Force on Practice Guidelines (Committee on Exercise Testing). Available at: www.acc.org/clinical/guidelines/exercise/dirIndex.htm. Accessed October 1, 2004.
18. Yasue H, Omote S, Takizawa A, Nagao M, Miwa K, Tanaka S. Circadian variation of exercise capacity in patients with Prinzmetal's variant angina: role of exercise-induced coronary arterial spasm. *Circulation* 1979;59:938-48.
19. Mark DB, Califf RM, Morris KG, et al. Clinical characteristics and long-term survival of patients with variant angina. *Circulation* 1984;69:880-8.
20. Gordon JB, Ganz P, Nabel EG, et al. Atherosclerosis influences the vasomotor response of epicardial coronary arteries to exercise. *J Clin Invest* 1989;83:1946-52.
21. Waters DD, Szelachic J, Bourassa MG, Scholl JM, Theroux P. Exercise testing in patients with variant angina: results, correlation with clinical and angiographic features and prognostic significance. *Circulation* 1982;65:265-74.
22. Hamilton KK, Pepine CJ. A renaissance of provocative testing for coronary spasm? *J Am Coll Cardiol* 2000;35:1857-9.
23. Gibbons RJ, Abrams J, Chatterjee K, et al. ACC/AHA 2002 guideline update for the management of patients with chronic stable angina—summary article: a report of the American College of Cardiology/American Heart Association Task Force on Practice Guidelines (Committee on the Management of Patients With Chronic Stable Angina). *Circulation* 2003;107:149-58.
24. Shanoudy H, Raggi P, Gasperetti C, et al. Detection of coronary vasospasm by posthyperventilation technetium-99m sestamibi single-photon emission computed tomography imaging in patients with coronary artery disease. *Am J Cardiol* 1998;81:573-7.
25. Miller LW, Schlant RC, Kobashigawa J, Kubo S, Renlund DG. 24th Bethesda Conference: cardiac transplantation. Task force 5: complications. *J Am Coll Cardiol* 1993;22:41-54.
26. Uretsky BF, Kormos RL, Zerbe TR et al. Cardiac events after heart transplantation: incidence and predictive value of coronary arteriography. *J Heart Lung Transplant* 1992;11:S45-51.
27. Schroeder JS, Gao SZ, Hunt SA, Stinson EB. Accelerated graft coronary artery disease: diagnosis and prevention. *J Heart Lung Transplant* 1992;11:S258-65.
28. Spes CH, Klauss V, Mudra H, et al. Diagnostic and prognostic value of serial dobutamine stress echocardiography for noninvasive assessment of cardiac allograft vasculopathy: a comparison with coronary angiography and intravascular ultrasound. *Circulation* 1999;100:509-15.
29. Spes CH, Klauss V, Rieber J, et al. Functional and morphological findings in heart transplant recipients with a normal coronary angiogram: an analysis by dobutamine stress echocardiography, intracoronary Doppler and intravascular ultrasound. *J Heart Lung Transplant* 1999;18:391-8.
30. Larsen RL, Applegate PM, Dyar DA, et al. Dobutamine stress echocardiography for assessing coronary artery disease after transplantation in children. *J Am Coll Cardiol* 1998;32:515-20.
31. Akosah KO, McDaniel S, Hanrahan JS, Mohanty PK. Dobutamine stress echocardiography early after heart transplantation predicts development of allograft coronary artery disease and outcome. *J Am Coll Cardiol* 1998;31:1607-14.
32. Mohlenkamp S, Hort W, Ge J, Erbel R. Update on myocardial bridging. *Circulation* 2002;106:2616-22.
33. Morales AR, Romanelli R, Tate LG, Boucek RJ, de Marchena E. Intramural left anterior descending coronary artery: significance of the depth of the muscular tunnel. *Hum Pathol* 1993;24:693-701.
34. Maron BJ, Shirani J, Poliac LC, Mathenge R, Roberts WC, Mueller FO. Sudden death in young competitive athletes: clinical, demographic, and pathological profiles. *JAMA* 1996;276:199-204.
35. Betriu A, Tubau J, Sanz G, Magrina J, Navarro-Lopez F. Relief of angina by periarterial muscle resection of myocardial bridges. *Am Heart J* 1980;100:223-6.
36. Hill RC, Chitwood WR Jr., Bashore TM, Sink JD, Cox JL, Wechsler AS. Coronary flow and regional function before and after supraarterial myotomy for myocardial bridging. *Ann Thorac Surg* 1981;31:176-81.

Appendix 1. Author Relationships With Industry and Others

Name	Consultant	Research Grant	Scientific Advisory Board	Speakers' Bureau	Stock Holder	Expert Witness Testimony
Dr. Gary J. Balady	None	None	None	None	None	None
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Dr. Luther T. Clark	None	None	None	None	None	None
Dr. Benjamin D. Levine	None	None	None	None	None	None
Dr. Robert J. Myerburg	• Guidant • Procter & Gamble	None	• Procter & Gamble • Reliant Pharmaceutical	None	None	• 2000, Defense, Lewis vs. Mudge • 2002, Defense, Weiner vs. Vitello • 2005, Defense, Ephedra Multi-District Litigation
Dr. Paul D. Thompson	• Astra Zeneca • Bristol Myers Squibb	• Astra Zeneca • Merck • Pfizer • Schering	None	• Astra Zeneca • Merck • Pfizer • Schering	• Pfizer • Schering	• 2005, Plaintiff, Sudden death in college athlete • 2004, Defense, Stress test/recreational sports • 2002, Sudden death, World Gym • 2002, Plaintiff, Sudden death, truck driver • 1998, Plaintiff, Sudden death, recreational sports

Task Force 7: Arrhythmias

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GENERAL CONSIDERATIONS

Guidelines for athletic participation are needed to reduce the risk for arrhythmia-related morbidity or mortality. However, it is often difficult to establish the importance of a cardiac rhythm disturbance in assessing an athlete's eligibility for competition. Few data exist that have been obtained prospectively from well-designed, scientifically acceptable studies to determine whether a particular rhythm disturbance predisposes an athlete to sudden death or to symptoms, such as syncope or presyncope, that could precipitate severe injury. Sudden unexpected cardiac death in the young is rare, estimated at less than 1% of that observed in adults. Nonetheless, a significant proportion of these deaths occur in relation to exercise (1). The deaths of several prominent athletes have focused attention on athletes with known arrhythmias.

Arrhythmias commonly are evanescent, often disappearing unpredictably for long periods of time, in some cases years. If they recur when the athlete is not exercising, the arrhythmia may not be noted or may not produce significant symptoms. The same arrhythmia may minimally affect a competitive golfer but severely incapacitate other athletes, such as cross-country skiers, performing at peak physical effort. The athlete may not develop the arrhythmia during each sporting event. Although the reasons for this are not known, factors related to the autonomic nervous system probably play a very important role in determining whether an arrhythmia occurs and its rate and effect on hemodynamic responses and symptoms. Autonomic "tone" probably varies greatly and perhaps unpredictably between and within athletic events and from one athlete to another. Mental stress during competition can produce important electrophysiologic and hemodynamic changes that are probably mediated through the autonomic nervous system.

It is important to understand the range of normal heart rate and rhythm for the trained athlete. Heart rates of 25 beats/min and sinus pauses lasting greater than 2 s may be found on 24-h Holter ambulatory electrocardiographic (ECG) recordings. Type I second-degree atrioventricular (AV) block and single uniform premature ventricular complexes each may occur in approximately 40% of athletes. Complex ventricular arrhythmias (multiform premature ventricular complexes, couplets, nonsustained ventricular tachycardia) are less common (2,3).

Many of our conclusions result from data obtained in non-athletes, from general perceptions, or experience and from a heavy input of "what seems reasonable." Decision-

making based on this type of logic is often faulty but is the best available. Recommendations and guidance need to be balanced between an effort to avoid restricting activity unduly and the hope of reducing the risk of death and injury due to a rhythm disturbance.

Despite the lack of complete information, some firm conclusions can be reached. Certain arrhythmias, such as ventricular tachyarrhythmias, create symptoms and are dangerous in and of themselves regardless of the clinical situation in which they occur (4). These arrhythmias generally are characterized by very rapid or very slow heart rates that significantly compromise cardiac output, coronary or cerebral blood flow, or maintenance of blood pressure. Such arrhythmias may include atrial flutter or atrial fibrillation with uncontrolled ventricular rates of 200 to 300 beats/min, usually (but not exclusively) in athletes with Wolff-Parkinson-White (WPW) syndrome, rapid sustained ventricular tachycardias, and AV block, or sinus node disease with very slow ventricular rates. Certain persistent arrhythmias, such as chronic tachycardias, can worsen cardiac function by a process called "remodeling" (5). Other arrhythmias, such as AV nodal re-entrant tachycardia, generally well-tolerated in most people, may not produce important symptoms at rest but only during exercise in athletes with structural heart disease due, in part, to an increase in the tachycardia rate. Arrhythmias that might otherwise be innocuous and no more than a nuisance might, under conditions of participation in certain sports involving body contact or high speed, place the athlete at risk of injury or death because of transient impaired mental function causing loss of physical control. For example, it is possible that an athlete with supraventricular tachycardia participating in potentially dangerous sports, such as diving, downhill skiing, or auto racing, may be at greater risk because of dizziness, near syncope (a feeling of impending loss of consciousness), or syncope than if he or she were playing basketball or baseball.

The search for significant structural heart disease is an important element in evaluating athletes with arrhythmias prior to sports participation. Some athletes with coronary artery disease (CAD), hypertrophic cardiomyopathy, arrhythmogenic right ventricular cardiomyopathy (ARVC), aortic stenosis, some inherited cardiac channelopathies like congenital long QT syndrome (LQTS) (6), and other forms of congenital heart disease, including repaired congenital heart disease, are probably at greater risk for cardiac arrest and sudden death during and, perhaps, just after exercise.

This is probably true whether or not arrhythmias have been recognized previously. In general, athletes with symptoms possibly related to cardiac arrhythmia, such as exertional- or auditory-triggered syncope, near syncope, and palpitations, should be carefully evaluated (see Task Force 2: Congenital Heart Disease) before being permitted to participate in competitive sports. A consideration of cardiac hemodynamic status is critical because right or left ventricular dysfunction is an additional important predictor of arrhythmic death. The presence of a significant rhythm disturbance, such as a rapid supraventricular or ventricular tachyarrhythmia in athletes with abnormal cardiac hemodynamic status (from any cause), itself is definitely incompatible with participation in all competitive sports. However, it is important to emphasize that some disease states, such as a myocarditis, can produce arrhythmias that can be self-limited, with subsequent full recovery.

In general, all athletes with significant cardiac arrhythmias being considered for athletic activity should have a careful cardiac examination, a 12-lead ECG, echocardiogram, exercise test, and, in some, a long-term 24-h Holter ambulatory ECG recording, if possible during the specific type of exercise being considered. Arrhythmias, as discussed in the various Task Force documents, are usually identified by exercise testing or some form of long-term monitoring (including ambulatory Holter and event recording). Arrhythmias precipitated during the specific type of exercise being considered can be important because a conventional exercise test may not replicate the specific clinical situation produced by actively participating in the sport. In this regard, exercise tests may need to be adapted specifically for the athlete; that is, to begin exercise at peak energy expenditure, as a sprinter in a race might, rather than with the slow increase in workload commonly used in testing athletes with CAD. Resuscitation equipment and trained personnel may be needed on a standby basis.

All athletes with an arrhythmia who are permitted to engage in athletics should be re-evaluated at 6 to 12 month intervals after they are trained, to determine whether the conditioning process affected the arrhythmia. It should also be stressed that athletes with arrhythmias controlled by antiarrhythmic drugs may stop taking these drugs for a variety of reason and, therefore, compliance with recommended therapy, as well as evaluation for recurrence of symptoms, must be established periodically. Abuse with drugs like cocaine or ephedra can precipitate life-threatening arrhythmias, and such considerations are an important part of the evaluation. Of note, the use of certain cardioactive drugs, such as beta-adrenergic blocking agents, is banned in some competitive sports (see the Introduction). In addition, it is important to realize that the catecholamines released during exercise may counteract the salutary effects of some antiarrhythmic agents. For some arrhythmias, an ablation approach, usually with a catheter and usually with radiofrequency energy, to eliminate the arrhythmia may be preferable to drug treatment. Other

ablation energies, such as cryoablation, or other approaches such as surgery, can be used as indicated. After successful ablation of the arrhythmia, a return to athletics can be within days for those in whom repeated attempts at tachycardia induction during isoproterenol administration is unsuccessful, and in whom the tachycardia was easily induced prior to ablation. For those in whom such provocative testing is not performed, waiting two to four weeks seems advisable.

It may be difficult for team physicians and consultants from the local community to make objective decisions to restrict or proscribe sports for a competitive athlete. Borderline cases can be reviewed by non-biased experts for the purpose of helping make these decisions. Whether that approach can be practically implemented has not been determined. Alternatively, it has also been suggested that in such borderline cases, both appropriate emergency equipment and medical professionals versed in the operation of that equipment be present at all practices and games. However, there is considerable concern with regard to this practice because its safety and efficacy have not been established. Indeed, the practical implementation and reliability of this approach to reverse potentially lethal arrhythmias in out-of-hospital settings for the competing athlete is highly questionable.

SYNCOPE

Unexplained syncope in an athlete is a potentially important symptom that mandates a thorough evaluation. It may be due to a variety of causes, including cardiovascular disease; alternatively, it may not be associated with either structural heart disease or a primary electrical disorder but rather to mechanisms such as vasovagal syncope, which is a common finding in highly trained athletes. Although vasovagal syncope may be compatible with continued participation in all competitive sports, caution should be used in making this diagnosis in highly trained athletes without first definitively excluding underlying structural cardiovascular disease. A cardiac arrhythmia should be considered, particularly when syncope occurs during or immediately after exercise.

A cause of syncope can be established in approximately 50% of patients. Often, a careful history and physical examination will identify the etiology (7). When such an evaluation does not determine the cause, further testing in search of an arrhythmia is indicated. Ambulatory ECG recordings are often unrevealing but nevertheless probably worthwhile to obtain during the initial evaluation. Event or loop recorders or implantable devices can be used to increase the ECG sampling time. Tilt-table testing has been used to assess patients at risk for vasovagal syncope, but the lack of specificity of this test (particularly in endurance-trained athletes in whom false-positive results can occur) requires a particularly cautious interpretation of the results. Exercise testing is useful and is optimally performed while recording the ECG during the athletic activity in which the person

participates. Provocative catecholamine stress testing with either epinephrine, procainamide, or isoproterenol may be useful to unmask cases of concealed LQTS, Brugada syndrome (BrS), or catecholaminergic polymorphic ventricular tachycardia (CPVT) (8). Invasive electrophysiologic testing is most likely to identify an arrhythmia responsible for syncope in those patients with structural heart disease or an abnormal ECG but can be considered in other athletes when no other cause of the syncope has been identified, remembering that the diagnostic yield of invasive electrophysiologic testing is low in the absence of structural heart disease.

TYPES OF ARRHYTHMIAS

Disturbances of sinus node function. Sinus tachycardia and sinus bradycardia appropriate for the clinical situation are not considered abnormal, and no tests are necessary. Sinus arrhythmia and wandering pacemaker are generally considered normal, and no tests are necessary unless the arrhythmias result in inappropriately slow rates accompanied by symptoms. Sinus arrhythmia and sinus bradycardia are particularly common in the trained athlete.

Asymptomatic sinus pause or sinus arrest (less than 3 s) is probably of no significance. Longer symptomatic pauses, sinoatrial exit block, and sick sinus syndrome are considered abnormal, and athletes should have a 12-lead, 24-h ECG, and an exercise test. Mutations involving the cardiac sodium channel encoded by *SCN5A* has been demonstrated in some patients with progressive cardiac conduction disease and congenital sick sinus syndrome (9,10). In an occasional athlete experiencing syncope or near syncope, an electrophysiologic study may be indicated and reveal abnormal sinus node function, but in general, invasive electrophysiologic testing is not helpful. Echocardiography should be performed to exclude structural heart disease; other tests to evaluate ventricular or valvular function may be indicated.

Recommendations:

1. Athletes with a normal or structurally abnormal heart in whom the bradycardic rate is increased appropriately by physical activity can participate in all competitive sports consistent with the limitations imposed by the structural heart disease. They should be reassessed periodically to determine that training does not aggravate the bradycardia.
2. Athletes with syncope or near syncope should not participate in sports where the likelihood of even a momentary loss of consciousness may be hazardous until the cause has been determined and treated, if necessary.
3. Athletes with symptoms such as impaired consciousness and fatigue clearly attributed to the arrhythmias should be treated and if asymptomatic for two to three months during treatment, they can participate in all competitive sports after physician re-evaluation.

4. Athletes with symptomatic tachycardia/bradycardia syndrome or inappropriate sinus tachycardia should be treated. If no structural heart disease and asymptomatic for two to three months, they can participate in all competitive sports.
5. Athletes with pacemakers should not engage in sports with a danger of bodily collision because such trauma may damage the pacemaker system. This restriction should clearly exclude activities where direct blows to the chest are a part of the sport, such as football, rugby, boxing, martial arts, hockey, and lacrosse. Protective padding for the device is advisable for other sports such as soccer, basketball, baseball, and softball where trauma is possible but less likely.

Premature atrial complexes. In the absence of evidence of structural heart disease and in the absence of symptoms other than occasional palpitation, no evaluation other than a 12-lead ECG is necessary.

Recommendation:

1. Athletes can participate in all competitive sports.

Atrial flutter (in the absence of WPW syndrome). In the absence of an acute, limiting illness, sustained atrial flutter is an uncommon rhythm disturbance in athletes without structural heart disease. Therefore, an echocardiogram should be performed to evaluate cardiovascular structure and function. Because the potential for very rapid ventricular rates exists if the atrial flutter conducts 1:1 to the ventricles, ECG determination of the ventricular response during an exercise test or athletic event during treatment is essential. For some patients with paroxysmal atrial flutter, induction of the arrhythmia by electrical stimulation may be considered before the exercise test, recognizing the difficult logistics in doing this. A 12-lead ECG and, at times, long-term 24-h ECG may be necessary. Asymptomatic athletes who have transient episodes of atrial flutter lasting less than 10 s that do not increase in duration during exercise can participate in all sports.

Recommendations:

1. Athletes with atrial flutter in the absence of structural heart disease who maintain a ventricular rate that increases and slows appropriately comparable to that of a normal sinus response in relation to the level of activity, while receiving no therapy or therapy with AV nodal blocking drugs, can participate in class IA competitive sports with the warning that rapid 1:1 conduction still may occur. However, full participation in all competitive sports should not be allowed unless the athlete has been without atrial flutter for two to three months with or without drug treatment. Note that the use of beta-blockers is prohibited in some competitive sports (see Introduction).

2. Athletes with structural heart disease who have atrial flutter can participate in class IA competitive sports only after two to four weeks have elapsed without an episode of atrial flutter.
3. Athletes without structural heart disease who have elimination of the atrial flutter by an ablation technique or surgery can participate in all competitive sports after two to four weeks without a recurrence, or in several days after an electrophysiologic study showing non-inducibility of the atrial flutter in the presence of bi-directional isthmus block.
4. Athletes in whom anticoagulation is deemed necessary cannot participate in competitive sports where the danger of bodily collision is present.

Atrial fibrillation (in the absence of WPW syndrome). Atrial fibrillation is far more common than atrial flutter and may be present intermittently or chronically (11,12). Evaluation should include a search for the cause, such as thyrotoxicosis. More often atrial fibrillation occurs in association with diseases such as CAD or hypertension. Evaluation includes determination of the ventricular response during athletic activity or an exercise test comparable to the intended athletic competition. For some patients with paroxysmal atrial fibrillation, electrical induction of atrial fibrillation before the exercise test may be necessary, recognizing the difficult logistics in doing this. A 12-lead ECG is necessary, and long-term 24-h ECG recordings and an echocardiogram are helpful in establishing the presence of structural heart disease. Asymptomatic athletes who have episodes of atrial fibrillation of 5 to 15 s that do not increase in duration during exercise can participate in all sports.

Recommendations:

1. Athletes with asymptomatic atrial fibrillation in the absence of structural heart disease who maintain a ventricular rate that increases and slows appropriately and is comparable to that of a normal sinus response in relation to the level of activity, while receiving no therapy or therapy with AV nodal-blocking drugs, can participate in all competitive sports. Note that the use of beta-blockers is prohibited in some competitive sports.
2. Athletes who have atrial fibrillation in the presence of structural heart disease who maintain a ventricular rate comparable to that of an appropriate sinus tachycardia during physical activity while receiving no therapy or therapy with AV nodal-blocking drugs can participate in sports consistent with the limitations of the structural heart disease.
3. Athletes who require anticoagulation should not participate in sports with danger of bodily collision (13).
4. Athletes without structural heart disease who have elimination of atrial fibrillation by an ablation technique, including surgery, may participate in all com-

petitive sports after four to six weeks without a recurrence or after an electrophysiologic study has confirmed non-inducibility.

Sinus node re-entry, inappropriate sinus tachycardia, and atrial tachycardia (in the absence of WPW syndrome). Sinus node re-entry, inappropriate sinus tachycardia, and atrial tachycardia should be evaluated as described for atrial flutter. Asymptomatic athletes who have episodes of tachycardia of 5 to 10 s that do not increase in duration during exercise can participate in all sports.

Recommendations:

1. Athletes with sinus node re-entry, inappropriate sinus tachycardia, or atrial tachycardia in the absence of structural heart disease who maintain a ventricular rate that increases and slows appropriately and is comparable to that of a normal sinus response in relation to the level of activity, with or without therapy, can participate in all competitive sports.
2. Athletes with underlying structural heart disease can participate only in competitive sports consistent with the limitations of the heart disease.
3. Athletes without structural heart disease who have elimination of the atrial tachyarrhythmia by an ablation technique, including surgery, may participate in all competitive sports after two to four weeks without a recurrence or in several days after an electrophysiologic study showing non-inducibility.

AV junctional escape beats/rhythm. Atrioventricular junctional escape beats and junctional rhythm are common in athletes. The clinical approach and final recommendations are the same as those given earlier for symptomatic athletes with disturbances of sinus node function.

Premature AV junctional complexes. If the athlete is asymptomatic except for occasional episodes of palpitations that do not suggest a sustained tachycardia, evaluation need include only a 12-lead ECG. In some athletes, a 24-h ECG recording (during athletic activity if possible), echocardiogram, and an exercise test may be indicated.

Recommendations:

1. Athletes with a structurally normal heart and a normal heart rate response to activity without evidence of a sustained tachycardia can participate in all competitive sports.
2. Athletes with an abnormal heart, depending on the type and extent of the heart disease, can participate in competitive sports consistent with the limitations of the structural cardiac disease.

Non-paroxysmal AV junctional tachycardia. A form of junctional tachycardia, so-called junctional ectopic tachycardia, can be found in a permanent form in infants less than six months old, whereas a transient form occurs mostly in children but occasionally in adults (13). Most adults have a

slower form of non-paroxysmal junctional tachycardia. Evaluation generally includes a 12-lead ECG, echocardiogram, exercise test, and 24-h ECG recording during activity. Invasive studies may be necessary for some symptomatic patients or for those with very rapid ventricular rates.

Recommendations:

1. Athletes without structural heart disease or symptoms who have a controlled ventricular rate that increases and slows appropriately and is comparable to that of a normal sinus response in relation to the level of activity, with or without therapy, can participate in all sports.
2. Athletes who have no symptoms but who have structural heart disease or incompletely controlled ventricular rates can engage in class IA competitive sports depending on the nature and extent of the structural heart disease and the ventricular rate.
3. Athletes with inappropriately rapid ventricular rates, with or without structural heart disease, should be considered for treatment to control the ventricular rate before participating in any sports. Athletes whose tachycardia is controlled by therapy and verified by appropriate testing can participate in all competitive sports consistent with their cardiac status.

Supraventricular tachycardia. Atrioventricular nodal re-entrant tachycardia and AV re-entry over a concealed accessory pathway (with only retrograde conduction) (4,14) are included in this category. If the diagnosis of the supraventricular tachycardia cannot be made with certainty, and if other clinical therapeutic circumstances warrant it, invasive electrophysiologic studies might be indicated. It is important to identify the rate response of the supraventricular tachycardia during exercise. If the exercise does not induce the tachycardia, attempts to induce the supraventricular tachycardia (possibly with atrial or esophageal pacing) may be useful, followed once again by an exercise test performed by the athlete when the supraventricular tachycardia has been initiated. The logistics to accomplish this may be difficult. Asymptomatic athletes who have episodes of supraventricular tachycardia of 5 to 10 s that do not increase in duration during exercise can participate in all sports.

Recommendations:

1. Athletes without structural heart disease who are asymptomatic and have reproducible exercise-induced supraventricular tachycardia prevented by therapy and verified by appropriate testing can participate in all competitive sports.
2. Athletes who do not have exercise-induced supraventricular tachycardia but experience sporadic recurrences should be treated. However, because of the unpredictable nature of the tachycardia, end points

for adequate therapy may be difficult to achieve; but once established, these athletes can participate in all activities consistent with their cardiac status. Asymptomatic athletes who have episodes of supraventricular tachycardia of 5 to 15 s that do not increase in duration during exercise can participate in all sports consistent with their cardiac status.

3. Athletes with syncope, near-syncope, or significant symptoms secondary to arrhythmia or who have significant structural heart disease in addition to the arrhythmia should not participate in any competitive sports until they have been adequately treated and have no recurrence for two to four weeks (4). At that time they can participate in class IA competitive sports.
4. For those athletes with no structural heart disease who have had successful catheter or surgical ablation, are asymptomatic, and have no inducible arrhythmia on follow-up electrophysiologic testing, all competitive sports are permitted in several days. If no electrophysiologic testing is done, full participation is permitted if no spontaneous recurrence of tachycardia for two to four weeks after ablation.

Ventricular pre-excitation (WPW syndrome). Required noninvasive tests include a 12-lead ECG, exercise test, and echocardiogram to exclude associated cardiovascular abnormalities. In some instances, a 24-h ECG recording during athletic activity may be indicated. Electrophysiologic studies are indicated in athletes with symptoms of impaired consciousness, long-lasting palpitations, or rapid rates in whom an ablation procedure is indicated.

In asymptomatic athletes with no history of palpitations or tachycardia and no evidence of structural cardiac abnormalities, further evaluation may not be necessary. However, the optimal management for these athletes is uncertain, and continues to be debated (15,16). Furthermore, the younger the patient, the less time the patient has had to develop symptoms, and so the distinction between symptomatic and asymptomatic WPW syndrome may be less meaningful in the pediatric population. Sudden death in athletes with pre-excitation is rare, and it appears to be confined largely to those with accessory pathways that have short refractory periods. Therefore, it may be advisable in selected asymptomatic athletes who anticipate moderate or high level activity to undergo electrophysiology study to determine the anterograde refractory period of the accessory pathway, the minimum RR interval between pre-excited complexes in atrial fibrillation, and the number of accessory pathways. Individuals with multiple accessory pathways or ventricular rates exceeding 240 beats/min should be offered catheter ablation of the accessory pathway (17,18). For those athletes with a history of palpitations, syncope, or near syncope, it is mandatory to assess the functional capabilities and electrophysiologic properties of the accessory pathway.

Recommendations:

1. Athletes without structural heart disease, without a history of palpitations, or without tachycardia (particularly those 20 to 25 years old or more) can participate in all competitive sports. However, in younger age groups, a more in-depth evaluation including an electrophysiologic study may be recommended before allowing participation in moderate- to high-intensity competitive sports.
2. Athletes with episodes of AV reciprocating tachycardia should be treated as previously recommended (see section on Supraventricular Tachycardia). However, it should be appreciated that they can develop atrial fibrillation with rapid ventricular rates. Electrical induction of atrial fibrillation to determine the shortest QRS interval between two complexes conducted over the accessory pathway during isoproterenol administration or exercise is recommended. Those athletes in whom the shortest cycle length is less than 250 ms should undergo ablation of the accessory pathway.
3. Athletes with episodes of atrial flutter/fibrillation and syncope or near syncope whose maximal ventricular rate at rest (without therapy) as a result of conduction over the accessory pathway exceeding 240 beats/min should be considered for catheter ablation therapy of the accessory pathway prior to continuing competition. Those whose ventricular rate during isoproterenol administration is less than 240 beats/min and who have no episodes of syncope or near syncope appear to be at low risk for sudden cardiac death.
4. Athletes with no structural heart disease who have had successful catheter or surgical ablation of the accessory pathway, are asymptomatic, and have normal AV conduction and no inducible arrhythmia by follow-up electrophysiologic study can participate in all competitive sports in several days. Those without an electrophysiologic study and no spontaneous recurrence of tachycardia for two to four weeks after ablation can participate in all competitive sports.

Premature ventricular complexes. Noninvasive tests recommended include a 12-lead ECG and exercise test. If there is evidence to suggest the presence of structural heart disease, an echocardiogram is indicated, and a 24-h ECG recording may be beneficial. Even without evidence of structural heart disease, if an increase in the number of premature ventricular complexes or complex ventricular arrhythmias occurs during exercise (with or without treatment, or substantial reduction or abolition following a period of deconditioning), further evaluation may be indicated. In some of these athletes thought to have a structurally normal heart, cardiac catheterization and angiography may reveal otherwise undetected abnormalities, including occult CAD, congenital coronary anomalies, ARVC, car-

diac tumor, or evidence of cardiomyopathy. The more recently recognized channelopathy known as CPVT should be considered (see the following text).

Frequent and complex ventricular tachyarrhythmias are common in trained athletes; they are usually unassociated with underlying cardiovascular abnormalities and do not appear to convey increased risk (18). Deconditioning usually results in loss or diminution of these arrhythmias, providing evidence of their benign clinical nature (19).

Recommendations:

1. Athletes *without* structural heart disease who have premature ventricular complexes at rest and during exercise, and exercise testing (comparable to the sport in which they compete) can participate in all competitive sports. Should the premature ventricular complexes increase in frequency during exercise or exercise testing to the extent that they produce symptoms of impaired consciousness, significant fatigue, or dyspnea, the athlete can participate in class IA competitive sports only.
2. Athletes *with* structural heart disease who are in high-risk groups and have premature ventricular complexes (with or without treatment) can participate in class IA competitive sports only. Such athletes with premature ventricular complexes that are suppressed by drug therapy (as assessed by ambulatory ECG recordings) during participation in the sport can compete in only class IA competitive sports.

Ventricular tachycardia (VT). Nonsustained or sustained monomorphic or polymorphic VT is always a potentially serious occurrence. Noninvasive tests to be performed include a 12-lead ECG, exercise test, and echocardiography. In some patients, 24-h ECG recording during exercise may be indicated. Echocardiography, cardiac catheterization, and electrophysiologic study should be considered to verify that the heart is structurally normal and to establish the mechanism or location, or both, of the VT. A possible exception is the patient with accelerated idioventricular rhythm, in which the ventricular rate is similar to the sinus rate. In such patients, if they have no significant structural heart disease, the approach should be similar to that in individuals with premature ventricular complexes.

Recommendations:

1. Athletes with a structurally normal heart and monomorphic nonsustained or sustained VT that can be localized to a specific site(s) in the heart are candidates for a catheter ablation procedure that may potentially offer a cure. Following such a successful ablation procedure, with subsequent failure to induce VT during electrophysiologic study (EPS) with/without isoproterenol when the VT was reproducibly induced before ablation, the athlete can resume full

competitive activity within two to four weeks. A more conservative approach is recommended for the athlete who chooses drug suppression because catecholamines released during athletic activity can counter the suppressive effects of the drug, and the VT can re-emerge. In that situation, generally the athlete should not compete in any sports for at least two to three months after the last VT episode. If there have been no clinical recurrences, and the VT is not inducible by exercise/exercise testing or EPS, and the athlete has no structural heart disease, all competitive sports may be permitted. Because deconditioning can result in the loss or lessening of ventricular arrhythmias (19), a short period of deconditioning and retesting can be considered in some athletes.

2. For the athlete with structural heart disease and VT, moderate- and high-intensity competition is contraindicated regardless of whether the VT is suppressed or ablated. Only class IA competitive sports are permitted.
3. An exception to this general recommendation is the asymptomatic athlete with brief (generally less than 8 to 10 consecutive ventricular beats) episodes of non-sustained monomorphic VT, rates generally less than 150 beats/min, and no structural heart disease established by noninvasive and invasive tests. These athletes do not appear to be at increased risk for sudden cardiac death. If exercise testing (preferably by ambulatory ECG recording during the specific competitive activity) demonstrates suppression of the VT or no significant worsening compared with baseline, participation in all competitive sports is permissible.
4. The desire of the athlete to continue athletic competition should not represent the primary indication for use of an implantable cardioverter-defibrillator (ICD). The efficacy with which these devices will terminate a potentially lethal arrhythmia under the extreme conditions of competitive sports, with the associated metabolic and autonomic changes, and possible myocardial ischemia, is unknown. In addition, sports with physical contact may result in damage to the ICD and/or lead, preventing normal function. For athletes with ICDs, all moderate and high intensity sports are contraindicated. Class IA sports are permitted.

Ventricular flutter and ventricular fibrillation.

Recommendation:

1. Athletes with conditions that result in cardiac arrest in the presence or absence of structural heart disease generally are treated with an ICD and cannot participate in any moderate- or high-intensity competitive sports. However, athletes with ICDs and who have had no episodes of ventricular flutter or ventricular fibrillation requiring device therapy for six months

may engage in class IA competitive sports. Recommendations in the section on VT also apply.

First-degree AV block. In asymptomatic athletes with structurally normal hearts, if the QRS complex is normal, no further evaluation other than a 12-lead ECG is necessary. If the QRS complex is abnormal, or the PR interval is excessively prolonged (0.3 s or more), an exercise stress test, 24-h ECG recording and echocardiogram may be indicated. Possibly an EPS might be necessary to determine the site and duration of conduction delay.

Recommendation:

1. Asymptomatic athletes without evidence of structural heart disease, in whom the first-degree AV block does not worsen with exercise, can participate in all competitive sports. If underlying heart disease is present, its nature and severity can independently dictate alternative restrictions.

Type 1 second-degree (Wenckebach) AV block. Wenckebach AV nodal block can be present in otherwise normal, well-trained endurance athletes (4). Recommended evaluations include a 12-lead ECG, exercise test, and echocardiogram. A 24-h ECG recording during athletic activity may be indicated in some athletes. In those athletes with type 1 second-degree AV block and co-existing bundle-branch block, EPS may be indicated to identify the presence of His-Purkinje Wenckebach block.

Recommendations:

1. Asymptomatic athletes with a structurally normal heart and no worsening or actual improvement of AV block with exercise or recovery can participate in all competitive sports.
2. Asymptomatic athletes with a structurally abnormal heart in whom AV block disappears or does not worsen with exercise or recovery can participate in all competitive sports, as determined by the limitations of the cardiac abnormality.
3. Asymptomatic athletes in whom type 1 second-degree AV block initially appears or worsens with exercise or during the recovery period should be evaluated further (e.g., for possible intra- or infra-His block) and may require pacemaker therapy. Such athletes can participate in class IA competitive sports.
4. Athletes treated with pacemakers should not engage in competitive sports with a danger of bodily collision because such trauma may damage the pacemaker system.

Type 2 second-degree (Mobitz) AV block. The evolution and treatment of this abnormality is considered to be the same as in acquired complete heart block. It should be treated with permanent pacing before any athletic activity. Athletes with pacemakers should not participate in compet-

itive sports that pose a danger of bodily collision because such trauma may damage the pacemaker system. Before allowing athletes to engage in these activities, an exercise test should be done at the level of activity demanded by the particular sport to be certain that the paced heart rate increases appropriately.

Congenital complete heart block. The clinical approach to evaluating the severity of the cardiovascular abnormality includes an echocardiogram, 12-lead ECG, 24-h ECG recording during exercise, and exercise stress test (exercise testing should be performed at the same exercise level as that during the sports activity).

Recommendations:

1. Athletes with a structurally normal heart and normal cardiac function, with no history of syncope or near syncope, a narrow QRS complex, ventricular rates at rest greater than 40 to 50 beats/min increasing appropriately with exertion, no or only occasional premature ventricular complexes, and no VT during exertion can participate in all competitive sports.
2. Athletes with ventricular arrhythmia, symptoms of fatigue, near-syncope, or syncope should have a pacemaker implanted before they participate in competitive sports. Athletes with pacemakers should not participate in competitive sports when the danger of bodily collision exists because such trauma may damage the pacemaker system. Before allowing athletes to engage in these activities, an exercise test should be conducted at the level of activity demanded by the particular sport so as to be certain that the paced heart rate increases appropriately.
3. Athletes with abnormal hemodynamic status, as those with an intracardiac shunt, cannot participate in any competitive sports without a pacemaker. Restrictions are the same as those in recommendation 2.

Acquired complete heart block.

Recommendations:

1. Patients with acquired complete heart block should be treated with pacing before any athletic activity.
2. Athletes with a pacemaker should not participate in competitive sports posing a danger of bodily collision because such trauma may damage the pacemaker system.

Complete right bundle-branch block. Evaluation includes a 12-lead ECG, exercise test, and echocardiogram. In some instances, a 24 h ECG recording may be indicated.

Recommendation:

1. Athletes without ventricular arrhythmias who do not develop AV block with exercise and who have no symptoms can participate in all competitive sports

consistent with their cardiac status. This also applies to athletes with associated left-axis deviation.

Complete left bundle-branch block. Evaluation includes a 12-lead ECG, exercise test, and echocardiogram. In some instances, a 24-h ECG recording may be indicated. Because of the rarity of acquired left bundle-branch block in children and its association with syncope from presumed paroxysmal AV block, an invasive EPS should be considered in young patients.

Recommendations:

1. Adult athletes with acquired left bundle-branch block should follow the recommendations under the section entitled Complete Right Bundle-Branch Block.
2. Athletes with a normal HV interval and a normal AV conduction response to pacing can participate in all competitive sports consistent with their cardiac status.
3. Athletes with abnormal AV conduction characterized by an HV interval greater than 90 ms or a His-Purkinje block should have pacemaker implantation. They should be restricted from competitive sports that hold a danger of bodily collision because such trauma may damage the pacemaker system.

Inherited arrhythmia syndromes. LONG QT SYNDROME. The definitive clinical diagnosis of congenital long QT syndrome (LQTS) can be complex (20). Debate continues as to what QTc constitutes the upper limit of normal. An increasing proportion of asymptomatic individuals with genetically proven LQTS are found to have a normal resting ECG with a heart rate corrected QT interval (QTc) by Bazett's formula of less than 460 ms (genotype positive/phenotype negative LQTS). In addition, a QTc of 440 ms, used in the past as an upper limit of normal, is present in far too many normal individuals (greater than 25%) to serve as a meaningful upper limit cut-off value. In general, a QTc of 470 ms or more in males and 480 ms or more in females requires further investigation as to the presence of congenital (or acquired) causes of QT prolongation. A patient with LQTS and a resting QTc of 500 ms or more is generally considered at increased clinical risk for a significant arrhythmia (21). One approach to the diagnosis of congenital LQTS is to utilize the "Priori-Schwartz" score that incorporates QTc, T-wave morphology, symptomatic presentation, and family history into the diagnostic algorithm (21). A "Priori-Schwartz" score of 4 or more suggests high clinical probability for LQTS. In addition, genetic testing for the five cardiac ion-channel genes responsible for 75% of LQTS (LQT1, LQT2, LQT3, LQT5, and LQT6) is now available as a commercial diagnostic test (22).

Mutations involving the structural protein ankyrin-B underlie the rare LQT4 form (23). Mutations involving the *KCNJ2*-encoded IK1 potassium channel account for approximately one-half of Andersen-Tawil syndrome (ATS1)

characterized by abnormal U waves, a prolonged QU interval, periodic paralysis, and facial and skeletal dysmorphisms. The ATS1 has been annotated in the past as LQT7.

Physical exertion (particularly swimming) appears to be a common trigger for ventricular arrhythmias in LQT1, whereas individuals with LQT2 seem more at-risk to auditory/emotional triggers, and patients with LQT3 may be at greater risk during rest and inactivity (24,25). However, exceptions to these genotype-phenotype correlations hinder genotype-specific tailoring of competitive sports recommendations. The entire personal and family phenotype must be incorporated before any eligibility or disqualification decision is rendered.

Recommendations:

1. Regardless of QTc or underlying genotype, all competitive sports, except those in class IA category should be restricted in a patient who has previously experienced either: 1) an out-of-hospital cardiac arrest, or 2) a suspected LQTS-precipitated syncopal episode.
2. Asymptomatic patients with baseline QT prolongation (QTc of 470 ms or more in males, 480 ms or more in females) should be restricted to class IA sports. The restriction limiting participation to class IA activities may be liberalized for the asymptomatic patient with genetically proven type 3 LQTS (LQT3).
3. Patients with genotype-positive/phenotype-negative LQTS (i.e., identification of a LQTS-associated mutation in an asymptomatic individual with a nondiagnostic QTc) may be allowed to participate in competitive sports. Although the risk of sudden cardiac death is not zero in such individuals, there is no compelling data available to justify precluding these individuals (who are being identified with increasing frequency) from competitive activities. Because of the strong association between swimming and LQT1, persons with genotype-positive/phenotype-negative LQT1 should refrain from competitive swimming.
4. LQTS patients with an ICD/pacemaker should not engage in sports with a danger of bodily collision because such trauma may damage the pacemaker system. The presence of an ICD should restrict individuals to class IA activities.

SHORT QT SYNDROME. Individuals with the short QT syndrome (SQTS) (QTc less than 300 ms) have a short QT interval and ventricular refractory period, and at least some of them have “gain-of-function” abnormalities in either I_{Kr} (*KCNH2*) or I_{Ks} (*KCNQ1*) (26).

Recommendation:

1. Until the phenotype of SQTS is better understood, a universal restriction from competitive sports with the

possible exception of class IA activities seems to represent the most prudent recommendation (27).

CATECHOLAMINERGIC POLYMORPHIC VENTRICULAR TACHYCARDIA (CPVT). Approximately one-half of patients with CPVT is pursuant to mutations involving the *RyR2*-encoded ryanodine receptor (sarcoplasmic reticulum calcium release channel). Such individuals are vulnerable to exercise-induced VT/ventricular fibrillation.

Recommendations:

1. Symptomatic patients have a poor prognosis unless treated with an implantable cardioverter-defibrillator (ICD) (28) and all such patients are restricted from competitive sports with the possible exception of minimal contact, class IA activities. As with LQT1, patients with CPVT should be restricted from competitive swimming. Asymptomatic patients detected as part of familial screening with documented exercise- or isoproterenol-induced VT should refrain from all competitive sports except possible class IA activities. A less restrictive approach may be possible for the genotype-positive/phenotype-negative (asymptomatic, no inducible VT) athlete.

BRUGADA SYNDROME (BrS). Brugada syndrome (29), characterized by an accentuated J-wave primarily in leads V_1 through V_3 , with ST-segment elevation, often followed by a negative T-wave and an R prime, may be the cause of sudden unexplained death syndrome, typically during sleep. Only 15% to 20% of BrS is established as a channelopathy due to mutations involving the *SCN5A*-encoded alpha subunit of the cardiac sodium channel (30). Individuals with BrS and no previous cardiac arrests may have a high risk of sudden death if they have inducibility of ventricular arrhythmias and a previous history of syncope (28). Hyperthermia can potentially unmask the Brugada ECG pattern in patients with BrS, who can then display fever-induced polymorphic VT. Death often occurs with mild activity or during sleep.

Recommendations:

1. Although a clear association between exercise and sudden death has not been established, and because of the potential impact of hyperthermia, restriction to participation in class IA sports seems advisable.
2. The presence of an ICD device warrants the same restrictions to class IA sports as previously outlined.

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TASK FORCE 7 REFERENCES

1. Maron BJ. Sudden death in young athletes. *N Engl J Med* 2003;349:1064–75.
2. Zehender M, Meinertz T, Keul J, Just H. ECG variants and cardiac arrhythmias in athletes: clinical relevance and prognostic importance. *Am Heart J* 1990;119:1378–91.

3. Bjornstad H, Storstein L, Meen HD, Hals O. Ambulatory electrocardiographic findings in top athletes, athletic students and control subjects. *Cardiology* 1994;84:42–50.
4. Olgin JE, Zipes DP. Specific arrhythmias: diagnosis and treatment. In: Zipes DP, Libby P, Bonow RO, Braunwald E, editors. *Heart Disease: A Textbook of Cardiovascular Medicine*. Philadelphia, PA: Saunders, 2005:803–63.
5. Allesie M, Ausma J, Schotten U. Electrical, contractile and structural remodeling during atrial fibrillation. *Cardiovasc Res* 2002;54:230–46.
6. Antzelevitch C. Molecular genetics of arrhythmias and cardiovascular conditions associated with arrhythmias. *J Cardiovasc Electrophysiol* 2003;14:1259–72.
7. Calkins H, Zipes DP. Hypotension and syncope. In: Zipes DP, Libby P, Bonow RO, Braunwald E, editors. *Heart Disease. A Textbook of Cardiovascular Medicine*. Philadelphia, PA: Saunders, 2005:909–19.
8. Ackerman MJ, Khositseth A, Tester DJ, Hejlik JB, Shen WK, Porter CB. Epinephrine-induced QT interval prolongation: a gene-specific paradoxical response in congenital long QT syndrome. *Mayo Clin Proc* 2002;77:413–21.
9. Schott JJ, Alshinawi C, Kyndt F, et al. Cardiac conduction defects associated with mutations in SCN5A. *Nat Genet* 1999;23:20–1.
10. Benson DW, Wang DW, Dymant M, et al. Congenital sick sinus syndrome caused by recessive mutations in the cardiac sodium channel gene (SCN5A). *J Clin Invest* 2003;112:1019–28.
11. Nattel S, Erlich J. Atrial fibrillation. In: Zipes D, Jalife J, editors. *Cardiac Electrophysiology: From Cell to Bedside*. Philadelphia, PA: Saunders, 2004:512–22.
12. Furlanello F, Bertoldi A, Dallago M, et al. Atrial fibrillation in elite athletes. *J Cardiovasc Electrophysiol* 1998;9:S63–8.
13. Oral H, Strickberger SA. Junctional rhythms and junctional tachycardia. In: Zipes D, Jalife J, editors. *Cardiac Electrophysiology: From Cell to Bedside*. Philadelphia, PA: Saunders, 2004:523–7.
14. Lockwood D, Otomoto K, Wang Z. Electrophysiologic characteristics of atrioventricular nodal reentrant tachycardia. In: Zipes D, Jalife J, editors. *Cardiac Electrophysiology: From Cell to Bedside*. Philadelphia, PA: Saunders, 2004:537–57.
15. Pappone C, Santinelli V, Rosanio S, et al. Usefulness of invasive electrophysiologic testing to stratify the risk of arrhythmic events in asymptomatic patients with Wolff-Parkinson-White pattern: results from a large prospective long-term follow-up study. *J Am Coll Cardiol* 2003;41:239–44.
16. Klein GJ, Bashore TM, Sellers TD, Pritchett EL, Smith WM, Gallagher JJ. Ventricular fibrillation in the Wolff-Parkinson-White syndrome. *N Engl J Med* 1979;301:1080–5.
17. Pappone C, Santinelli V, Manguso F, et al. A randomized study of prophylactic catheter ablation in asymptomatic patients with the Wolff-Parkinson-White syndrome. *N Engl J Med* 2003;349:1803–11.
18. Biffi A, Pelliccia A, Verdile L, et al. Long-term clinical significance of frequent and complex ventricular tachyarrhythmias in trained athletes. *J Am Coll Cardiol* 2002;40:446–52.
19. Biffi A, Maron BJ, Verdile L, et al. Impact of physical deconditioning on ventricular tachyarrhythmias in trained athletes. *J Am Coll Cardiol* 2004;44:1053–8.
20. Ackerman MJ. Cardiac channelopathies: it's in the genes. *Nat Med* 2004;10:463–4.
21. Priori SG, Schwartz PJ, Napolitano C, et al. Risk stratification in the long-QT syndrome. *N Engl J Med* 2003;348:1866–74.
22. Genaisance Pharmaceuticals, Inc. Genaisance Pharmaceuticals, Inc. (GNSC) launches its proprietary FAMILION test for genetic mutations associated with sudden cardiac death. Available at: http://www.biospace.com/news_story.cfm?StoryID=16229920&full=1. Accessed October 1, 2004.
23. Mohler PJ, Schott JJ, Gramolini AO, et al. Ankyrin-B mutation causes type 4 long-QT cardiac arrhythmia and sudden cardiac death. *Nature* 2003;421:634–9.
24. Schwartz PJ, Priori SG, Spazzolini C, et al. Genotype-phenotype correlation in the long-QT syndrome: gene-specific triggers for life-threatening arrhythmias. *Circulation* 2001;103:89–95.
25. Choi G, Kopplin LJ, Tester DJ, Will ML, Hauglund CM, Ackerman MJ. Spectrum and frequency of cardiac channel defects implicated in swimming-triggered arrhythmia syndromes. *Circulation* 2004;110:2119–24.
26. Brugada R, Hong K, Dumaine R, et al. Sudden death associated with short-QT syndrome linked to mutations in HERG. *Circulation* 2004;109:30–5.
27. Gaita F, Giustetto C, Bianchi F, et al. Short QT syndrome: a familial cause of sudden death. *Circulation* 2003;108:965–70.
28. Sumitomo N, Harada K, Nagashima M, et al. Catecholaminergic polymorphic ventricular tachycardia: electrocardiographic characteristics and optimal therapeutic strategies to prevent sudden death. *Heart* 2003;89:66–70.
29. Brugada J, Brugada R, Brugada P. Determinants of sudden cardiac death in individuals with the electrocardiographic pattern of Brugada syndrome and no previous cardiac arrest. *Circulation* 2003;108:3092–6.
30. Brugada P, Brugada R, Mont L, Rivero M, Geelen P, Brugada J. Natural history of Brugada syndrome: the prognostic value of programmed electrical stimulation of the heart. *J Cardiovasc Electrophysiol* 2003;14:455–7.

Appendix 1. Author Relationships With Industry and Others

Name	Consultant	Research Grant	Scientific Advisory Board	Stock Holder	Expert Witness Testimony
Dr. Michael J. Ackerman	None	None	None	None	None
Dr. N. A. Mark Estes III	<ul style="list-style-type: none"> • Guidant • Medtronic 	<ul style="list-style-type: none"> • Guidant • Medtronic 	<ul style="list-style-type: none"> • Guidant (Executive Committee) 	None	None
Dr. Augustus O. Grant	None	None	None	None	None
Dr. Robert J. Myerburg	<ul style="list-style-type: none"> • Guidant • Procter & Gamble 	None	<ul style="list-style-type: none"> • Procter & Gamble • Reliant Pharmaceutical 	None	<ul style="list-style-type: none"> • 2000, Defense, Lewis vs. Mudge • 2002, Defense, Weiner vs. Vitello • 2005, Defense, Ephedra Multi-District Litigation
Dr. George Van Hare	None	<ul style="list-style-type: none"> • Medtronic • St. Jude Medical 	None	None	None
Dr. Douglas P. Zipes	<ul style="list-style-type: none"> • Cardiofocus • Janssen • Medtronic 	<ul style="list-style-type: none"> • Medtronic 	<ul style="list-style-type: none"> • Medtronic 	<ul style="list-style-type: none"> • MVMD 	<ul style="list-style-type: none"> • 1996, Defense, Knapp vs. Northwestern

Task Force 8: Classification of Sports

Jere H. Mitchell, MD, FACC, *Chair*

William Haskell, PhD, Peter Snell, PhD, Steven P. Van Camp, MD, FACC

This classification of sports has been developed to allow a fundamental question to be addressed: whether it is reasonably safe to recommend that an athlete with a specific cardiovascular abnormality be eligible for a particular competitive sport (1,2). We recognize that cardiovascular disease assessments are imprecise and may change over time and be influenced by exercise training. Furthermore, there are potentially life-threatening aspects to the nature of the risk involved. We have attempted to incorporate these realities into the classification system.

Sports can be classified according to the type and intensity of exercise performed and also with regard to the danger of bodily injury from collision, as well as the consequences of syncope. Exercise can be divided into two broad types: dynamic (isotonic) and static (isometric) (3-6).

Dynamic exercise involves changes in muscle length and joint movement with rhythmic contractions that develop a relatively small intramuscular force; static exercise involves development of a relatively large intramuscular force with little or no change in muscle length or joint movement. These two types of exercise should be thought of as the two opposite poles of a continuum, with most physical activities involving both static and dynamic components. For example, distance running has low static and high dynamic demands, water skiing has principally high static and low dynamic demands, and rowing has both high static and dynamic demands.

The terms *dynamic* and *static* exercise characterize activity on the basis of the mechanical action of the muscles involved and are different from the terms *aerobic* and *anaerobic* exercise. The latter characterize activity on the basis of the type of muscle metabolism. Most high-intensity static exercise is performed anaerobically, whereas high-intensity dynamic exercise lasting for more than several minutes is performed aerobically. However, some dynamic exercises, such as sprinting or jumping, are performed primarily anaerobically. Thus, many sports are placed in the high dynamic category, including such diverse activities as skiing (cross country), running (distance), soccer, and squash. Because the cardiovascular demands of very high resistance dynamic exercise are similar to sustained static exercise, those sports that have either a sustained static component or a very high resistance dynamic component are classified together as high-intensity static exercise (e.g., weightlifting, gymnastics, and field events [throwing]).

The two primary factors determining the cardiovascular risk of competitive sports are, clearly, the athlete's abnormality and the stress under which it is placed by the sport. This involves: 1) the specific cardiovascular diagnosis and its

pathophysiological consequences; and 2) the cardiovascular response to the demands of the sport during both competition and training, which a competitive athlete in a sport may typically or reasonably be expected to undertake. The stress of the sport involves both static and dynamic components that determine the cardiovascular demands of the sport. Thus, for athletes with cardiovascular abnormalities, recommendations regarding eligibility for competition will recognize these factors as well as the attendant psychological stresses that invariably accompany competitive athletics. The cardiovascular demands produced by training or competition in a particular sport involve the type, intensity, and duration of the activity, with both peak intensity and total work performed as well as attendant neurohumoral effects and environmental factors.

RESPONSE AND ADAPTATION TO EXERCISE

The acute responses of the cardiovascular system to dynamic and static exercise are summarized in Figure 1 (5,6). Dynamic exercise performed with a large muscle mass causes a marked increase in oxygen consumption (Fig. 1, Panel A). There is a substantial increase in cardiac output, heart rate, stroke volume, and systolic blood pressure; a moderate increase in mean arterial pressure; and a decrease in diastolic blood pressure. Also, there is a marked decrease in total peripheral resistance. Static exercise, in contrast, causes a small increase in oxygen consumption, cardiac output, and heart rate, and no change in stroke volume (Fig. 1, Panel B). Moreover, there is a marked increase in systolic, diastolic, and mean arterial pressure and no appreciable change in total peripheral resistance. Thus, dynamic exercise primarily causes a volume load on the left ventricle, whereas static exercise causes a pressure load. The cardiovascular responses during dynamic exercise of a small muscle mass at low resistance or during dynamic exercise of a large muscle mass at high resistance are similar to the responses during static exercise.

The acute response to both dynamic and static exercise changes several factors that are important in determining myocardial oxygen demand: heart rate, wall tension, and contractile state of the left ventricle (LV) (7,8). Wall tension is affected by pressure development and ventricular volume. In high-intensity dynamic exercise, there is a large increase in heart rate and an increase in stroke volume that is achieved by both an increase in end-diastolic volume (Frank-Starling mechanism) and a decrease in end-systolic volume (increased contractile state). In high-intensity static exercise, a smaller increase occurs in heart rate and little change occurs in end- and end-systolic volumes of the LV.

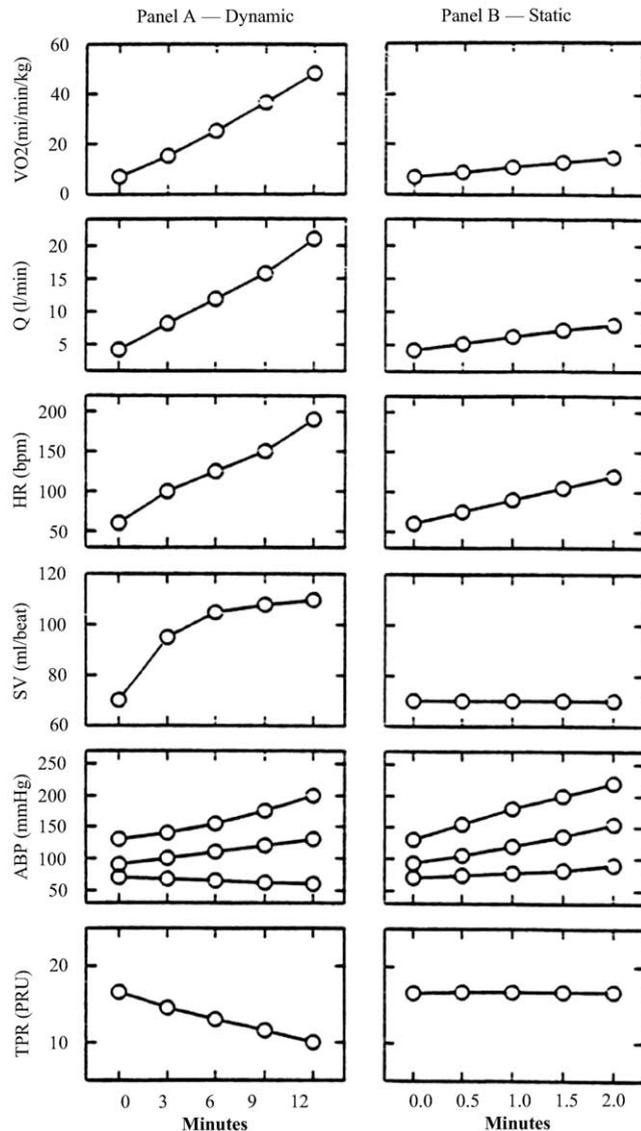


Figure 1. Cardiovascular response to exercise. (A) Response to dynamic exercise of progressively increasing workload to maximal oxygen consumption. (B) Response to a static handgrip contraction at 30% maximal voluntary contraction. ABP (mm Hg) = systolic, mean and diastolic arterial blood pressures; HR (bpm) = heart rate (beats/min); Q (l/min) = cardiac output (liters/min); SV (ml/beat) = stroke volume; TPR (PRU) = total peripheral resistance in peripheral resistance units; VO₂ (ml/min/kg) = oxygen consumption (ml/min × body weight in kg). Reprinted with permission from Mitchell JH, Raven PB. Cardiovascular adaptation to physical activity. In: Bouchard C, Shephard RJ, Stephens T, editors. Physical Activity, Fitness and Health: International Proceedings and Consensus Statement (Fig. 17.2). Champaign, IL: Human Kinetics Publishers. Copyright 1994 by Human Kinetics Publishers, Inc.

However, arterial pressure and contractile state of the ventricle are increased. Thus, both dynamic and static exercise increase factors that are important in determining myocardial oxygen demand.

The chronic adaptation of the cardiovascular system to repeated bouts of dynamic exercise (training) results in an increase in maximal oxygen uptake (5,6). This is due to an increased maximal stroke volume and arteriovenous oxygen difference. Athletes who train in sports with a high dynamic

component have a large absolute LV mass and chamber size (eccentric hypertrophy) (6,9,10). This eccentric hypertrophy develops gradually and correlates with a high maximal stroke volume and high maximal oxygen uptake. Also, the skeletal muscles involved in the dynamic exercise training become more oxidative and less glycolytic with an increase in the number and size of the mitochondria and an increased number of capillaries. These changes contribute to the larger maximal arteriovenous oxygen difference seen in endurance athletes.

The chronic adaptation of the cardiovascular system to static exercise training results in little or no increase in maximal oxygen uptake. However, athletes who participate in sports with a high static component also have a large LV mass but no increase in chamber size (concentric hypertrophy) (6,9,10). In addition, the skeletal muscles involved in the static exercise training become more glycolytic and less oxidative, and there is an increase in skeletal muscle mass primarily by fiber hypertrophy with a small degree of hyperplasia via stem cell activation.

ATHLETE'S HEART

As previously mentioned, participation in sports with a high dynamic demand (endurance) or with a high static demand (power) causes an increased cardiac mass and structural remodeling in many athletes. This finding has been shown in detail over three decades in a multitude of echocardiographic studies and more recently by cardiac magnetic resonance imaging (6,9,10). The changes resulting from training include enlargement and increased volume of the right and LV chambers, sometimes accompanied by increased thickness of the LV wall, and increased size of the left atrium, with preservation of systolic and diastolic function. Extreme changes in cavity dimensions and LV wall thickness are most commonly associated with training in rowing, cross-country skiing, cycling, and swimming, but paradoxically are uncommon as a consequence of training in ultra-endurance sports (10). The increased cardiac dimensions associated with athletic training are related to body surface area or lean body mass and consequently are less pronounced in women (10).

Participation in sports with a high static demand (e.g., weightlifting or wrestling) is associated with LV wall thickness that is usually normal in absolute terms (less than 12 mm) but disproportionately increased in relation to cavity size. More substantial LV wall thickness (13 mm or more in men and 12 mm or more in women), occasionally encountered in competitive athletes, creates a differential diagnosis with hypertrophic cardiomyopathy (9,10) (also see Task Force 1).

CLASSIFICATION OF SPORTS

A classification of sports is provided in Figure 2, which relates individual competitive sports to the two general types of exercise: dynamic and static (3-6). Each sport is catego-

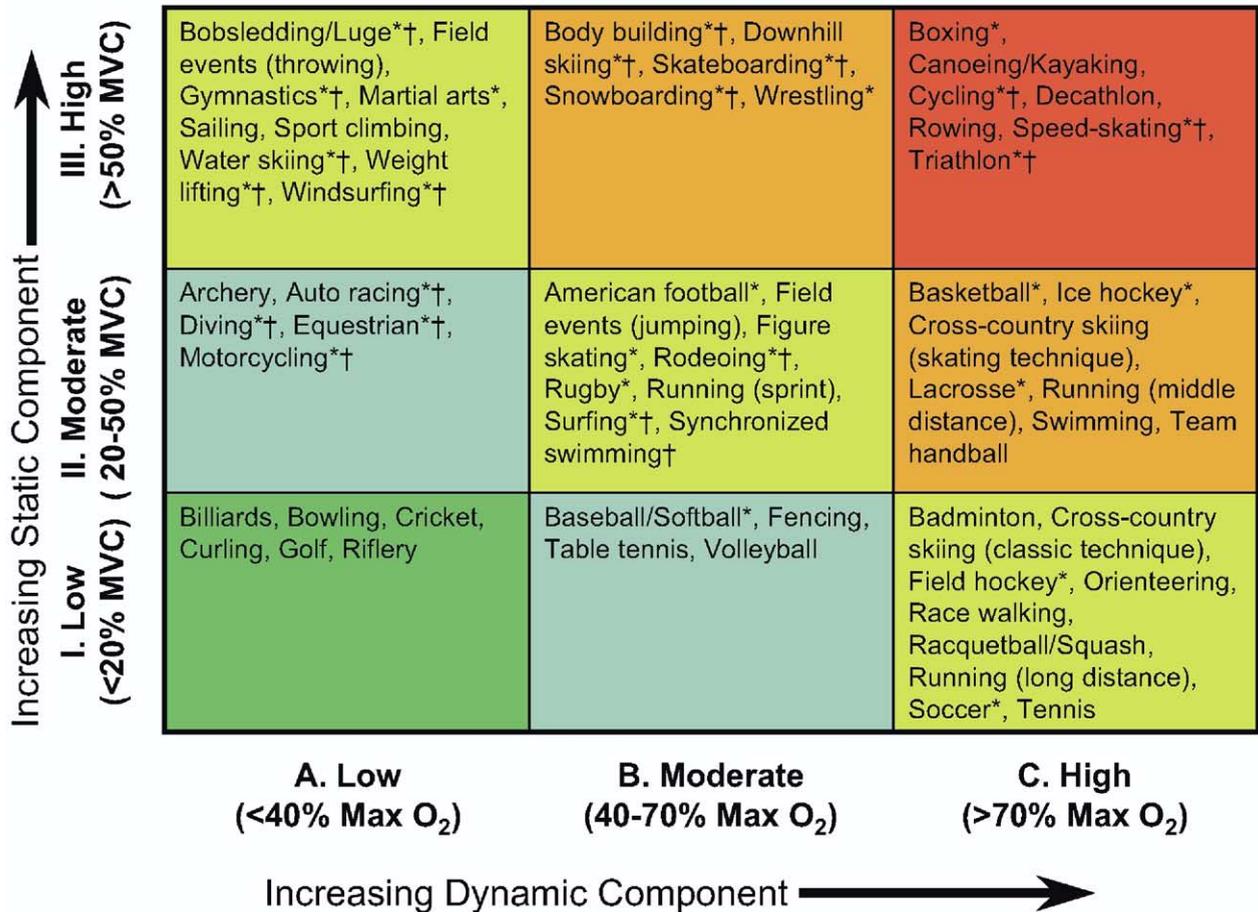


Figure 2. Classification of sports. This classification is based on peak static and dynamic components achieved during competition. It should be noted, however, that higher values may be reached during training. The increasing dynamic component is defined in terms of the estimated percent of maximal oxygen uptake (MaxO₂) achieved and results in an increasing cardiac output. The increasing static component is related to the estimated percent of maximal voluntary contraction (MVC) reached and results in an increasing blood pressure load. The lowest total cardiovascular demands (cardiac output and blood pressure) are shown in **green** and the highest in **red**. **Blue, yellow, and orange** depict low moderate, moderate, and high moderate total cardiovascular demands. *Danger of bodily collision. †Increased risk if syncope occurs.

ized by the level of intensity (low, medium, high) of dynamic or static exercise generally required to perform that sport during competition. It also recognizes those sports that pose significant risk due to bodily collision, either because of the probability of hard impact between competitors or between a competitor and an object, projectile, or the ground; as well as the degree of risk to the athlete or others if a sudden syncopal event occurs. Thus, in terms of their dynamic and static demands, sports can be classified (Fig. 2) as IIIC (high static, high dynamic), IIB (moderate static, moderate dynamic), IA (low static, low dynamic), and so forth. For example, an athlete with a cardiovascular abnormality that contraindicates a sport that produces a high pressure load on the LV may be advised to avoid sports classified as IIIA, IIIB, and IIIC. It should be emphasized that in terms of the classification of sports matrix presented in Figure 2, cardiovascular abnormalities designated as compatible with a high level of intensity in any particular category also (by definition) permit participation in levels of lesser intensity. For example, if class IC sports are appro-

priate (low static/high dynamic), then so are classes IA and IB (low static/low and moderate dynamic).

The sports matrix in Figure 2 should not be regarded as a rigid classification, but rather a spectrum in which some athletes in the same sport could possibly deserve placement in different categories. Furthermore, some sports involve heterogeneity with respect to static and dynamic cardiovascular demands in either different athletic disciplines—such as parallel bars and floor exercises in gymnastics or positions such as lineman and running back in football, or goalkeeper and mid-fielder in soccer. We have not formulated such distinctions in the matrix, but these should be taken into consideration when making clinical decisions regarding the eligibility and disqualification for competitive sports.

LIMITATIONS OF CLASSIFICATION

There are important limitations to the present classification of sports according to the type and intensity of exercise performed, as presented in Figure 2. For example, it does not consider the emotional stress that an athlete experiences

during a competitive event, the effects of environmental factors, electrolyte abnormalities, or the specific training regimen used by the athlete. Also, for team sports the classification is based on the highest cardiovascular demands that are experienced during competition and does not consider the different cardiovascular demands of specific positions.

During all athletic competitions, the athlete's emotional involvement can substantially increase sympathetic drive, and the resulting catecholamine concentrations can increase blood pressure, heart rate, and myocardial contractility, thereby increasing myocardial oxygen demand. Also, the increase in sympathetic tone can trigger arrhythmias and aggravate existing myocardial ischemia. Thus, even in competitive sports such as golf or riflery, which have low myocardial oxygen demands owing to the exercise required, substantial increases may occur because of emotional involvement during competition. This problem is difficult (if not impossible) to quantitate, but it needs to be considered in determining the eligibility for sports participation of athletes with existing cardiovascular abnormalities.

Environmental exposure during athletic competition or training also needs to be considered. Performance at high altitudes or under water may decrease oxygen availability, whereas excessively hot or cold temperatures and high humidity can increase myocardial workload for the same intensity of exercise. Another potentially relevant environmental factor is air pollution, such as elevated carbon monoxide levels in a sport such as auto racing.

With the modern application of exercise science to competitive sports, training for competition can, in fact, be more demanding on the cardiovascular system than the competition itself. Many training regimens now use heavy resistance weight training (high static and low dynamic demand) for increasing strength and power in sports that do not include heavy static demands during competition (e.g., tennis, basketball). This concept that both the dynamic and static demands of a sport may be greater during training than in competition must be seriously considered when the eligibility of an athlete in a given sport is being determined. Also, in some cases where it is found acceptable for the athlete to participate in the competitive aspect of a specific sport but the existing training program is considered too vigorous, it may be possible to modify the training regimen

so as to reduce the cardiovascular demands to an acceptable level.

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TASK FORCE 8 REFERENCES

1. Mitchell JH, Blomqvist CG, Haskell WL, et al. Classification of sports. 16th Bethesda Conference: cardiovascular abnormalities in the athlete: recommendations regarding eligibility for competition. *J Am Coll Cardiol* 1985;6:1198–9.
2. Mitchell JH, Haskell WL, Raven PB. Classification of sports. 26th Bethesda Conference: cardiovascular abnormalities in the athlete: recommendations for determining eligibility for competition in athletes with cardiovascular abnormalities. *J Am Coll Cardiol* 1994;24:864–6.
3. Asmussen E. Similarities and dissimilarities between static and dynamic exercise. *Circ Res* 1981;48 Suppl 1:I3–10.
4. Mitchell JH, Wildenthal K. Static (isometric) exercise and the heart: physiological and clinical considerations. *Annu Rev Med* 1974;25:369–81.
5. Mitchell JH, Raven PB. Cardiovascular adaptation to physical activity. In: Bouchard C, Shephard R, Stephen T, editors. *Physical Activity, Fitness, and Health: International Proceedings and Consensus Statement*. Champaign, IL: Human Kinetics, 1994:286–98.
6. Gallagher KM, Raven PB, Mitchell JH. Classification of sports and the athlete's heart. In: Williams RA, editor. *The Athlete and Heart Disease: Diagnosis, Evaluation and Management*. Philadelphia, PA: Lippincott Williams & Wilkins, 1999:9–21.
7. Sonnenblick EH, Ross JJ, Braunwald E. Oxygen consumption of the heart: newer concepts of its multifactorial determination. *Am J Cardiol* 1968;22:328–36.
8. Mitchell JH, Hefner LL, Monroe RG. Performance of the left ventricle. *Am J Med* 1972;53:481–94.
9. Pelliccia A, Maron BJ, Spataro A, Proschan MA, Spirito P. The upper limit of physiologic cardiac hypertrophy in highly trained elite athletes. *N Engl J Med* 1991;324:295–301.
10. Maron BJ. Sudden death in young athletes. *N Engl J Med* 2003;349:1064–75.

Appendix 1

Dr. Steven P. Van Camp declared that he served as an expert witness for the following court cases which may be related to the 36th Bethesda Conference: 2001—Defense, young athlete with cardiomyopathy dying suddenly; 2002—Defense, ephedra ingestion and exercise-related death; 2003—Defense, two cases of ephedra ingestion and exercise-related death; 2005—Defense, ephedra ingestion and exercise-related death; Defense, ephedra ingestion and myocardial infarction. The other authors of this report declared that they have no financial relationships with industry or others pertinent to this topic.

Task Force 9: Drugs and Performance-Enhancing Substances

N. A. Mark Estes III, MD, FACC, *Chair*

Robert Kloner, MD, PhD, FACC, Brian Olshansky, MD, FACC, Renu Virmani, MD, FACC

GENERAL CONSIDERATIONS

Athletes commonly use drugs and dietary supplements because they hope to improve athletic performance. These performance-enhancing substances include ergogenic and thermogenic supplements, stimulants, anabolic steroids, peptide hormones, and others. Despite aggressive marketing and user testimonials, scientific studies assessing the benefits and risks of any of these substances have not been conducted (1-5). Clinical observations indicate some supplements may have serious side effects including fatal adverse reactions (6-8). Athletes should make informed decisions regarding the use of drugs and dietary supplements with careful consideration of what is known and unknown. Health care professionals should ask about drug and dietary supplements and serve as an educational resource for athletes and athletic organizations (9).

Many drugs and supplements are marketed to improve exercise duration or physical strength, to shorten recovery time from exertion, to reduce fat, or in other ways to improve athletic performance (1-3,10-13). These substances include anabolic-androgenic steroids and the more than 30 natural and synthetic derivatives including tetrahydrogestrinone (THG). Androstenedione, commonly referred to as "andro," and dehydroepiandrosterone (DHEA) are available in oral form and are sold as nutritional supplements. Stimulants include amphetamines, cocaine, dexadrine, ephedra, ritalin, beta-2 agonists, and others (1-3,10-13). Peptide hormones and analogues, such as recombinant erythropoietin (EPO), are used as a pharmacologic alternative to "blood doping" or autotransfusion (1-3,10-13). Human growth hormone (hCG), chorionic gonadotropin, pituitary and synthetic gonadotropins (LH), and corticotropins (ACTH, tetracosactide) also are used because they are believed to improve athletic performance (1-3,10-13).

Dietary supplements are products, other than tobacco, containing vitamins, minerals, amino acids, herbs, or other botanical dietary substances (1-3,10-13). Some substances such as vitamins, minerals, bee pollen, caffeine, glycine, carnitine, lecithin, brewer's yeast, gelatin, creatine, protein supplements, and others probably have minimal toxicity when used in recommended doses. Based on serious concerns regarding the safety of a popular energy drink with taurine, caffeine, and glucuronolactone, multiple European countries have banned its sale (10). Like most dietary supplements this drink has not been assessed for efficacy or toxicity in rigorous scientific studies (1-3,10-13).

Other banned drugs that are not considered performance-enhancing would come under the designation of recreational

drugs (1-5,13). These include alcohol, cannabinoids, sedatives, narcotics, LSD, and others that have the potential to impair cognitive and physical function and are prohibited (1-3,12,13). The dietary supplement ephedra (ma huang) is associated with life-threatening toxicity and death resulting on a ban of its sale by the Food and Drug Administration (6-8). Inadequate labeling and insufficient quality control in the production of many nutritional supplements are also reasons to recommend that athletes not take dietary supplements. Contamination or poor labeling of nutritional supplements are not regarded as adequate defenses by athletic governing bodies. Recognizing that there may be exceptional circumstances where an athlete will require an otherwise prohibited substance for medical purposes, formal therapeutic exemption mechanisms are available with prior consideration and approval (11-13).

Athletic governing bodies should provide comprehensive lists of prohibited drugs and dietary supplements (1,11-13). They should develop a rigorous approach to prevent performance-enhancing and recreational drug and dietary supplement use. The crucial elements of any program should include education, counseling, treatment, detection, and enforcement. Governing athletic bodies should use all available resources to enhance, supplement, and coordinate existing efforts to educate athletes and reinforce the ethical principles inherent in athletic participation. Without such oversight, the integrity of athletics is threatened. Ultimately, athletes must accept responsibility for the decisions they make regarding the usage of drugs and performance-enhancing substances.

Athletes taking or considering the use of such substances should be aware that the safety and efficacy of supplements used for improving athletic performance have not been addressed in systematic scientific studies. Trainers, exercise physiologists, sports nutritionists, athletic governing bodies, or medical organizations should discourage the use of dietary supplements by athletes. Serious side effects may result from the use of these substances, including cardiac hypertrophy, myocyte necrosis, myocarditis, fibrosis, coronary thrombosis, and sudden death (even at recommended dosing) (2,3,6-8,10).

Recommendation:

- 1. Athletes should have their nutritional needs met through a healthy, balanced diet without dietary supplements.**

TASK FORCE 9 REFERENCES

1. Estes NAM III, Link MS, Cannom D, et al. Report of the NASPE policy conference on arrhythmias and the athlete. *J Cardiovasc Electrophysiol* 2001;12:1208–19.
2. Kloner RA. Illicit drug use in the athlete as a contributor to cardiac events. In: Estes NAM III, Salem D, Wang P, editors. *Sudden Cardiac Death in the Athlete*. Armonk, NY: Futura Pub. Co., 1998:441–52.
3. Cregler LL. Substance abuse in sports: the impact of cocaine, alcohol, steroids, and other drugs on the heart. In: Williams R, editor. *The Athlete and Heart Disease: Diagnosis, Evaluation and Management*. Philadelphia, PA: Lippincott Williams & Wilkins, 1998:131–54.
4. DeAngelis CD, Fontanarosa PB. Drugs alias dietary supplements. *JAMA* 2003;290:1519–20.
5. Stout CW, Weinstock J, Homoud MK, Wang PJ, Estes NAM III, Link MS. Herbal medicine: beneficial effects, side effects, and promising new research in the treatment of arrhythmias. *Curr Cardiol Rep* 2003;5:395–401.
6. Samenuk D, Link MS, Homoud MK, et al. Adverse cardiovascular events temporally associated with ma huang, an herbal source of ephedrine. *Mayo Clin Proc* 2002;77:12–6.
7. U.S. Food and Drug Administration. FDA announces plans to prohibit

sale of dietary supplements containing ephedra. December 30, 2003. Available at: <http://www.fda.gov/oc/initiatives/ephedra/february2004/>. Accessed October 1, 2004.

8. U.S. Food and Drug Administration. RAND report. Available at: <http://www.fda.gov/OHRMS/DOCKETS/98fr/95n-0304-bkg0003-ref-07-01-index.htm>. Accessed October 1, 2004.
9. Maron BJ, Thompson PD, Puffer JC, 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 the young), American Heart Association. *Circulation* 1996;94:850–6.
10. Ireland to review safety of energy drinks. Available at: <http://www.foodanddrinkurope.com/news/ng.asp?id=49957>. Accessed October 1, 2004.
11. The National Center for Drug Free Sport, Inc. Nutritional supplements. Available at: <http://www.drugfreesport.com/choices/supplements/index.html>. Accessed October 1, 2004.
12. NCAA Web site. Drug testing policy. Available at: http://www1.ncaa.org/membership/ed_outreach/health-safety/drug_testing/index.html. Accessed October 1, 2004.
13. World Anti-Doping Agency. Available at: <http://www.wada-ama.org>. Accessed November 30, 2004.

Appendix 1. Author Relationships With Industry and Others

Name	Consultant	Research Grant	Scientific Advisory Board	Speakers' Bureau
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Dr. Robert Kloner	<ul style="list-style-type: none"> • Bayer GSK • Lilly ICOS • Pfizer • Schering Plough 	<ul style="list-style-type: none"> • CV Therapeutics • Lilly ICOS • McNeill 	<ul style="list-style-type: none"> • Bayer GSK • Lilly ICOS • Pfizer 	<ul style="list-style-type: none"> • Bayer GSK • Lilly ICOS • Pfizer
Dr. Brian Olshansky	<ul style="list-style-type: none"> • Guidant • Medicorp 	<ul style="list-style-type: none"> • Medtronic 	None	<ul style="list-style-type: none"> • Astra Zeneca • Baxter • Bayer GSK • Reliant Pharmaceutical
Dr. Renu Virmani	None	None	None	None

Task Force 10: Automated External Defibrillators

Robert J. Myerburg, MD, FACC, *Chair*
 N. A. Mark Estes III, MD, FACC, John M. Fontaine, MD, FACC, Mark S. Link, MD, FACC,
 Douglas P. Zipes, MD, MACC

GENERAL CONSIDERATIONS OF CARDIAC ARREST RISK AMONG ATHLETES

The incidence of out-of-hospital cardiac arrest among the general population of adults is 1 to 2 deaths per 1,000 subjects per year; a figure that represents 50% of all cardiovascular deaths (1). For the adolescent and young adult subgroups, the estimated incidence is 1 per 100,000 per year or less. Available data suggest that among the younger population, competitive athletes account for a disproportionately higher-risk subset, compared to the gen-

eral population in a comparable age group (2–4). In addition, among the adult population performing conditioning activities in health clubs, the probability of cardiac arrest during exertion appears higher than the expected rate for comparable groups generally, especially among persons who exercise despite being habitually sedentary (5).

Within the subgroup of the population age 35 years and older, coronary heart disease accounts for approximately 80% of all sudden cardiac deaths (SCDs), with the cardiomyopathies accounting for another 10% to 15%. In the younger age groups, hypertrophic cardiomyopathy, anoma-

lous coronary arteries, myocarditis, and various other inherited disorders that are associated with arrhythmic risk dominate the etiologies (1). Although the absolute risk for the young athlete remains low, the excess compared to the general population in their age group and the life expectancy associated with the underlying diseases in the absence of a cardiac arrest, suggests the need for aggressive approaches to the recognition of individuals at risk, and for systems to respond to unexpected events.

ROLE OF AUTOMATED EXTERNAL DEFIBRILLATORS IN RESPONSE TO CARDIAC ARREST

In attacking the problem of SCD, it is unrealistic to assume that any single approach—epidemiological surveillance, primary prevention of disease states, clinical interventions of established diseases, or community-based response systems—will have a major impact alone. Each strategy has a role, in part because a large majority of events occur unexpectedly in the out-of-hospital environment and are not predictable with great accuracy by risk profiling in most clinical circumstances (6).

Because the majority of out-of-hospital cardiac arrests occur by the initial mechanism of ventricular fibrillation, community-based defibrillation strategies have emerged as one approach to the SCD problem. Time to defibrillation is the most important factor in survival from out-of-hospital cardiac arrest due to ventricular fibrillation (7). Thus, community deployment of rapid access systems has been evolving since the late 1960s, when portable defibrillators initially became available. The first systems were fire department-based paramedical programs, and were followed years later by the placement of automated external defibrillators (AEDs) in the hands of non-conventional trained responders, such as security guards, police, and flight attendants (8–15). Most recently, defibrillators were made available to minimally trained or untrained lay responders in public locations such as airports, commercial aircraft, supermarkets, stadiums, and hospitals (14–17), and have also been suggested for private residences or neighborhoods (18).

Although survival rates from out-of-hospital cardiac arrest remain far lower than desirable, the various out-of-hospital response strategies have improved the survival rates, largely based on more rapid response times. Those settings and strategies that provide response times from witnessed onset to initial defibrillation of less than 2 to 3 min can provide survival outcomes hovering about 50% (16); but rates fall sharply with each passing minute thereafter. By 4 to 5 min, survival is 25% or less, and less than 10% after 10 min (19). As a consequence, despite the apparent value of conventional emergency medical systems and other vehicle-based systems such as police responders, additional public access systems are anticipated to provide even faster access to defibrillation. Such systems are being deployed and tested (20,21). Each has potential or demonstrates added benefit to survival rates.

AEDs AT SITES OF TRAINING AND COMPETITION

Among young athletes, cardiac arrests generally occur during or shortly after intense training sessions or competition. Although the incidence of cardiac arrest is extremely low (approximately 1% of that reported in middle-age and older adult populations), the value of prompt and successful resuscitation and long-term survival is enhanced by the potential of extended life over many decades (i.e., much longer than is the case for older adults, among whom the extent of underlying disease results in substantial risk over shorter time periods). Thus, it is considered reasonable to have an AED available for use at educational facilities, training centers, and sports arenas and stadiums, in addition to trained responders among the staff at each (5,21). The AEDs should be deployed in a distribution that can achieve an anticipated response time of 5 min or less (22). When the time from loss of consciousness to availability of an AED is 5 min or longer, 30 to 60 s of CPR prior to the first attempt to defibrillate has been shown to provide a survival benefit (23).

Although there are only anecdotal observations supporting the feasibility and impact of this strategy, the rationale is clear and should be promoted. In addition, it should be recognized that the availability of AEDs during competitive sporting events also provides the potential for life-saving support to spectators and other bystanders. Nonetheless, the availability of an AED at a sporting event should not be construed as absolute protection against a fatal outcome from a cardiac arrest. Neither should it supersede restrictions against participation in competitive sports, based upon underlying cardiac abnormalities, as defined in this document.

Recommendations:

1. **The AEDs should be available at educational facilities that have competitive athletic programs (including intramural sports and conditioning classes), stadiums, arenas, and training sites, with trained responders identified among the permanent staff. Devices should be deployed so as to provide a response time of less than 5 min.**
2. **The initial response to a suspected or identified cardiac arrest should be to contact emergency medical services (e.g., 9-1-1), followed immediately by, or concomitant with, initiating CPR and deploying the AED.**

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TASK FORCE 10 REFERENCES

1. Myerburg RJ, Castellanos A. Cardiac arrest and sudden cardiac death. In: Zipes DP, Libby P, Bonow RO, Braunwald E, editors. Braunwald's Heart Disease: A Textbook of Cardiovascular Medicine. 7th edition. Philadelphia, PA: WB Saunders Company, 2005:865–908.
2. Thiene G, Basso C, Corrado D. Is prevention of sudden death in young athletes feasible? *Cardiologia* 1999;44:497–505.
3. Corrado D, Basso C, Schiavon M, Thiene G. Screening for hypertrophic cardiomyopathy in young athletes. *N Engl J Med* 1998;339:364–9.
4. Maron BJ. Sudden death in young athletes. *N Engl J Med* 2003;349:1064–75.
5. Albert CM, Mittleman MA, Chae CU, Lee IM, Hennekens CH, Manson JE. Triggering of sudden death from cardiac causes by vigorous exertion. *N Engl J Med* 2000;343:1355–61.

6. Myerburg RJ. Sudden cardiac death: exploring the limits of our knowledge. *J Cardiovasc Electrophysiol* 2001;12:369-81.
7. Cobb LA, Weaver WD, Fahrenbruch CE, Hallstrom AP, Copass MK. Community-based interventions for sudden cardiac death: impact, limitations, and changes. *Circulation* 1992;85:198-102.
8. Mosesso VN Jr., Davis EA, Auble TE, Paris PM, Yealy DM. Use of automated external defibrillators by police officers for treatment of out-of-hospital cardiac arrest. *Ann Emerg Med* 1998;32:200-7.
9. White RD, Hankins DG, Bugliosi TF. Seven years' experience with early defibrillation by police and paramedics in an emergency medical services system. *Resuscitation* 1998;39:145-51.
10. Kette F, Sbrojavacca R, Rellini G, et al. Epidemiology and survival rate of out-of-hospital cardiac arrest in north-east Italy: the F.A.C.S. study: Friuli Venezia Giulia Cardiac Arrest Cooperative Study. *Resuscitation* 1998;36:153-9.
11. Newman MM, Mosesso VN Jr., Ornato JP, et al. Law Enforcement Agency Defibrillation (LEA-D): position statement and best practices recommendations from the National Center for Early Defibrillation. *Prehosp Emerg Care* 2002;6:346-7.
12. Myerburg RJ, Fenster J, Velez M, et al. Impact of community-wide police car deployment of automated external defibrillators on survival from out-of-hospital cardiac arrest. *Circulation* 2002;106:1058-64.
13. Capucci A, Aschieri D, Piepoli MF, Bardy GH, Iconomu E, Arvedi M. Tripling survival from sudden cardiac arrest via early defibrillation without traditional education in cardiopulmonary resuscitation. *Circulation* 2002;106:1065-70.
14. O'Rourke MF, Donaldson E, Geddes JS. An airline cardiac arrest program. *Circulation* 1997;96:2849-53.
15. Page RL, Joglar JA, Kowal RC, et al. Use of automated external defibrillators by a U.S. airline. *N Engl J Med* 2000;343:1210-6.
16. Valenzuela TD, Roe DJ, Nichol G, Clark LL, Spaite DW, Hardman RG. Outcomes of rapid defibrillation by security officers after cardiac arrest in casinos. *N Engl J Med* 2000;343:1206-9.
17. Caffrey SL, Willoughby PJ, Pepe PE, Becker LB. Public use of automated external defibrillators. *N Engl J Med* 2002;347:1242-7.
18. Zipes DP. President's page: the neighborhood health watch program: Save A Victim Everywhere (SAVE). *J Am Coll Cardiol* 2001;37:2004-5.
19. Holmberg M, Holmberg S, Herlitz J. Incidence, duration and survival of ventricular fibrillation in out-of-hospital cardiac arrest patients in Sweden. *Resuscitation* 2000;44:7-17.
20. Nichol G, Hallstrom AP, Kerber R, et al. American Heart Association report on the Second Public Access Defibrillation Conference, April 17-19, 1997. *Circulation* 1998;97:1309-14.
21. Marengo JP, Wang PJ, Link MS, Homoud MK, Estes NA III. Improving survival from sudden cardiac arrest: the role of the automated external defibrillator. *JAMA* 2001;285:1193-200.
22. Hazinski MF, Markenson D, Neish S, et al. Response to cardiac arrest and selected life-threatening medical emergencies: the medical emergency response plan for schools: a statement for healthcare providers, policymakers, school administrators, and community leaders. *Circulation* 2004;109:278-91.
23. Wik L, Hansen TB, Fylling F, et al. Delaying defibrillation to give basic cardiopulmonary resuscitation to patients with out-of-hospital ventricular fibrillation: a randomized trial. *JAMA* 2003;289:1389-95.

Appendix 1. Author Relationships With Industry and Others

Name	Consultant	Research Grant	Scientific Advisory Board	Stock Holder	Expert Witness Testimony
Dr. N. A. Mark Estes III	<ul style="list-style-type: none"> • Guidant • Medtronic 	<ul style="list-style-type: none"> • Guidant • Medtronic 	<ul style="list-style-type: none"> • Guidant (Executive Committee) 	None	None
Dr. John M. Fontaine	None	None	None	None	None
Dr. Mark S. Link	None	<ul style="list-style-type: none"> • Guidant • Medtronic 	None	None	None
Dr. Robert J. Myerburg	<ul style="list-style-type: none"> • Guidant • Procter & Gamble 	None	<ul style="list-style-type: none"> • Procter & Gamble • Reliant Pharmaceutical 	None	<ul style="list-style-type: none"> • 2000, Defense, Lewis vs. Mudge • 2002, Defense, Weiner vs. Vitello • 2005, Defense, Ephedra Multi-District Litigation
Dr. Douglas P. Zipes	<ul style="list-style-type: none"> • Cardiofocus • Janssen • Medtronic 	<ul style="list-style-type: none"> • Medtronic 	<ul style="list-style-type: none"> • Medtronic 	<ul style="list-style-type: none"> • MVMD 	<ul style="list-style-type: none"> • 1996, Defense, Knapp vs. Northwestern

Task Force 11: Commotio Cordis

Barry J. Maron, MD, FACC, *Chair*
 N. A. Mark Estes III, MD, FACC, Mark S. Link, MD, FACC

GENERAL CONSIDERATIONS

Sudden and unexpected deaths of young athletes are most frequently the consequence of unsuspected cardiovascular diseases (1). However, participants in organized sports are also subject to another risk for sudden death that occurs in the

absence of cardiovascular disease—namely, blunt, non-penetrating, and usually innocent-appearing chest blows, triggering ventricular fibrillation unassociated with structural damage to the ribs, sternum, or heart itself (*commotio cordis*) (2,3). Although the precise incidence during competitive

and recreational sports is unknown, commotio cordis events may be a more frequent cause of sudden death than previously believed, and also more common than many of the cardiovascular diseases that cause these catastrophes (1).

Knowledge of the demographics and clinical profile of commotio cordis is based largely on information from the U.S. Registry (Minneapolis, Minnesota) (2,3). Precordial blows that cause commotio cordis usually are not perceived to be unusual for the sport or activity involved, nor of sufficient magnitude to trigger arrhythmic sudden death. Although reported at a wide range of ages (3 months to 50 years), commotio cordis has a predilection for children and adolescents (mean age 13 years) probably because the young characteristically have narrow, pliable chest walls that facilitate transmission of energy from the chest impact to the myocardium.

Commotio cordis occurs in a wide variety of sports, but most commonly youth baseball (and softball), ice hockey, football, and lacrosse, with death often caused by projectiles that are implements of the competition. Although blows are typically of low energy, projectiles may strike the precordium with a range of velocities—paradoxically, and most commonly, with only modest force such as a pitched baseball striking a batter at 30 to 40 mph, but also with high-velocity blows from hockey pucks or lacrosse balls (up to an estimated 90 mph), and frequently with bodily contact to the precordium such as by karate blows or collisions between outfielders tracking a baseball in-flight. Collapse can be instantaneous or preceded by brief periods of consciousness and physical activity. Despite a structurally normal heart, survival from commotio cordis is uncommon (i.e., only 15%). However, survival from commotio cordis has been reported with increasing frequency associated with prompt cardiopulmonary resuscitation and defibrillation (3). Survivors of commotio cordis should undergo a complete cardiac evaluation including electrocardiogram (ECG), Holter ambulatory monitoring, echocardiogram, and possibly cardiac catheterization to exclude underlying structural cardiac abnormalities.

Also, many deaths from commotio cordis occur around the home or on the playground in informal activities related or unrelated to recreational sports (often involving close relatives) in which the chest impact is delivered in an innocent fashion; for example, such events have occurred as a result of light blows during playful “shadow boxing” or as a remedy for hiccups (3). Unfortunately, some commotio cordis events have even triggered criminal convictions for manslaughter or murder (4).

A swine model that replicates commotio cordis has provided important insights into the mechanisms responsible for the devastating electrophysiologic consequences of these precordial blows (5–7). Determinants of ventricular fibrillation following a chest blow include impact delivered at a wide range of velocities directly over the heart, and timing within a narrow 15-to-30-ms window just prior to the T-wave peak during the vulnerable phase of repolariza-

tion (representing only 1% of the cardiac cycle) (5–8). The requirement for such an exquisite confluence of circumstances may largely explain the uncommon occurrence of commotio cordis.

In addition, spontaneously aborted commotio cordis events may occasionally result from blows sustained during the QRS complex (depolarization), which trigger transient complete heart block or nonsustained polymorphic ventricular tachycardia in the animal model (5). Basic cellular mechanisms responsible for commotio cordis are incompletely understood, although selective activation of K^+ ATP channels may play a pivotal role (9).

Several strategies for prevention of commotio cordis events, including innovations in the design of sports equipment, have been considered. Softer-than-normal (“safety”) baseballs reduce risk for ventricular fibrillation under laboratory conditions (5,7). Although such projectiles do not provide absolute protection from sudden death on the baseball field (3), nevertheless there is sufficient reason to encourage the use of such baseballs in organized play at appropriate ages (10). Chest barriers with proven efficacy for youth sports (e.g., baseball, lacrosse, and hockey) have not yet been developed, and many of the commercially available products offer no or only incomplete protection against provoked arrhythmias (3,11). The continued reports of commotio cordis events during organized and recreational sports emphasize the importance of more timely resuscitative efforts, including immediate access to automated external defibrillators (AEDs) (12,13), and also development of preventive strategies including design of effective chest barriers (11).

Recommendations:

1. **Age-appropriate safety baseballs are recommended for use in children up to 13 years of age.**
2. **Although chest wall protectors may prevent traumatic injury in goalies and baseball catchers, insufficient evidence is available to recommend universal use of commercially available chest barriers for all participants in sports, specifically to prevent commotio cordis events.**
3. **AEDs should be available within 5 min after participant collapse at sporting events.**
4. **Survivors of a commotio cordis with ventricular fibrillation (or a presumed aborted event without documented ventricular fibrillation) should undergo a thorough cardiac evaluation, including at least 12-lead ECG, ambulatory Holter monitoring, and echocardiogram. Standard electrophysiologic testing and an implantable cardioverter-defibrillator are not usually recommended.**
5. **Because data are lacking with regard to the susceptibility for recurrent events, eligibility for returning to competitive sports in survivors is at present a decision left to individual clinical judgment.**

TASK FORCE 11 REFERENCES

1. Maron BJ. Sudden death in young athletes. *N Engl J Med* 2003;349:1064-75.
2. Maron BJ, Poliac LC, Kaplan JA, Mueller FO. Blunt impact to the chest leading to sudden death from cardiac arrest during sports activities. *N Engl J Med* 1995;333:337-42.
3. Maron BJ, Gohman TE, Kyle SB, Estes NAM III, Link MS. Clinical profile and spectrum of commotio cordis. *JAMA* 2002;287:1142-6.
4. Maron BJ, Mitten MJ, Greene BC. Criminal consequences of commotio cordis. *Am J Cardiol* 2002;89:210-3.
5. Link MS, Wang PJ, Pandian NG, et al. An experimental model of sudden death due to low-energy chest-wall impact (commotio cordis). *N Engl J Med* 1998;338:1805-11.
6. Link MS, Maron BJ, VanderBrink BA, et al. Impact directly over the cardiac silhouette is necessary to produce ventricular fibrillation in an experimental model of commotio cordis. *J Am Coll Cardiol* 2001;37:649-54.
7. Link MS, Maron BJ, Wang PJ, Pandian NG, VanderBrink BA, Estes NAM III. Reduced risk of sudden death from chest wall blows (commotio cordis) with safety baseballs. *Pediatrics* 2002;109:873-7.
8. Link MS, Maron BJ, Wang PJ. Upper and lower limits of vulnerability to sudden arrhythmic death with chest wall impact (commotio cordis). *J Am Coll Cardiol* 2003;41:99-104.
9. Link MS, Wang PJ, VanderBrink BA, et al. Selective activation of the K(+)(ATP) channel is a mechanism by which sudden death is produced by low-energy chest-wall impact (commotio cordis). *Circulation* 1999;100:413-8.
10. Kyle SB. Youth Baseball Protective Equipment Project Final Report. United States Consumer Product Safety Commission. Washington, DC: 1996.
11. Weinstock J, Maron BJ, Song C, Mane PP, Estes NAM III, Link MS. Commercially available chest wall protectors fail to prevent ventricular fibrillation induced by chest wall impact (commotio cordis) (abstr). *Heart Rhythm* 2004;1:692.
12. Strasburger JF, Maron BJ. Images in clinical medicine: commotio cordis. *N Engl J Med* 2002;347:1248.
13. Link MS, Maron BJ, Stickney RE, et al. Automated external defibrillator arrhythmia detection in a model of cardiac arrest due to commotio cordis. *J Cardiovasc Electrophysiol* 2003;14:83-7.

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Name	Consultant	Research Grant	Scientific Advisory Board	Expert Witness Testimony
Dr. N. A. Mark Estes III	<ul style="list-style-type: none"> • Guidant • Medtronic 	<ul style="list-style-type: none"> • Guidant • Medtronic 	<ul style="list-style-type: none"> • Guidant (Executive Committee) 	None
Dr. Mark S. Link	None	<ul style="list-style-type: none"> • Guidant • Medtronic 	None	None
Dr. Barry J. Maron	None	<ul style="list-style-type: none"> • Medtronic 	None	<ul style="list-style-type: none"> • 1996, Defense, Knapp vs. Northwestern

Task Force 12: Legal Aspects of the 36th Bethesda Conference Recommendations

Matthew J. Mitten, JD, *Chair*
 Barry J. Maron, MD, FACC, Douglas P. Zipes, MD, MACC

GENERAL CONSIDERATIONS

In 1994, when the 26th Bethesda Conference recommendations were formulated, no court had yet considered whether an athlete with a cardiovascular abnormality could be involuntarily excluded from a competitive sport if physicians disagreed in their participation recommendations (1-3). However, new data have subsequently become available, and several highly visible cases involving the sudden deaths of elite competitive athletes (4,5) have brought medical-legal and liability considerations into prominent focus. A 1996 lawsuit brought by a student-athlete claiming the legal right to play intercollegiate basketball contrary to a university team physician's medical recommendation has established a developing legal framework for medical decisions regarding the eligibility or disqualification of trained athletes with cardiovascular disease to participate in competitive sports. In this case, Knapp vs. Northwestern University (6,7), a federal appellate court recognized the appropriateness of a physician's reliance on current consensus medical guidelines when making a participation recommendation for an athlete with a cardiovas-

cular abnormality. Consequently, judicial precedent now provides some guidance regarding the role of the present 36th Bethesda Conference recommendations in resolving legal issues relating to athletic participation disputes.

In the Knapp case, the court upheld Northwestern University's legal right to accept its team physician's recommendation, which was consistent with the then-current 26th Bethesda Conference guidelines, to medically disqualify a student-athlete from playing college basketball (6). As a high school senior, Nicholas Knapp suffered sudden cardiac arrest while playing in an informal basketball game, which required cardiopulmonary resuscitation and defibrillation to restore sinus rhythm. Thereafter, he had a cardioverter-defibrillator (ICD) implanted. He resumed playing recreational basketball without any subsequent cardiovascular events, and three cardiologists medically cleared him to play college basketball.

Knapp had received a full athletic scholarship at Northwestern University to play intercollegiate basketball. While Northwestern honored Knapp's scholarship, it barred him

from playing on its basketball team based on the team physician's medical recommendation. The team physician considered Knapp's medical records and history, the 26th Bethesda Conference recommendations (1), and the opinions of two consulting cardiologists who concluded that Knapp would expose himself to a medically unacceptable risk for ventricular fibrillation during competitive athletics.

All medical experts agreed on the following facts: 1) Knapp had suffered a cardiac arrest; 2) even with the ICD, playing college basketball placed Knapp at a higher risk of sudden cardiac death as compared to other male college basketball players; 3) the ICD had never been tested under the conditions of intercollegiate basketball; and 4) no person currently played or had ever played college or professional basketball after having an ICD implanted. However, the experts disagreed whether Knapp should be medically disqualified from playing intercollegiate basketball.

The trial court ruled that Northwestern should restore Knapp's eligibility to play on its basketball team and mandated a courtside defibrillator and cardiologist to be present at all games and practices. However, the appeals court overruled that decision and held that a university has a legal right to establish legitimate physical qualifications for its intercollegiate athletes. Northwestern did not violate the Rehabilitation Act of 1973, a federal law prohibiting discrimination against persons with disabilities, by following its team physician's reasonable medical advice. An athlete may be medically disqualified and excluded from a sport if necessary to avoid an enhanced risk of death or serious injury during competitive athletics that cannot be eliminated through the use of medication, monitoring, or protective equipment. The court explained that Northwestern's decision to exclude Knapp from its basketball team was legally justified:

"We do not believe that, in cases where medical experts disagree in their assessment of the extent of a real risk of serious harm or death, Congress intended that the courts—neutral arbiters but generally less skilled in medicine than the experts involved—should make the final medical decision. Instead, in the midst of conflicting expert testimony regarding the degree of serious risk of harm or death, the court's place is to ensure that the exclusion or disqualification of an individual was individualized, reasonably made, and based upon competent medical evidence. . . . [W]e wish to make clear that we are not saying Northwestern's decision is necessarily the right decision. We say only that it is not an illegal one under the Rehabilitation Act" (6).

Knapp eventually left Northwestern and pursued collegiate basketball at another university where the team physician cleared him to play. Shortly thereafter, his ICD delivered an apparently appropriate shock during a basketball game.

The present 36th Bethesda Conference recommendations update the 26th Bethesda Conference guidelines of 1994 (1) (which modified the 16th Bethesda Conference guidelines of 1984), taking into account the most recent and relevant developments in the diagnosis and management of

cardiovascular disease. These new guidelines represent the most current consensus opinion of a distinguished group of cardiologists regarding the medical risks of participation in competitive sports by athletes with cardiovascular abnormalities. Thus, we anticipate that the 36th Bethesda Conference recommendations will be recognized and accepted by physicians but also by the legal community and courts, as the most contemporary consensus opinion of a distinguished expert panel of cardiologists regarding medical eligibility and disqualification recommendations for competitive athletes with cardiovascular disease.

The Knapp case recognized the appropriateness of physician reliance on current consensus guidelines in making medical clearance recommendations. However, the court did not rule that the 26th Bethesda Conference guidelines would always be legally determinative in resolving athletic participation disputes involving athletes with cardiovascular abnormalities. Therefore, consistent with legal precedent established by *Knapp vs. Northwestern University*, a physician may justifiably consider and rely upon the updated 36th Bethesda Conference recommendations in making medical eligibility recommendations for competitive athletes with cardiovascular disease. Nevertheless, the law continues to require that these recommendations be applied on an individualized basis rather than used to exclude categorically all competitive athletes who have a particular cardiovascular abnormality.

The Knapp case establishes an important precedent regarding the medical exclusion of college and high school athletes with cardiovascular disease from intercollegiate and interscholastic athletics, for whom sports is an avocation or extracurricular activity incidental to one's education (6,7). It is presently uncertain whether this same legal framework will be applied to resolve future participation disputes that involve professional athletes (for whom sports is an income-generating livelihood) (8). However, it is notable that the U.S. Supreme Court recently held that an enhanced risk of significant harm to personal health is a legitimate ground for exclusion from employment (9), which suggests that the legal framework developed in the Knapp case may be applied to professional sports.

Currently there is no well-defined legal precedent regarding a physician's potential malpractice liability for medically clearing an athlete with a cardiovascular abnormality to participate in a competitive sport contrary to consensus recommendations (10). The law generally requires a physician to have and use the current knowledge, skill, and care ordinarily possessed and employed by members of the medical profession in good standing. The applicable legal standard of physician conduct is "good medical practice" within the physician's area of specialty practice, which depending on the jurisdiction means either "reasonable," "customary," or "accepted" medical care under the circumstances (10). This general standard applies to physicians who provide cardiovascular medical treatment to a competitive athlete, including evaluation of his or her medical fitness to participate in a sport.

Courts generally have recognized “guidelines” established by national medical associations as evidence of good medical practice, but they are not conclusive evidence of the standard of care (11–13). Indeed, consistent with the requirements of the federal disability discrimination laws as interpreted in the Knapp case, it is important to emphasize that the Bethesda Conference recommendations permit the exercise of a physician’s medical judgment in individual cases. The recommendations do not, per se, rigidly restrict clinical practice or medical decision making. A clinician has the flexibility to deviate from the recommendations if he or she believes it is in the best interests of a patient-athlete to reach an alternative decision and strategy.

The controlling legal issue is whether adherence to (or deviation from) consensus recommendations is consistent with reasonable, customary, or accepted medical practice in an individual patient’s case. Although the recommendations of the 36th Bethesda Conference do not represent formal guidelines endorsed by the American College of Cardiology, 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. Therefore, deviations from the 36th Bethesda Conference recommendations that are nevertheless consistent with good medical practice and are protective of an athlete’s health may be appropriate in particular cases and do not necessarily create physician liability for medical malpractice. Conversely, compliance with the 36th Bethesda Conference recommendations is some evidence that a physician has satisfied this legal

requirement, and in future legal disputes may form the basis of a successful defense against allegations of malpractice (14).

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TASK FORCE 12 REFERENCES

1. Maron BJ, Mitchell JH. 26th Bethesda Conference: recommendations for determining eligibility for competition in athletes with cardiovascular abnormalities. *J Am Coll Cardiol* 1994;24:845–99.
2. Maron BJ, Brown RW, McGrew CA, Mitten MJ, Caplan AL, Hutter AM Jr. Ethical, legal, and practical considerations affecting medical decision-making in competitive athletes. *J Am Coll Cardiol* 1994;24: 854–60.
3. Mitten MJ, Maron BJ. Legal considerations that affect medical eligibility for competitive athletes with cardiovascular abnormalities and acceptance of Bethesda Conference recommendations. *J Am Coll Cardiol* 1994;24:861–3.
4. Maron BJ. Sudden death in young athletes: lessons from the Hank Gathers affair. *N Engl J Med* 1993;329:55–7.
5. Maron BJ. Sudden death in young athletes. *N Engl J Med* 2003;349: 1064–75.
6. 101 F. 3d 473 (7th Cir. 1996), cert. denied, 520 U.S. 1274 (1997).
7. Maron BJ, Mitten MJ, Quandt EF, Zipes DP. Competitive athletes with cardiovascular disease—the case of Nicholas Knapp. *N Engl J Med* 1998;339:1632–5.
8. Mitten MJ. Enhanced risk of harm to one’s self as a justification for exclusion from athletics. *Marq Sports L J* 1998;8:189–223.
9. *Chevron U.S.A. Inc. vs. Echazabal*, 536 U.S. 73 (2002).
10. Mitten MJ. Emerging legal issues in sports medicine: a synthesis, summary, and analysis. *St John’s L Rev* 2002;76:5–86.
11. *Stone vs. Proctor*, 131 S.E.2d 297, 299 (N.C. 1963).
12. *Pollard vs. Goldsmith*, 572 P.2d 1201, 1203 (Ariz. Ct. App. 1977).
13. *Swank vs. Halivopoulos*, 260 A.2d 240, 242–43 (N.J. Super. Ct. App. Div. 1969).
14. Mitten M. Team physicians and competitive athletes: allocating legal responsibility for athletic injuries. *Univ Pitt L Rev* 1993;55:129–60.

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Mr. Matthew J. Mitten	None	None	None	None	None
Dr. Douglas P. Zipes	• Cardiofocus • Janssen • Medtronic	• Medtronic	• Medtronic	• MVMD	• 1996, Defense, Knapp vs. Northwestern