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## Recommendations for Physical Activity and Recreational Sports Participation for Young Patients With Genetic Cardiovascular Diseases

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**Abstract**—A group of relatively uncommon but important genetic cardiovascular diseases (GCVDs) are associated with increased risk for sudden cardiac death during exercise, including hypertrophic cardiomyopathy, long-QT syndrome, Marfan syndrome, and arrhythmogenic right ventricular cardiomyopathy. These conditions, characterized by diverse phenotypic expression and genetic substrates, account for a substantial proportion of unexpected and usually arrhythmia-based fatal events during adolescence and young adulthood. Guidelines are in place governing eligibility and disqualification criteria for competitive athletes with these GCVDs (eg, Bethesda Conference No. 26 and its update as Bethesda Conference No. 36 in 2005). However, similar systematic recommendations for the much larger population of patients with GCVD who are not trained athletes, but nevertheless wish to participate in any of a variety of recreational physical activities and sports, have not been available. The practicing clinician is frequently confronted with the dilemma of designing noncompetitive exercise programs for athletes with GCVD after disqualification from competition, as well as for those patients with such conditions who do not aspire to organized sports. Indeed, many asymptomatic (or mildly symptomatic) patients with GCVD desire a physically active lifestyle with participation in recreational and leisure-time activities to take advantage of the many documented benefits of exercise. However, to date, no reference document has been available for ascertaining which types of physical activity could be regarded as either prudent or inadvisable in these subgroups of patients. Therefore, given this clear and present need, this American Heart Association consensus document was constituted, based largely on the experience and insights of the expert panel, to offer recommendations governing recreational exercise for patients with known GCVDs. (*Circulation*. 2004;109:2807-2816.)

**Key Words:** AHA Scientific Statements ■ exercise ■ cardiomyopathy ■ genetics ■ arrhythmia

Genetic cardiovascular diseases (GCVDs) include hypertrophic cardiomyopathy (HCM), arrhythmogenic right ventricular cardiomyopathy (ARVC), Marfan syndrome, and the ion-channel diseases, including long-QT syndrome (LQTS), Brugada syndrome, and catecholaminergic polymorphic ventricular tachycardia (CPVT).<sup>1-16</sup> Although relatively uncommon in the general population and most cardiology practices, these conditions have nevertheless been associated with an increased risk for sudden death during

exercise and account for a substantial proportion of the unexpected and usually arrhythmogenic fatal events during adolescence and young adulthood.<sup>1-6,8-27</sup> These deaths are devastating to the families, community, and physicians, particularly in light of the youthful age of the victims.

Previous expert recommendations and the attention of the medical community generally focused on trained athletes with heart disease engaged in organized and competitive sports programs for whom sudden death often occurs during

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or shortly after vigorous exertion on the athletic field.<sup>25–29</sup> Indeed, the Bethesda Conference No. 26 guidelines (to be updated in 2005) offer widely accepted eligibility and disqualification recommendations for competitive athletes with established cardiovascular abnormalities.<sup>29</sup> These recommendations are predicated on the view that intense physical exertion may create greater susceptibility to sudden death in those athletes with heart disease and unstable electrophysiological substrates.<sup>19,29,30</sup> Conversely, restriction from the unique lifestyle of competitive sports is likely to diminish that risk for an arrhythmia-based catastrophe. Nevertheless, in GCVD, most sudden death events, whether or not associated with some form of physical exertion, occur among the numerically larger subgroup of young persons who are not, in fact, trained competitive athletes.<sup>23,25</sup>

The practicing clinician is frequently confronted with the dilemma of designing noncompetitive exercise programs for those athletes with GCVDs after disqualification from competition, as well as for those patients with such conditions who do not aspire to organized sports. Indeed, many asymptomatic (or mildly symptomatic) patients with GCVDs desire a physically active lifestyle and participate in recreational and leisure-time activities to take advantage of the many established benefits of exercise.<sup>31,32</sup> Recently, published guidelines have focused narrowly on individuals participating in recreational sports settings such as health and fitness facilities<sup>31</sup> or in Master's sports competition.<sup>32</sup>

The conflict between the known benefits and potentially adverse consequences of exercise and the desire of young individuals to participate in various levels of physical activity creates a demand for appropriate information to resolve such uncertainty. At present, no systematic practice recommendations are available concerning the risk of recreational (non-competitive) exercise in adolescents and young adults with GCVDs. Therefore, there was a clear and present need to develop the present consensus recommendations governing recreational exercise for patients with known GCVDs. It is the aspiration of the panel that this document will be useful for cardiology subspecialists and practicing clinicians in pediatrics, internal medicine, primary care, or sports medicine.

## Definitions

For the purpose of this discussion, recreational sports activities are defined in juxtaposition to competitive sports. A competitive athlete is one who participates in an organized team or individual sport that requires systematic training and regular competition against others and that places a high premium on athletic excellence and achievement.<sup>33</sup> Characteristic of competitive athletes is the strong inclination to extend themselves to extremely high levels of exertion, often exceeding their native physical limits and sometimes for prolonged periods of time, regardless of other considerations.

Conversely, individuals participating in a variety of informal recreational sports and circumstances engage in a range of exercise levels from modest to vigorous on either a regular or an inconsistent basis, which do not require systematic training or the pursuit of excellence and are without the same pressure to excel against others that characterizes competitive

sports. The lack of systematic athletic conditioning in the definition of recreational sports is expected to decrease the risk of cardiovascular events.

Sudden cardiac death is usually the result of an interaction between acute triggers and the underlying heart disease (ie, substrate). Triggers for life-threatening ventricular tachyarrhythmias and sudden death during sports include emotional stress, environmental factors, myocardial ischemia, sympathetic-vagal imbalance, and hemodynamic changes. Intensive and systematic athletic training itself may increase the risk of sudden death in the presence of heart disease by promoting disease progression or worsening of the arrhythmogenic substrate (either structurally or electrically) over time. For example, in patients with HCM, recurrent episodes of exercise-induced myocardial ischemia during intensive training may result in cell death and myocardial replacement fibrosis, which in turn enhances ventricular electrical instability.<sup>34</sup> In patients with ARVC, regular and intense physical activity may provoke right ventricular volume overload and cavity enlargement, which in turn may accelerate fibrofatty atrophy.<sup>35</sup> In Marfan syndrome, the hemodynamic stress placed on the aorta by increased blood pressure and stroke volume during intense activity (particularly with rapid acceleration and deceleration) may promote and increase the rate of aortic enlargement.<sup>36</sup>

However, in the majority of patients with Brugada syndrome, the malignant ventricular arrhythmias occur at rest and, in many cases, at night as a consequence of an increased vagal activity and/or withdrawal of sympathetic activity.<sup>7,37,38</sup> Enhanced adrenergic drive such as occurs during sports activity could have an inhibitory effect and theoretically reduce sudden death risk.

The panel also recognizes that in formulating this definition, some individuals participating in recreational sports nevertheless train systematically (similar to, and as a surrogate for, competitive athletics). Indeed, it is far easier to formulate recommendations for competitive sports, which are easily defined forms of exercise, than for recreational sports, which may include a multitude of physical activities that are part of ordinary daily life. We have not included recommendations for physical activity associated with performing art forms such as dance and ballet.

## Scope of the Problem

A number of largely congenital and/or inherited cardiovascular diseases have been linked causally to unexpected sudden cardiac deaths in young people, including those engaged in either leisure sporting activities or organized and truly competitive athletics.<sup>6,7,18–25</sup> Although regional differences have been reported, in US-based surveys the most common causes of these deaths have been HCM and congenital coronary artery anomalies with coronary artery origin from the wrong sinus of Valsalva.<sup>17,19,20,22,23,25,26</sup> In Italy, ARVC and premature atherosclerotic coronary artery disease predominate.<sup>24,30,39,40</sup>

On the basis of data from the United States, GCVDs account for at least 40% of sudden deaths in young athletes.<sup>17,19,25</sup> Many patients with these conditions aspire to participate in a variety of recreational sporting activities, and

some may have even been previously withdrawn from competitive sports in accordance with the recommendations of Bethesda Conference No. 26.<sup>29</sup> Furthermore, the number of young patients and family members identified with these cardiac diseases has greatly increased because of advances in diagnostic molecular genetics and enhanced survival attributable to contemporary management strategies. Therefore, there is now a substantial population of youthful active patients with GCVDs who require prudent recommendations regarding daily exercise programs.

The health benefits of exercise at all ages have been emphasized repeatedly and promoted as a national public health agenda.<sup>41,42</sup> Certainly, there is substantial evidence that considerable medical advantage is derived from even regular moderate exercise and fitness, such as improvement in aerobic power and maximum oxygen uptake, blood lipid levels and glucose tolerance, as well as enhanced self-assurance, a sense of psychological and physical well-being, and improved overall quality of life.<sup>31,32,41,42</sup> Undoubtedly, similar benefits from regular exercise probably also accrue in a young patient population with GCVDs.

In addition, recent recognition in the United States that obesity is an emerging major health problem in young people has focused attention on the importance of regular exercise as a weight loss and maintenance strategy in adults.<sup>43</sup> In general terms, we believe these principles are also relevant to young patients with GCVD. Certainly, this panel acknowledges that involvement in sports is of particular importance to the physical and psychological well-being of children and adolescents, and abrupt removal from such activities can be devastating.

### **Objectives of the Panel**

The present statement represents the consensus of an international panel appointed by the American Heart Association Scientific Advisory and Coordinating Committee comprising clinical cardiovascular specialists and molecular biologists. This group has extensive experience with athletes of all ages but special expertise on the relationship between exercise and cardiovascular disease. The panel deliberated on the benefits and risks of recreational exercise in patients with GCVD and in this document has formulated prudent, contemporary, and practical consensus recommendations for physical activity in this group of cardiac patients.

The panel focused on physical activity for young people (<40 years of age) with those inherited and nonischemic cardiovascular diseases implicated most often in sudden cardiac death in young people, and specifically for HCM, the ion-channel diseases (LQTS and Brugada syndrome), ARVC, and diseases of connective tissue such as Marfan syndrome (and related vascular conditions, Ehlers-Danlos syndrome and other fibrillin disorders). Other uncommon familial conditions such as dilated cardiomyopathy or certain congenital heart malformations such as atrial or ventricular septal defect and mitral valve prolapse have not been specifically addressed here. However, it is possible to draw reasonable inferences from the recommendations presented and to extrapolate generally to these and other conditions.

Of particular note, the present recommendations assume the absence of important limiting cardiac symptoms, as well as an unequivocal cardiac diagnosis previously made on the basis of clinically overt features. Therefore, young people with negligible manifestations of disease, who may harbor only preclinical (ie, molecular) evidence of a particular disease-causing mutation predisposing to GCVD, are excluded.<sup>1,2,5,8,14,16,26,44,45</sup> Furthermore, at present, it is unresolved as to whether genotype-positive/phenotype-negative individuals warrant any restrictions from either recreational or competitive sports.

Issues directly related to preparticipation screening for cardiovascular disease<sup>46,47</sup> are beyond the scope of this document. The panel also recognizes that the present recommendations, in some instances, may be reasonably applied to patients older than 40 years (in the absence of important comorbidity), as well as to employment- and occupation-related activities involving physically vigorous and intense lifestyles (eg, firemen and other emergency personnel).

### **Premises**

The panel operated under several major conceptual premises in formulating the recommendations. First, vigorous physical activity may trigger sudden death in susceptible individuals with underlying heart disease.<sup>17,19–26,48,49</sup> Undoubtedly, there are other potential mechanisms, because sudden death is also known to occur with modest or sedentary activity or even during sleep, or to be triggered by abrupt or loud noises (such as in LQTS), but these variables remain largely undefined. Second, the vast majority of sudden deaths associated with GCVDs are due to primary ventricular tachyarrhythmias, although in Marfan syndrome sudden death is usually secondary to aortic dissection and rupture<sup>36</sup>; however, arrhythmic sudden death has also been reported.<sup>18</sup> Third, the risk for cardiac events associated with exercise in patients with GCVDs is, in theory, amenable to interventional strategies and therefore avoidable to a large extent. This latter tenet is fundamental to the present document because it supports the principle that sudden death may be prevented (or the risk substantially reduced) through lifestyle management and restriction of exercise or through the use of implantable devices.<sup>47,50–54</sup>

In formulating the present consensus recommendations, the expert panel considered their individual and collective experiences, as well as available and pertinent scientific data. However, panel members were required to confront areas in which there is a paucity of precise published evidence. Indeed, the panel encountered few absolutes or truly quantitative data that could be used to formulate recommendations regarding exercise programs, physical activity, or lifestyle in noncompetitive recreational situations.

Given these limitations, the panel has nevertheless endeavored to offer recommendations that can be regarded as a reasonable, practical, and conservative framework for practicing physicians in advising patients with GCVDs. These recommendations are not intended to be (and cannot be) rigid dicta but rather should be viewed as general guidelines, and not specific standards, that allow sufficient latitude to the managing physician for individual clinical judgment. Finally,

it was not the objective of the panel to restrict all exercise that could conceivably be associated with some increased risk but to develop a prudent balance between risk and benefit. Given the relative paucity of evidence in this area of medicine, particularly stringent recommendations would potentially (and unnecessarily) exclude a large proportion of patients with GCVD and in the process create a sedentary cardiac population deprived of the many benefits afforded by exercise for cardiovascular health.

### Role of Genetic Analysis

Mutation analysis for the purpose of risk stratification of sudden death, although prominently portrayed in the subspecialty literature,<sup>1-4,7-12,16,55-67</sup> has limited practical impact on the present considerations. First, the diagnostic strategy of DNA analysis is time consuming, expensive, and limited to a small number of highly specialized research-oriented laboratories<sup>1,2,8,64-67</sup> and thus is not yet routinely available to the practicing clinician for the purpose of patient management and formulation of exercise recommendations. Therefore, in the vast majority of circumstances, the clinician will be required to make exercise recommendations without data from genetic testing. Second, for the genetic diseases discussed here, genotype-phenotype correlations are too early in development to permit precise predictions of risk levels associated with recreational exercise for individual patients. Given these circumstances, the present exercise recommendations assume the absence of genetic data and presume the presence of a clinically apparent phenotype.

### Other Specific Considerations and Potential Limitations

The panel acknowledges the difficulties inherent in formulating arbitrary exercise recommendations, considering the subjectivity and many uncertainties or "gray areas" that are unavoidably involved, particularly for the diverse group of diseases addressed in this document. For example, there is marked heterogeneity in the phenotypic expression among these diseases and the variations in gene expression that influence individual patients. HCM is typically but not invariably associated with substantial left ventricular hypertrophy.<sup>26,68</sup> Conversely, LQTS and Brugada syndromes characteristically have no evidence of structural heart disease or abnormalities on gross or histopathologic examination.<sup>17,25,69</sup>

Consequently, it is not possible to tailor precise exercise recommendations for each of the many phenotypic and genetic patient subsets with varying levels of risk that have been defined within the broad clinical spectrum of each disease state. For example, although data are scarce, there is presently little evidence to suggest that the vast majority of genetically affected but phenotypically normal family members with HCM are at substantially increased risk for sudden death. Indeed, such individuals are probably at low risk; however, should the phenotype in HCM convert morphologically from nonthickened to hypertrophied,<sup>70,71</sup> it is possible for the risk level to increase in some patients. Therefore, there is insufficient formal scientific evidence for restricting such phenotypically negative individuals with HCM<sup>65</sup> from most modest recreational sports activities. However, this recom-

mendation is less compelling for LQTS, for which heightened risk has been reported in some phenotype-negative family members.<sup>3</sup>

In addition to the phenotypic and genetic variability among patients with GCVD, individual responses may differ within the context of the same sporting activities, ie, a recreational activity for one patient may be equivalent to a competitive sport for another. Also, the effect of exercise on a given patient with GCVD is dependent on several variables, including the intensity of the sporting activity, its physiological characteristics (eg, dynamic versus static) and duration (continuous versus intermittent), environmental conditions, and individual degree of hydration or use of medications. Although the impact that emotional and psychological investment in a recreational sports activity has on the underlying disease substrate and electrophysiological stability is not quantifiable, psychological stress is undoubtedly an important trigger influencing the likelihood of a sudden cardiac event.

Therefore, implementation of exercise recommendations ultimately depends in large measure on the interaction between physician and patient. It may often be necessary for clinicians to individualize exercise recommendations for particular patients, the specific GCVD involved, and the physical activity under consideration. These clinical decisions regarding the structure of exercise programs are also unavoidably influenced by liability issues and concerns, the possibility that physician recommendations may be ignored by some patients, and the variable tolerance for sudden death risk among patients and their families. Finally, the panel found it difficult to design recommendations that rely on obtaining truly quantitative measurements dependent on monitoring, such as maximum heart rate or metabolic equivalents with exercise, given the diverse sporting disciplines involved, the multitude of variables that impact these activities, and the impracticality of making accurate assessments during athletic activity.

## Recommendations

### General Principles

The conveyance of exercise and lifestyle recommendations to young patients with GCVD requires substantial physician-patient interaction to ensure that recommendations are translated in sufficient detail. On the other hand, arbitrary and rigid directives would be impractical and most likely ineffective.

In communicating with patients, it is useful to define informal recreational sports by contrasting such activities with competitive sports. Because of the unique structure and pressures of organized sports, athletes with heart disease engaged in competition may not always use proper judgment in prudently extricating themselves immediately from vigorous exercise, even should they recognize the potential medical need to terminate the activity. For example, dizziness, palpitations, fatigue, excessive dyspnea, or chest discomfort (or any other potential warning sign of cardiovascular disease) experienced during competitive sports may be difficult for the participant to distinguish reliably from those innocent sensations that can normally accompany intense and extreme exercise and mimic symptoms of cardiac disease.

However, such considerations are not generally part of truly recreational sports activity, constituting a clear distinction on which exercise recommendations can be effectively conveyed to patients with GCVD. Participants in recreational physical activities have greater opportunity to exert reasonable control over their level of exercise and therefore are more likely to reliably detect cardiac symptoms and willfully terminate physical activity. On the other hand, some recreational sports (eg, soccer, tennis, squash, and racquetball)<sup>72</sup> can become truly competitive, largely owing to the style of play and attitudes of the participants, and thereby may obscure these important distinctions.

We have not included within the definition of recreational sports those neighborhood and elementary school activities for young children that involve lesser degrees of physical intensity and which should be an allowed component of daily activity for individuals with GCVD. Of note, some mildly symptomatic GCVD patients may be under the misconception that their functional limitation can be overcome by physical activity, rather than regarding symptoms provoked by exercise as “warning signs” triggered by their underlying heart disease.

### Patient Recommendations

Sports activities have been categorized with regard to the high, moderate, and low levels of physical intensity required (Table). These 3 partitions relate to the generally expected degree of physiological exertion implied by a sporting discipline. Patients with GCVD can safely participate in most forms of recreational exercise judged to be of moderate or low intensity (equivalent to 4 to 6 and <4 metabolic equivalents, respectively; Table).<sup>73</sup>

Eligibility for exercise in specific recreational sports activities was assessed on a graded scale (from 0 to 5), with 0 to 1 designating activities generally not advised or strongly discouraged, 4 to 5 indicating activities probably permitted, and 2 to 3 indicating intermediate activities, which should be assessed on an individual basis. The assigned grades represent only an estimate, which assumes the usual level of physical exertion for a given recreational activity. However, in large measure, this grading system cannot take into account other potentially important variables such as the psychological burden and physical intensity uniquely brought to a sport by an individual participant or the potential effects of cardioactive drugs, environmental conditions, and the precise clinical profile. Therefore, these recommendations are, to a certain extent, necessarily subjective and represent only a starting point for clinical judgments in individual asymptomatic (or only very mildly symptomatic) patients with clinically evident GCVD. Consequently, their application requires substantial reliance on the practice of the “art of medicine” and the weighing of perceived risk with respect to benefit for each patient.

Furthermore, these recommendations are not intended for individuals with the following clinical features: history of important cardiac symptoms including syncope or other important episodes of impaired consciousness; prior cardiac operation (including surgical septal myectomy for obstructive HCM and aortic root reconstruction for Marfan syndrome)<sup>26,73</sup>

or heart transplantation; presence of an implanted cardioverter-defibrillator or pacemaker; and clinically overt and potentially life-threatening arrhythmias or other evidence of high-risk status. The presence of any of these features may require individual clinical judgment in adapting the present exercise recommendations.

The panel also found it useful to express specific exercise recommendations in terms of those activities that should be avoided by patients with clinically diagnosed GCVD:

- “Burst” exertion (or sprinting), characterized by rapid acceleration and deceleration over short distances. Exercise of this type is encountered in a variety of sports, such as basketball (particularly full-court play), soccer, and tennis. Therefore, preference is given to recreational sporting activities such as informal jogging without a training regimen, biking on level terrain, or lap swimming, in which energy expenditure is largely stable and consistent, even over relatively long distances or periods of time.
- Extremely adverse environmental conditions, which may be associated with alterations in blood volume, electrolytes, and state of hydration and thereby increase risk, such as greatly elevated or particularly cold temperatures disproportionate to that which the athlete is accustomed to in temperate climates (ie, >80°F [27°C] and <32°F [0°C]), high humidity, or substantial altitude.
- Exercise programs (even if recreational in nature) that require systematic and progressive levels of exertion and are focused on achieving higher levels of conditioning and excellence, as in road running, cycling, and rowing. Patients with GCVDs such as HCM, in which limiting dyspnea may occur with exercise, should be discouraged from any exertion that provokes these symptoms. These individuals are also advised against systematic training during which they are extended beyond the physical limits imposed by their underlying disease and the average aerobic state expected at that age.
- Excessive participation in sporting activities that otherwise would be regarded as recreational if performed in moderation, eg, downhill skiing continuously over an entire day versus more limited and selective skiing over the same time period.
- Exercise-related and adrenergic-type activities or stress that conveys a risk for cardiac events, specific to certain disease states. For example, in LQTS, swimming, abrupt loud noises (such as from a race starter’s pistol), and diving have been implicated as triggers for sudden death, particularly with certain mutant genes (ie, *KCNQ1* [or *LQT1*] for swimming and *KCNH2/HERG* [or *LQT2*] for auditory triggers).<sup>1,3,4,8,55–61</sup> However, such laboratory-based molecular information is unlikely to be available to clinicians prospectively making exercise recommendations. Patients with rare conditions such as CPVT,<sup>13,15</sup> in which many forms of exercise are associated with catecholamine release that triggers ventricular tachycardia, should be cautioned against virtually all forms of vigorous physical activity. The same restriction should be adopted for that subgroup of ARVC patients that shares with CPVT both effort-induced polymorphic ventricular tachycardia and a mutant ryan-

**Recommendations for the Acceptability of Recreational (Noncompetitive) Sports Activities and Exercise in Patients With GCVs\***

Intensity Level	HCM†	LQTS†	Marfan Syndrome‡	ARVC	Brugada Syndrome
<b>High</b>					
Basketball					
Full court	0	0	2	1	2
Half court	0	0	2	1	2
Body building§	1	1	0	1	1
Ice hockey§	0	0	1	0	0
Racquetball/squash	0	2	2	0	2
Rock climbing§	1	1	1	1	1
Running (sprinting)	0	0	2	0	2
Skiing (downhill)§	2	2	2	1	1
Skiing (cross-country)	2	3	2	1	4
Soccer	0	0	2	0	2
Tennis (singles)	0	0	3	0	2
Touch (flag) football	1	1	3	1	3
Windsurfing	1	0	1	1	1
<b>Moderate</b>					
Baseball/softball	2	2	2	2	4
Biking	4	4	3	2	5
Modest hiking	4	5	5	2	4
Motorcycling§	3	1	2	2	2
Jogging	3	3	3	2	5
Sailing	3	3	2	2	4
Surfing	2	0	1	1	1
Swimming (lap)	5	0	3	3	4
Tennis (doubles)	4	4	4	3	4
Treadmill/stationary bicycle	5	5	4	3	5
Weightlifting (free weights)§¶	1	1	0	1	1
Hiking	3	3	3	2	4
<b>Low</b>					
Bowling	5	5	5	4	5
Golf	5	5	5	4	5
Horseback riding§	3	3	3	3	3
Scuba diving	0	0	0	0	0
Skating#	5	5	5	4	5
Snorkeling	5	0	5	4	4
Weights (non-free weights)	4	4	0	4	4
Brisk walking	5	5	5	5	5

\*Recreational sports are categorized with regard to high, moderate, and low levels of exercise and graded on a relative scale (from 0 to 5) for eligibility with 0 to 1 indicating generally not advised or strongly discouraged; 4 to 5 indicating probably permitted; and 2 to 3 indicating intermediate and to be assessed clinically on an individual basis. The designations of high, moderate, and low levels of exercise are equivalent to an estimated >6, 4 to 6, and <4 metabolic equivalents, respectively.

†Assumes absence of laboratory DNA genotyping data; therefore, limited to clinical diagnosis.

‡Assumes no or only mild aortic dilatation.

§These sports involve the potential for traumatic injury, which should be taken into consideration for individuals with a risk for impaired consciousness.

||The possibility of impaired consciousness occurring during water-related activities should be taken into account with respect to the clinical profile of the individual patient. Barotrauma is a primary risk associated with the use of the scuba apparatus in Marfan syndrome.<sup>73</sup>

¶Recommendations generally differ from those for weight-training machines (non-free weights), based largely on the potential risks of traumatic injury associated with episodes of impaired consciousness during bench-press maneuvers; otherwise, the physiological effects of all weight-training activities are regarded as similar with respect to the present recommendations.

#Individual sporting activity not associated with the team sport of ice hockey.

dine receptor.<sup>10</sup> It is also of note that a temperature-dependent dysfunction of the *SCN5A* gene for cardiac sodium channel has been characteristically observed in patients with Brugada syndrome (and Thr1620Met missense mutation).<sup>74</sup> This increased temperature sensitivity could predispose some Brugada patients to life-threatening arrhythmias either during a febrile state or when body temperature increases during intense physical exertion.

- Intense static (isometric) exertion, such as lifting free weights, may prove to be adverse by inducing a Valsalva maneuver and dynamic left ventricular outflow obstruction in HCM<sup>75</sup> (as well as the risk for traumatic injury in the event of impaired consciousness) or by increasing wall stress and weakening of the aortic media in Marfan syndrome, particularly if aortic dilatation is already present.<sup>62,73</sup>
- Patients with diseases associated with impaired consciousness (eg, syncope and near-syncope) are subject to considerably higher risk for traumatic injury while engaged in certain sports such as free weight and bench-pressing maneuvers, downhill skiing, diving, ice hockey, rock climbing, motorcycling, and horseback riding, and this factor should be taken into consideration in making recommendations to individual patients with GCVD.
- Although data are lacking, it is reasonable to specifically caution patients with GCVD, particularly those with catecholamine-sensitive or auditory-triggered arrhythmia syndromes such as LQTS and CPVT, against amusement park rides (eg, roller coasters and thrill-related or frightening rides) because these are associated with intense stress and emotion due to sudden acceleration in heart rate and abrupt changes in centrifugal or centripetal forces.
- Paired athletic activities in which a second party may be at risk should the individual with GCVD suddenly incur bodily injury or impairment of consciousness and incapacitation, eg, in recreational sports such as scuba diving and rock or mountain climbing. Water sports such as scuba diving or diving from platforms into pools are also generally unacceptable by virtue of their exposure of patients with GCVD (for whom syncope is a not-uncommon manifestation) to the risk of underwater drowning and reduced probability of rescue.
- Extreme sports (such as hang gliding and bungee jumping) are activities that are best avoided because they require the expenditure of particularly substantial physical energy and incur psychological demands that are exceedingly unpredictable, placing individuals with GCVD in compromised circumstances in which the likelihood of injury is substantial and the possibility of rescue from a traumatic or cardiovascular event is greatly reduced.
- Concomitant use of substances or compounds professed to promote enhanced physical performance but that also harbor the potential for adverse effects, particularly when associated with disease states or extreme environmental conditions, ie, cocaine, anabolic steroids, or dietary and nutritional supplements such as ma huang, an herbal source of ephedrine (ie, elemental ephedra) and a cardiac stimulant that is potentially arrhythmogenic.<sup>76–79</sup>

## Special Circumstances

### Physical Education Class

Issues related to recreational exercise often arise with regard to compulsory physical education classes in junior high or elementary school. In the former, the required levels of exercise in physical education classes vary considerably but often involve vigorous exertion and circumstances that are difficult to control. For example, although many components of such classes may be truly recreational, others clearly are not and can be regarded as competitive in nature, ie, prolonged aerobic events such as the traditional timed 600-yard (or 400-meter) run and the President's Physical Fitness Award. On the other hand, in elementary school physical education class, sports activities may involve nothing more than innocent play.

Although in the view of some very conservative clinicians, it might seem most prudent for patients with GCVDs to largely avoid any involvement in school-structured physical education, we recognize that profound personal and psychological stigmata can be associated with such selective, targeted, and often unnecessary gym class prohibition. Indeed, in some states, such children are placed in alternative health classes, which results in further ostracism by their peers. In the event that the patient and family judge it important, for social and peer-related reasons, to retain some level of participation and normalcy among peers (as is often the case), careful and detailed review of physical education class requirements should be undertaken by the parents in concert with school officials and their physician, during which the principles of safe recreational activities are agreed upon.

This process will define acceptable portions of the curriculum and exclude activities that can be regarded as intense or competitive (particularly those involving burst exertion) although it may be challenging to distinguish recreational from competitive activity under these circumstances. In this regard, it should be determined whether physical education instructors will agree to (or are capable of) monitoring the patient's activity level in accord with the medical recommendations. Therefore, the degree to which a child or adolescent with GCVD can or should participate in physical education class requires a large measure of individualization.

### Implantable Cardioverter-Defibrillator

A growing number of young patients with GCVDs are receiving implantable defibrillators for primary or secondary prevention of sudden death.<sup>50,65,80,81</sup> Such patients may participate in a wide variety of noncompetitive and noncontact physical activities in concert with the exercise recommendations of the responsible electrophysiologist. However, the defibrillator itself may create some restrictions to these activities, particularly with regard to the possibility that bodily trauma may disrupt the lead system or that certain levels of physical exertion may trigger an inappropriate shock due to sinus tachycardia. Therefore, young patients with GCVD undergoing implantation of a cardioverter-defibrillator should receive a dual-chamber device with algorithms that improve discrimination between ventricular and supraventricular arrhythmias, thereby reducing the incidence

of inappropriate interventions.<sup>80</sup> Participation in intense exercise should not be advocated explicitly because of the presence of an implanted defibrillator and the antiarrhythmic protection afforded by the device.<sup>29,81</sup>

### Legal Considerations

These recommendations are intended to facilitate the provision of high-quality patient care, to provide guidance to physicians, and to enable clinicians to base recommendations regarding medically acceptable recreational exercise programs for patients with known GCVDs on more than their own personal expertise and experience. In *Knapp v. Northwestern University*,<sup>81</sup> a federal appellate court recently recognized the appropriateness of physician reliance on consensus recommendations and guidelines to determine medically reasonable levels of athletic activity for persons with cardiovascular abnormalities.

Currently, there is no well-defined legal precedent regarding potential legal liability if a physician deviates from consensus medical recommendations or guidelines based on the collective judgment of experts. The law generally requires a physician to have and use the knowledge, skill, and care ordinarily possessed and employed by members of the 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 particular jurisdiction, means either "reasonable," "customary," or "accepted" medical care under the circumstances.<sup>82</sup>

Courts 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.<sup>83–85</sup> In establishing a recreational exercise program for patients with known GCVDs, physician deviation from consensus guidelines in a particular individual case does not necessarily constitute medical malpractice if consistent with acceptable medical practices under the circumstances. The controlling legal issue is whether adherence to (or deviation from) consensus guidelines is consistent with reasonable, customary, and accepted medical practice in an individual patient.

Consistent with this legal principle, the medical recommendations set forth in this document should not and cannot be viewed as inflexible doctrine but rather should allow sufficient latitude to the managing physician without precluding individual clinical judgment. Compliance with these recommendations is evidence that a physician has satisfied these legal requirements and may form the basis of a successful defense to allegations of malpractice.<sup>86</sup>

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