Bethesda Conference #36 and the European Society of Cardiology Consensus Recommendations Revisited

A Comparison of U.S. and European Criteria for Eligibility and Disqualification of Competitive Athletes With Cardiovascular Abnormalities

Antonio Pelliccia, MD,* Douglas P. Zipes, MD,† Barry J. Maron, MD‡

Rome, Italy; Indianapolis, Indiana; and Minneapolis, Minnesota

Aspiration to reduce the risks of athletic field deaths prompted the American Heart Association and European Society of Cardiology (ESC) to establish consensus guidelines for eligibility/disqualification decisions in competitive athletes with cardiovascular abnormalities. Since 2005, the Bethesda Conference #36 and the ESC consensus documents have been relied upon by physicians from different parts of the world. The 2 consensus documents emanate from largely different cultural, social, and legal backgrounds existing in the U.S. and Europe and, although several recommendations are similar, in some instances the Bethesda Conference #36 and the ESC consensus documents suggest different approaches to disqualification decisions and implications for clinical practice, raising the possibility that confusion and discrepancies will contaminate the management of competitive athletes with cardiovascular disease. In the present article, the differences between the 2 documents are critically viewed, with special attention to genetic cardiovascular diseases relevant to sudden death in young athletes, through the prism of different cultural backgrounds, societal attitudes, and also perceptions regarding exposure to legal liability in the U.S. and Europe. In conclusion, it seems appropriate at some time to consider assembling updated recommendations for sports eligibility/disqualification that assimilate both the U.S. and European perspectives, with the aspiration of creating a unique and authoritative document applicable to the global sports medicine community. (J Am Coll Cardiol 2008;52:1990–6) © 2008 by the American College of Cardiology Foundation

Over the last several years, issues related to the sudden deaths of young competitive athletes due to underlying (and usually unsuspected) cardiovascular disease have become highly visible and ever more complex medical and public health topics (1–3). Although initially promoted largely in the U.S., interest in this problem has become increasingly international, now with an intense focus in Europe and particularly in Italy. In addition, competitive sports have progressively evolved toward globalization, as evident by the changing demographics of elite athletes engaged in professional sports, such as soccer, basketball, baseball, boxing, and ice hockey.

Aspirations to reduce the risks of the athletic field related to cardiovascular disease was initially formalized by the American College of Cardiology (ACC) in the 1985 Bethesda Conference #16, in which an expert panel was convened to establish consensus guidelines for eligibility/disqualification decisions in competitive athletes with cardiovascular abnormalities (4). In the ensuing 20 years there have been 2 updated Bethesda Conferences (#26 and #36 in 1994 and 2005, respectively) (5,6). Also, in 2005 a parallel consensus document from the European Society of Cardiology (ESC) addressed the same issues related to the management and participation in competitive sports of athletes with cardiovascular abnormalities (7).

The rationale for offering both the U.S. and European expert consensus documents is the widely accepted perception, supported by substantial circumstantial evidence (2,3), that certain susceptible athletes with clinically silent cardiovascular disease harbor increased risk for sudden death or disease progression, by virtue of their commitment to intensive training and competition. Conversely, the removal of athletes from this lifestyle is regarded as a mechanism by which this risk might be substantially reduced (8,9).

Both the ACC Bethesda Conference #36 (BC#36) and the ESC documents provide specific recommendations with respect to different cardiac abnormalities and sports, on the basis of available scientific data, as well as the personal experience of the panel participants. For the most part, the formulated recommendations cannot be viewed solely as evidence-based medicine but must also be viewed as the...
Hypertrophic Cardiomyopathy (HCM)

HCM is the most common cause of sudden death in U.S. competitive athletes (2,3) and is prominently assessed in both documents, which are similarly restrictive with respect to the recommendations for competitive athletes with this disease. Although the phraseology of disqualification guidelines differ, the sense is the same in both documents (i.e., athletes with HCM are excluded from most competitive sports, with the possible exception of those characterized by low static and low dynamic intensity, such as golf) (6,7). These recommendations are based on the assumption that competitive sports participation might itself constitute a risk factor for sudden death on the athletic field due to HCM, usually because of ventricular tachyarrhythmias (2). Such events can be promoted by those specific variables related to the stress of sports (e.g., electrolyte imbalance and hemodynamic and autonomic changes as well as other still undefined mechanisms). Therefore, a competitive athlete with HCM judged to be “low risk” in the absence of all traditional risk markers might nevertheless be at unacceptably increased risk solely by virtue of involvement in high-intensity competitive sports, a principle illustrated by the sudden death of the professional Cameroon soccer player Marc Vivien Foé (10).

However, the BC#36 and ESC panels made different recommendations for those HCM family members without the characteristic phenotype (i.e., left ventricular hypertrophy) but with disease-causing sarcomeric mutations (i.e., gene-positive, phenotype-negative individuals, known also as gene carriers) (6,7).

BC#36. Individuals identified as gene carriers are not precluded from participation in competitive sports, given that there has been, thus far, little or no evidence of adverse cardiac events in such individuals and therefore no compelling reason to deprive them of the many potential benefits (including economic) derived from sport participation.

ESC. In contrast, the ESC document is more restrictive with regard to HCM gene carriers, given that the natural history of such individuals is at present largely unknown. All competitive sports are excluded and only noncompetitive or leisure-time sporting activities are recommended. This recommendation is based on the hypothesis that regular exercise training and competitive sports can play a role in triggering cellular mechanisms leading to the HCM phenotype (i.e., left ventricular hypertrophy) and clinical evolution (i.e., tachyarrhythmia) in the presence of a predisposing gene abnormality.

However, there are no firm clinical or experimental data to support either position, and the recommendations are currently based on what seems to be most reasonable to the U.S. or ESC expert panel, respectively.

Ion Channelopathies

Ion channelopathies include long-QT syndrome (LQTS), Brugada syndrome, and catecholaminergic polymorphic ventricular tachycardia (CPVT). The greatest attention in the recommendations is on LQTS. The short-QT syndrome is not mentioned in either document.

Clinical diagnosis of LQTS is based predominantly on the measurement of the QT interval on the 12-lead electrocardiogram (ECG), corrected for heart rate (11). However, unequivocal identification of LQTS by this methodology can be challenging, because QT interval corrected for heart rate (QTc) might be borderline or even within normal limits in a large proportion of genetically proven LQTS patients. Conversely, occurrence of QTc interval above the widely used upper limits of 0.44 s in male subjects or 0.46 s in female subjects is not an uncommon finding in young...
trained athletes (12). Finally, and surprisingly, many cardiologists fail to measure the QT interval accurately (13).

**BC#36.** For diagnosis of LQTS in athletes the QTc interval should exceed 0.47 s in male subjects and 0.48 s in female subjects. When this diagnosis is made, the recommendation is for restriction of athletes from competitive sport, except those with low intensity.

Although the risk of sudden death is probably not zero in genotype-positive, phenotype-negative individuals, this document states that, on the basis of the available scientific evidence, it is not justifiable to preclude such individuals from competitive sports. Furthermore, most information indicates that serious arrhythmias are uncommon in individuals with QTc interval <500 ms. A unique recommendation applies to such individuals with LQT1 mutation, who should refrain specifically from competitive swimming, because of the strong association between this sport and cardiac events (6).

**ESC.** Threshold values accepted for diagnosis of LQTS are lower (i.e., QTc interval 0.44 s in male subjects and 0.46 s in female subjects). In athletes with QTc interval lengthening above these limits, genetic testing is recommended to increase the likelihood of definitive diagnosis. When the LQTS diagnosis is confirmed, the recommendation is for exclusion of the athlete from all competitive sports.

Asymptomatic genotype-positive, phenotype-negative individuals with proven mutation and normal QTc interval on 12-lead ECG are discouraged from participation in all competitive sports. Finally, no specific recommendations are provided for athletes with borderline QTc interval and negative genotyping (which might represent false negative results of genetic testing) (7). In current practice, such athletes are allowed to participate in competitive sports with close periodic surveillance.

With regard to individuals with definite diagnosis of Brugada syndrome and CPVT, both documents restrict participation. Although a clear association between exercise and sudden death in the Brugada syndrome has not been established (and because of the potential impact of hypothermia), disqualification from all competitive sports is nevertheless recommended by the ESC document, with a potential exception in BC#36 only for low-dynamic and low-static sport (6,7). Differences between the BC#36 and ESC documents are raised with regard to gene carriers of Brugada syndrome and CPVT. According to BC#36, gene carriers without the phenotype (in the absence of symptoms and ventricular tachyarrhythmia inducible at electrophysiologic study) should not be precluded from participation in competitive sports. In contrast, the ESC document states that all gene carriers (without the phenotype) should be restricted from competitive sport (6,7).

**Marfan Syndrome**

Marfan syndrome is characterized by a constellation of skeletal, cardiovascular, ocular, skin, pulmonary, and central nervous system anomalies that might vary considerably in severity, making clinical diagnosis challenging in individual patients. Both documents rely on the Ghent nosology for the Marfan diagnosis, which includes major and minor criteria for the affected systems and organs (14).

**BC#36.** Recommendations are based largely on aortic root dimension, with close monitoring by echocardiography suggested at 6-month intervals. The BC#36 recommends participation in low and even moderate competitive sports, provided that the following features are absent: aortic root dilation (i.e., transverse dimension ≥40 mm or more than 2 SDs from the mean for body surface area in children), moderate-to-severe mitral regurgitation, and family history of aortic dissection or sudden death (6).

**ESC.** For the Marfan diagnosis, the ESC document emphasizes the role of genetic screening for fibrillin 1 mutations. When the diagnosis of Marfan syndrome is definitive (by phenotypic recognition and/or genetic testing), athletes are disqualified from all competitive sports, independent of the precise aortic root dimension. When the Marfan diagnosis is uncertain, due to incomplete phenotypic expression in young athletes and/or if genetic testing is unavailable or negative, the ESC sanctions continued sport participation, although with periodic clinical and diagnostic re-evaluation (7).

**Arrhythmias**

In the evaluation of athletes with arrhythmias, the BC#36 document usually permits greater autonomy and individualized discretion compared with the ESC. For example, electrophysiologic study and radiofrequency ablation are often mandatory procedures for the ESC (but not in the BC#36) in the assessment and management of athletes with arrhythmias desiring to resume competitive sport activity.

**Wolff-Parkinson-White (WPW)**

**BC#36.** In asymptomatic athletes with WPW syndrome, this document does not consider electrophysiologic study to be mandatory for risk assessment, but such testing is required in athletes with symptoms such as impaired consciousness or persistent palpitations (due to documented supraventricular tachycardia) or when an ablation procedure is otherwise recommended. However, an electrophysiologic study is considered advisable in asymptomatic athletes if engaged in moderate- or high-level competitive sports (6).

**ESC.** In contrast, the ESC mandates that all athletes with WPW undergo complete risk assessment including electrophysiologic study (7). After risk stratification, the 2 documents are similar regarding the recommendations for sport participation. Namely, both BC#36 and ESC state that athletes judged to be at increased risk on the basis of electrophysiologic study and those who are symptomatic with atrial flutter/fibrillation or syncope should undergo radiofrequency ablation of the accessory pathway to retain athletic eligibility. In addition, the ESC recommends that
managing physicians advise athletes judged not to be at high risk of the potential benefits of radiofrequency ablation; however, such athletes are not restricted from competitive sports if they should refuse to undergo ablation.

Finally, BC#36 suggests resumption of competitive sport shortly after successful ablation (i.e., 4 weeks), but ESC postpones return to competition until 3 months after the procedure.

Premature Ventricular Complexes (PVCs)

Both the BC#36 and the ESC caution that PVCs might be the initial manifestation of clinically silent arrhythmogenic conditions associated with risk of sudden death, such as HCM, arrhythmogenic right ventricular cardiomyopathy, or myocarditis, and therefore require cardiac evaluation to exclude the underlying pathologic condition (6,7,15). Differences, however, emerge with regard to the criteria recommended for cardiac assessment and risk stratification.

BC#36. Evaluation is based largely on noninvasive testing (exercise testing and/or ECG Holter monitoring). Because deconditioning can result in a significant reduction in the frequency and complexity of ventricular arrhythmias, a short period of detraining can be considered in some athletes (16). Athletes without demonstrable cardiac disease have no limitations for competitive sports participation. Restriction to low-intensity sports is, nevertheless, indicated for athletes in whom PVCs increase in frequency during effort (or exercise testing) and produce symptoms (such as impaired consciousness, disproportionate fatigue, or dyspnea) (6).

ESC. Evaluation might include electrophysiologic study or selective invasive testing in individual athletes according to the suspected cardiac lesion. In the absence of cardiac disease, athletes with PVCs have no restriction from competitive sports; however, limitations might be sanctioned when there is a family history of sudden death, when there are symptoms on effort (syncope, palpitations, excessive fatigue), when arrhythmias worsen during exercise, or when frequent couplets with short RR interval are present (7).

Nonsustained Ventricular Tachycardia

BC#36. Asymptomatic athletes without structural cardiac disease are eligible for all competitive sports if nonsustained ventricular tachycardia bursts on ambulatory (Holter) ECG are short (<10 beats) at <150 beats/min and demonstrate suppression (or no significant worsening) during exercise (6).

ESC. Asymptomatic athletes without structural cardiac disease can participate in all competitive sports if nonsustained ventricular tachycardia is rare, is not triggered by exercise, presents without short RR interval, and occurs in the absence of a family history of sudden death (7).

Implantable Cardioverter-Defibrillators (ICDs)

The issue of whether athletes with ICDs (implanted for primary or secondary prevention) should have access to competitive sports has recently generated controversy (17,18). However, the BC#36 and the ESC recommendations are in agreement on this point. Both documents consistently recommend restriction from competitive sport activities in athlete patients with ICDs, with the possible exception of some low-intensity sports without associated risk of trauma to the device, such as golf (6,7). Furthermore, the desire of a high-risk athlete to remain in competitive athletics should not represent the primary indication for an ICD.

Pre-Participation Screening and Diagnosis of Cardiovascular Disease

The methodologies by which cardiovascular diseases are identified and competitive athletes come to evaluation for sports eligibility show notable differences between European countries (namely, Italy) and the U.S. In Italy, a national pre-participation screening and medical clearance program has been mandated for competitive athletes over the last 25 years (19). The Italian program has been unique in the world by virtue of requiring annual evaluations that routinely include 12-lead ECGs in addition to a medical history and physical examination. Other European countries have either limited or no medical screening programs for competitive athletes, although wide implementation of the Italian screening model has been promoted by the endorsement of the International Olympic Committee medical commission and the ESC in 2004 and 2005, respectively (20,21). Recently, scientific evidence has emerged showing this screening strategy to be useful for identification of HCM (and other cardiomyopathies) in asymptomatic athletes (22), and implementation of this program in Italy has been reported to be associated with substantial reduction in mortality due to cardiomyopathies (23).

Conversely, customary screening strategies for U.S. high school and college athletes is confined to medical history and physical examination (24), which is more limited in its power to consistently identify important cardiovascular diseases (3). Furthermore, in contrast to Italy, where screening is under the responsibility of specialized sports medicine physicians, in the U.S. physicians often volunteer or a variety of health care workers conduct pre-participation screening (24).

Current diagnosis of inherited arrhythmogenic cardiomyopathies might be greatly improved by the genetic testing of asymptomatic athletes. However, molecular analysis is currently not routine testing, either in the European or U.S. screening programs, but is performed selectively (e.g., when LQTS or Marfan syndrome is suspected). Moreover, protocol for protecting the personal genetic information of
highly visible athletes is not specifically discussed in either the European or U.S. recommendations.

Comment

The recommendations for eligibility/disqualification of competitive athletes with cardiovascular diseases can be regarded in many instances as more restrictive in the ESC consensus document than in BC#36, leaving much less discretion to the managing physician. This difference largely reflects the influence of the sports medicine model developed in Italy, which the ESC document greatly relies upon (7). In Italy, a state law has been implemented since 1982 by decree of the Ministry of Health that dictates that all citizens participating in official competitive sports events should undergo annual medical assessment and, if free of cardiovascular or systemic disease, obtain a certification of eligibility (19). Although no such state law exists in other European Union countries (or anywhere else in the world, including the U.S.), this influence is nevertheless evident in the ESC document, which was originally conceived and largely promoted by Italian clinical scientists.

In Italy, there is a standard and well-defined process grounded in the legislation, whereby the sport medicine specialist is the ultimate authority, entrusted with the responsibility for decision-making as well as for enforcement of eligibility/disqualification decisions. No such program exists in the U.S., where the process is generally more heterogeneous. The unique Italian situation unavoidably creates a tendency for managing physicians to be instinctively conservative in making decisions concerning athletic eligibility. In particular, disqualification from competitive sports is not necessarily confined to athletes with conclusive evidence of cardiovascular disease (as is generally the case in the U.S.) but also targets those with probable diagnoses of cardiovascular disease as well as in some circumstances in which the potentially deleterious impact of sport activities on clinical course and outcome is unresolved.

These more conservative attitudes are evident in many respects in the ESC document, with examples that include athletes occasionally identified without phenotypic expression but only as gene carriers for hypertrophic cardiomyopathy or ion channelopathies (i.e., LQTS, Brugada syndrome, and CPVT). Such athletes are currently restricted from competitive sports participation by ESC recommendations but not necessarily by BC#36 (6,7). Because the long-term consequences of an athletic lifestyle on gene carriers with HCM or ion channelopathies (namely, phenotypic and clinical evolution in the presence of a susceptibility mutation) are still largely unresolved, the ESC recommendations are more cautious and consistent with the practice endorsed by the sports medicine law in Italy (19). In the U.S., disqualification of young athletes with inherited cardiovascular (or other medical) abnormalities can be recommended by the managing physician, but the high-school or college officials are ultimately responsible for decisions regarding participation in competitive sports. Therefore, in contrast to Italy, where the sports medicine specialist retains the authority to enforce disqualification, the same circumstances do not apply to U.S. athletes for whom no federal or state law governs medical clearance for high school or college sports.

### Table 1 Summary of Selected Differences Between BC#36 and ESC Recommendations for Competitive Athletes With Selected CV Abnormalities

<table>
<thead>
<tr>
<th>Clinical Criteria and Sports Permitted</th>
<th>BC#36</th>
<th>ESC</th>
</tr>
</thead>
<tbody>
<tr>
<td>Gene carriers without phenotype (HCM, ARVC, DCM, ion channel diseases*)</td>
<td>All sports</td>
<td>Only recreational sports</td>
</tr>
<tr>
<td>LQTS</td>
<td>&gt;0.47 s in male subjects, &gt;0.48 s in female subjects</td>
<td>&gt;0.44 s in male subjects, &gt;0.46 s in female subjects</td>
</tr>
<tr>
<td>Marfan syndrome</td>
<td>If aortic root &lt;40 mm, no MR, no familial SD, then low-moderate intensity competitive sports permitted</td>
<td>Only recreational sports</td>
</tr>
<tr>
<td>Asymptomatic WPW</td>
<td>EPS not mandatory</td>
<td>EPS mandatory</td>
</tr>
<tr>
<td>Premature ventricular complexes</td>
<td>All competitive sports (restriction for sports in dangerous environment)†</td>
<td>All competitive sports (restriction for sports in dangerous environment)†</td>
</tr>
<tr>
<td>Nonsustained ventricular tachycardia</td>
<td>If no CV disease, all competitive sports</td>
<td>If no CV disease, all competitive sports</td>
</tr>
<tr>
<td></td>
<td>If CV disease, only low-intensity competitive sports</td>
<td>If CV disease, only recreational sports</td>
</tr>
</tbody>
</table>

*Long-QT syndrome (LQTS), Brugada syndrome, catecholaminergic polymorphic ventricular tachycardia; †sports in dangerous environments are restricted, given the risk should impaired consciousness occur, such as motor sports, rock climbing, and downhill skiing.

ARVC = arrhythmogenic right ventricular cardiomyopathy; BC#36 = Bethesda Conference #36; CV = cardiovascular; DCM = dilated cardiomyopathy; EPS = electrophysiologic study; ESC = European Society of Cardiology; HCM = hypertrophic cardiomyopathy; MR = magnetic resonance; PVC = premature ventricular complex; SD = sudden death; WPW = Wolff-Parkinson-White syndrome.
Disqualification decisions from competitive sports on the basis of a medical disability condition have on occasion been regarded by U.S. high-school or college athletes as a limitation on personal freedom or even as a matter of discrimination. For example, in one highly visible case regarding sanctioned college sports (i.e., Knapp v. Northwestern University) (25), a basketball player with HCM and a secondary prevention implantable defibrillator used the Rehabilitation Act to argue that Northwestern University had violated his rights in a discriminatory fashion by disqualifying him from the college basketball team on medical grounds. In this case, however, an appellate court upheld the right of the college to exclude such an athlete on the basis of the medical disability, given that college sports programs cannot be regarded as matters of civil liberty in which the sole consideration is individual responsibility: “playing intercollegiate sports cannot be held out as a necessary part of learning in all students…” (26). This case represents a unique legal precedent in the U.S., allowing the educational institution to selectively enforce disqualification in cases involving medical disability and, in that instance, cardiovascular disease.

For those athletes outside of sanctioned high school or college sports, such as professionals, participation in competitive sports (even in the presence of known cardiovascular disease) is not associated with any legal precedent or formalized disqualification and more often regarded largely as a matter of autonomous and individual responsibility. Indeed, the absence of a medical–legal framework, such as in Italy, permits U.S. professional athletes considerably more latitude in disputes over cardiovascular abnormalities and medical eligibility, which can lead athletes to look for multiple medical consultations until the desired recommendation is achieved (i.e., "shopping") (1,2). Indeed, for professional athletes the issue of criteria for disqualification is less formalized and therefore more complex, given that such individuals are usually not minors, enter a binding labor agreement with their teams, and face the potential cessation of employment, with substantial loss of economic benefits and other opportunities derived from professional sport activities should their eligibility be terminated.

Another paradoxical concern related to professional and some elite college athletes is the possibility of legal liability for the physician, by virtue of recommending disqualification from sports participation with the objective of protecting the athlete from the hazards of competition (27).

Conclusions

The ESC and BC#36 consensus documents, although generally similar with regard to recommendations, do demonstrate several important differences related to the assessment of sudden death risk during competitive sports and the criteria applied for disqualification of athletes with cardiovascular disease. It would seem that many of these distinctions, as outlined here, can be explained on the basis of differences in Europe and the U.S. with regard to cultural background, societal attitudes, and also perceived exposure to legal liability.

However, it would be useful to the sports medicine community to assemble an authoritative consensus document from both the ESC and the ACC (as well as sports medicine scientific associations) to provide common recommendations for sports eligibility/disqualification that could be implemented globally. Therefore, in the best interest of competitive athletes, it might be appropriate at some time in the near future to update recommendations in a collaborative fashion, assimilating all different perspectives, with the aspiration of creating a single and respected consensus document applicable to sports medicine worldwide.

Reprint requests and correspondence: Dr. Antonio Pelliccia, Institute of Sports Medicine and Science, Largo Piero Gabrielli 1, 00197 Rome, Italy. E-mail: antonio.pelliccia@coni.it or ant.pelliccia@libero.it.

REFERENCES

17. Maron BJ, Zipes DR. It is not prudent to allow all athletes with implantable-cardioverter defibrillators to participate in all sports. Heart Rhythm 2008;5:864–6.

Key Words: cardiovascular disease • competitive athletes • guidelines for eligibility/disqualification.