

HISTORICAL CONTEXT

There have been 3 prior documents, all sponsored by the American College of Cardiology (ACC) (1-3), that addressed eligibility and disqualification criteria for competitive athletes with cardiovascular diseases: Bethesda Conferences 16 (1985), 26 (1994), and 36 (2005), published and used over a 30-year period. Each of the 3 initiatives (and the present American Heart Association (AHA)/ACC scientific statement) were driven by the tenet that young trained athletes with underlying cardiovascular abnormalities are likely at some increase in risk for sudden cardiac death (usually on the athletic field) compared to nonathletes or competitive athletes without cardiovascular disease (4-8).

All 3 Bethesda Conferences and the present derived AHA/ACC document provide expert consensus recommendations. These insights use 1) the experience and expertise of the panelists (i.e., individual and collective judgments, using the “art of medicine”) and 2) available scientific evidence that estimates the medical risk in athletes with underlying acquired, genetic, and congenital heart abnormalities imposed by the unique lifestyle of engagement in competitive sports.

These insights can be applied to decision making for temporary or permanent disqualification versus eligibility of athletes with probable or conclusive evidence of cardiovascular disease; however, the scientific data supporting many of the recommendations in this document are unavoidably limited, as evidenced by the frequent assignment of a Level of Evidence C. Nevertheless, each of the 3 prior Bethesda Conferences has served the practicing community well, offering clinicians a consensus reference document that is potentially helpful in resolving predictably difficult clinical dilemmas. It is our expectation that the present conservative AHA/ACC scientific statement will follow in that tradition. The final document was approved by all participants and assigned outside reviewers.

IMPETUS FOR THE PRESENT DOCUMENT

There are a number of factors that support the decision to update the 36th Bethesda Conference here (3). First, sudden cardiac deaths in young healthy athletes remain tragic and counterintuitive events, subject to persistently high public visibility, emotion, and media scrutiny, with potential legal liability considerations. Therefore, a strong impetus remains to identify high-risk athletes to reduce their exposure to sudden death risk. Indeed, there is an ever-expanding population of competitive athletes, including those participating in new and emerging organized sports. Second, cardiovascular medicine changes rapidly. As evidence of this, in the almost 10 years since

publication of the 36th Bethesda Conference (3), new conditions associated with sudden death in the young have been recognized, and knowledge of the responsible diseases and inherent risks of sudden cardiac death in the young has evolved (4-8). As a result, some selected areas of the 36th Bethesda Conference may have become obsolete, and novel issues not previously addressed, have emerged. Third, an increasing number of adults with congenital heart disease and cardiomyopathies are now being recognized (often with surgical palliation or correction) who wish to engage in competitive athletics and require contemporary recommendations. In addition, the increasing penetration into cardiovascular practice of implantable devices (e.g., pacemakers and cardioverter-defibrillators) has created greater numbers of physically active young people with genetic heart diseases who have had devices implanted and who may aspire to participation in competitive athletics. Recently, there has been greater deployment of automatic external defibrillators at athletic events in recognition of sudden death risk in young athletes. Finally, the practicing cardiovascular community deserves and expects the most up-to-date information on which to make important clinical decisions regarding eligibility versus disqualification of competitive athletes.

DEFINITIONS

As in the 3 Bethesda Conferences (1-3), the basic definition of a competitive athlete remains unchanged: 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 principle concerns whether competitive athletes with either known or unsuspected cardiovascular disease can be expected to properly judge when it is prudent to terminate physical exertion. Indeed, the unique pressures of organized sports do not allow athletes to exert strict individual control over their level of exertion or reliably discern when cardiac-related symptoms or warning signs occur that should dictate termination of the activity.

Furthermore, it is emphasized that these AHA/ACC recommendations should not be regarded as a general overriding injunction against all forms of exercise. Notably, this document is concerned only with organized and sanctioned competitive sports participation, such as most commonly found in middle school, high school, and college (1-3), and not with purely recreational physical activities (9). The panel recognizes and strongly supports the well-documented health benefits of exercise, with

regular physical activities encouraged for those people who have been removed from organized competitive athletics, or who elect to participate in a wide range of recreational sporting activities.

Although the Bethesda Conferences and the present document are largely focused on student-athletes of high school and college age (primarily 12 to 25 years old), the panel recognizes the need to also be more expansive with regard to age of the athletes, that is, that participation in competitive athletics now increasingly begins earlier in a variety of youth sports before high school and continues beyond college. This consideration is also substantiated by the realization that inherited arrhythmia syndromes can impact very young people and that patients with genetic, congenital, or acquired heart diseases now engage in competitive athletics at more advanced ages. However, because systematic preparticipation screening in the United States does not usually begin before high school (4), recognition of cardiovascular disease in such younger athletes is unpredictable.

CAUSES OF SUDDEN DEATH IN ATHLETES

The cardiovascular causes of sudden death in young athletes have been well documented in forensic databases (5-9). These deaths occur in both sexes (although more commonly in males, by 9:1); in minorities, prominently including African-Americans and in a wide range of individual and team sports. In the United States, among people <35 years old, genetic heart diseases predominate, with hypertrophic cardiomyopathy being the most common, accounting for at least one-third of the mortality in autopsy-based athlete study populations (5-7). Congenital coronary anomalies (usually those of wrong sinus origin) are second in frequency, occurring in ≈15% to 20% of cases. Other less common diseases, each responsible for ≈5% or fewer of these sudden deaths, include myocarditis, aortic valve stenosis, aortic dissection/rupture (including cases of the Marfan phenotype), atherosclerotic coronary artery disease, ion channelopathies, and arrhythmogenic right ventricular cardiomyopathy. In addition, commotio cordis (i.e., sudden death caused by blunt, nonpenetrating chest blows, associated with structurally normal hearts) is more common as a cause of sudden death in young athletes than many of the aforementioned structural cardiovascular diseases (10).

Regional variations in the causes of sudden death may exist (6-9). Notable among these, arrhythmogenic right ventricular cardiomyopathy has been reported as the most common cause of sudden death in young athletes based on reports from the Veneto region of Italy (8), whereas this disease is a much less frequent cause of sudden death in U.S. athletes (6). In most athletes,

sudden death occurs in the setting of ventricular fibrillation, with the notable exception of aortic dilation that leads to dissection and rupture. For older athletes (>35 years of age), atherosclerotic coronary artery disease is the predominant cause of sudden death (7), but this occurs less frequently in younger participants.

HOW TO USE THE DOCUMENT

Of the 15 Task Forces that make up this document, 9 are disease (or multidisease) related. As before (1-3), specific recommendations for sports eligibility or temporary or permanent disqualification to reduce sudden death risk are formulated around the classification of sports (Task Force 1), which incorporates the principle that training and competition demands may vary considerably among competitive sports (often within sports as well), that the intensity of conditioning may exceed that of competition, and that different levels of physical activity are likely to impact underlying (and unsuspected) cardiovascular diseases unpredictably and in different ways. Furthermore, it is difficult to accurately grade or take into account exercise intensity in various sports because of a variety of factors, particularly motivational attitudes. Finally, as was the practice in prior Bethesda Conferences 16, 26, and 36 (1-3), the panel advises that sports participation recommendations or decisions be based on probable or confirmed diagnostic evidence for cardiovascular disease and not include diagnoses that are ambiguous, possible, or borderline.

The other 6 Task Forces deal with a variety of relevant topics and issues surrounding the risks of the athletic field. These include strategies for preparticipation screening (Task Force 2), use of the automatic external defibrillator on the athletic field (Task Force 12), the impact of dietary supplements and performance-enhancing substances (Task Force 11), commotio cordis as an acknowledged new risk of sudden death during sports (Task Force 13), and medical-legal perspectives (Task Force 15). However, we should underscore that it is not possible to foresee and include in this document every conceivable cardiovascular abnormality or clinical situation relevant to athletes. Eligibility and disqualification decisions in those particular situations would be made on a case-by-case basis with individual clinical judgment. Each of the 15 Task Forces is cited independently as a publication in PubMed.

WHO SHOULD USE THIS DOCUMENT?

These recommendations are designed primarily for cardiologists, internists, pediatricians, family medicine physicians, and other practitioners (including team physicians) charged with decision-making responsibilities

related to the eligibility and disqualification of those competitive athletes with cardiovascular disease.

Although this document essentially focuses on disqualification standards for trained competitive athletes, particularly those in organized sanctioned programs, we also recognize that the principles espoused herein may be, if appropriate, useful when translated to physically active people in other circumstances, for example, in occupations such as police officers, firefighters, and pilots (11), as well as to participants in certain recreational sports activities. In this regard, it should be underscored that many people independently choose to engage in recreational physical activities that may in fact involve high-intensity vigorous training at the same level of some competitive athletes. Therefore, the use of this document for decision making will require certain judgments and extrapolations to account for perceived differences in activity between trained competitive athletes in organized sports and some other physically active people. Hence, it may be possible to selectively apply the principles contained in this document to certain sporting activities that do not meet our precise definition of “competitive.” Nevertheless, excessive and unnecessary exercise restrictions for such people with heart disease could potentially create physical and psychological burdens (particularly in young children) and are discouraged (9).

If the underlying medical considerations are similar to high school- and college-aged athletes, the recommendations in this document could be used to guide decisions relevant to professional athletes with cardiovascular abnormalities. However, professional athletes represent a very small and unique subset of all competitive athletes compared with the millions of student-athlete participants and are generally highly compensated adults with employment contracts (12).

ASSESSMENT OF RISKS

Young people participating in competitive sports with cardiovascular abnormalities have limited control when exposed to extreme and unpredictable environmental conditions (associated with alterations in blood volume, hydration, and electrolytes), as a result of the unwavering demands of sport. These circumstances can enhance the risk for potentially lethal arrhythmias and sudden death, given underlying cardiovascular disease. For many athletes, removal from the lifestyle of athletic training and competition will reduce this risk for sudden death or disease progression, even in the absence of established risk factors related to their disease (13). However, appropriate sports disqualification is only one component of risk reduction, and each of the relevant cardiovascular diseases is attached to its own treatment algorithms, which

can include prophylactic implantation of a cardioverter-defibrillator should sudden death risk be judged unacceptably high (14-16).

The present recommendations, formulated with respect to allowable levels of sports activity, can be regarded as generally conservative. Certainly, this is a prudent posture when the available evidence is limited in many decision-making areas. In this regard, the panel acknowledges that the available data support the principle that participation in high-intensity sports is associated with an increased relative risk of sudden death in the setting of some cardiovascular diseases (6-17). On the other hand, this likelihood cannot be determined with certainty for each patient/athlete, and in fact may be low in certain people. However, at present, additional risk-stratifying tools are not available to independently (and more precisely) guide many of these difficult medical decisions in athletes, particularly for diseases such as hypertrophic cardiomyopathy (18).

Thus, it is possible that the recommendations of this consensus panel (as with the 3 previous Bethesda Conferences) (1-3) will occasionally cause some athletes to be unnecessarily withdrawn from competition. This is, of course, unfortunate, because athletes derive considerable self-assurance, confidence, physical well-being, and even on occasion financial security from these activities. Nevertheless, the increased sudden death risk associated with intense sports is a controllable variable (by disqualification from such sports), and we believe the devastating impact on families and communities of even infrequent sudden death events in this young population underscores the wisdom of our conservative recommendations.

In practice, individual athletes may be encouraged to change their competitive sport involvement from a prohibited high-intensity activity to a more permissible low-intensity one (i.e., usually to class IA). However, the strategy of changing the position in which an athlete competes (e.g., from running back to place kicker in football, or to goalie in hockey or soccer) may be difficult to accomplish in practical terms and therefore should be advised only if the training obligations outside of game situations can be controlled and modified adequately.

RELATION OF AHA/ACC GUIDELINES TO 36TH BETHESDA CONFERENCE

The present AHA/ACC recommendations are intended to update (and are derived from) the prior Bethesda Conferences (1-3). For the most part, the specific recommendations are similar or identical to those in the report on the 36th Bethesda Conference (3). However, selected recommendations of the present AHA/ACC document do in fact deviate from those of the 36th Bethesda

Conference, becoming less restrictive as certain data and observations have emerged since 2005. Nevertheless, numerous “gray areas” persist, for which the assessment of safe versus nonsafe sports participation continues to be uncertain from a medical and scientific perspective, with absolute certainty difficult to achieve for many cardiovascular issues. This may result in differences of opinion among physicians regarding the exercise of clinical judgment in individual cases. Thus, in making certain eligibility or disqualification decisions, some physicians may rely on the more liberal guidelines in portions of the present document, whereas others may take a more conservative approach by adopting the more restrictive recommendations from the 36th Bethesda Conference.

It is also important to underscore that the recommendations in this AHA/ACC document are not intended to establish absolute mandates or the general medical (and legal) standard of care applicable to all cases. These recommendations do not (and cannot) absolutely and arbitrarily replace individual clinical judgment and informed medical reasoning.

The panel recognizes that some practitioners, depending on their perception of risk for specific individual patients, may choose to prudently deviate from the published recommendations in selected clinical situations. Therefore, fully informed athletes with certain conditions may continue to engage in competitive sports in concert with recommendations made by their physician and athletic organization (i.e., high school or college). Individual athletes in the past have taken this option to continue or return to play, and we anticipate this will occur in the future. There will always be tolerance in the system for some flexibility and individual responsibility and choice, after the prevalent uncertainties have been acknowledged.

As with all guidelines, which cannot be regarded as rigid dictum, the specific medical clearance or disqualification recommendation in a particular case is ultimately the responsibility of the managing physician with medically relevant knowledge of the individual athlete-patient. Although neither the 36th Bethesda Conference or the present AHA/ACC recommendations arbitrarily establish the standard of care, these documents nevertheless do provide the framework for good medical practice (19).

It is important to recognize that protection of the athlete's health and avoidance of any unreasonable risks for sudden death during competitive athletics should be considered a priority in exercising individual clinical judgment and making medical recommendations regarding sports participation with a cardiovascular abnormality. The level of importance that the athlete personally attaches to engagement in competitive sports should not be a deciding factor in formulating eligibility recommendations.

Clinicians should also recognize that medical eligibility versus disqualification decisions have become increasingly complex. Also, these decisions may be fraught with potential legal liability risks. Therefore, it is unwise to be unduly influenced by the libertarian (free will) desires of athletes (with an important cardiovascular abnormality) willing to assume medically unreasonable risks to participate in a sport, nor by the managing clinician's personal willingness to comply with the desires of the individual athlete-patient. Finally, it is important to recognize that third-party interests (e.g., on behalf of high schools, colleges, or professional clubs) unavoidably contribute to the complexity in the decision-making process, but these should not outweigh the paramount concern for the athlete's health and safety when making medical eligibility recommendations.

DISCLOSURES

Writing Group Disclosures

Writing Group Member	Employment	Research Grant	Other Research Support	Speakers Bureau/Honoraria	Expert Witness	Ownership Interest	Consultant/Advisory Board	Other
Barry J. Maron	Minneapolis Heart Institute Foundation	None	None	None	None	None	None	None
Douglas P. Zipes	Indiana University	None	None	None	None	None	None	None
Richard J. Kovacs	Indiana University	None	None	None	None	None	None	None

This table represents the relationships of writing group members that may be perceived as actual or reasonably perceived conflicts of interest as reported on the Disclosure Questionnaire, which all members of the writing group are required to complete and submit. A relationship is considered to be “significant” if (a) the person receives \$10,000 or more during any 12-month period, or 5% or more of the person's gross income; or (b) the person owns 5% or more of the voting stock or share of the entity, or owns \$10,000 or more of the fair market value of the entity. A relationship is considered to be “modest” if it is less than “significant” under the preceding definition.

Reviewer Disclosures

Reviewer	Employment	Research Grant	Other Research Support	Speakers Bureau/Honoraria	Expert Witness	Ownership Interest	Consultant/Advisory Board	Other
Robert C. Hendel	University of Miami	None	None	None	None	None	None	None
Adolph M. Hutter, Jr	Massachusetts General Hospital	None	None	None	None	None	None	None
James L. Januzzi, Jr	Massachusetts General Hospital	None	None	None	None	None	None	None
Wojciech Krol	Medical University of Warsaw (Poland)	None	None	None	None	None	None	None
Geetha Raghuvier	Children's Mercy Hospital, Kansas City, MO	None	None	None	None	None	None	None
Barbara S. Wiggins	Medical University of South Carolina	None	None	None	None	None	None	None

This table represents the relationships of reviewers that may be perceived as actual or reasonably perceived conflicts of interest as reported on the Disclosure Questionnaire, which all reviewers are required to complete and submit. A relationship is considered to be "significant" if (a) the person receives \$10,000 or more during any 12-month period, or 5% or more of the person's gross income; or (b) the person owns 5% or more of the voting stock or share of the entity, or owns \$10,000 or more of the fair market value of the entity. A relationship is considered to be "modest" if it is less than "significant" under the preceding definition.

REFERENCES

- 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.
- Maron BJ, Mitchell JH. 26th Bethesda Conference: recommendations for determining eligibility for competition in athletes with cardiovascular abnormalities [published correction appears in *Med Sci Sports Exerc*. 1994;26:followi]. *J Am Coll Cardiol*. 1994;24:845-99.
- Maron BJ, Zipes DP. 36th Bethesda Conference: introduction: eligibility recommendations for competitive athletes with cardiovascular abnormalities. *J Am Coll Cardiol*. 2005;45:1318-21. <http://dx.doi.org/10.1016/j.jacc.2005.02.006>.
- Maron BJ, Friedman RA, Kligfield P, Levine BD, Viskin S, Chaitman BR, Okin PM, Saul JP, Salberg L, Van Hare GF, Soliman EZ, Chen J, Matherne GP, Bolling SF, Mitten MJ, Caplan A, Balady GJ, Thompson PD; on behalf of American Heart Association Council on Clinical Cardiology; Advocacy Coordinating Committee; Council on Cardiovascular Disease in the Young; Council on Cardiovascular Surgery and Anesthesia; Council on Epidemiology and Prevention; Council on Functional Genomics and Translational Biology; Council on Quality of Care and Outcomes Research and American College of Cardiology. Assessment of the 12-lead ECG as a screening test for detection of cardiovascular disease in healthy general populations of young people (12-25 years of age): a scientific statement from the American Heart Association and the American College of Cardiology. *J Am Coll Cardiol*. 2014;64:1479-514. <http://dx.doi.org/10.1016/j.jacc.2014.05.006>.
- Maron BJ. Sudden death in young athletes. *N Engl J Med*. 2003;349:1064-75. <http://dx.doi.org/10.1056/NEJMra022783>.
- Maron BJ, Doerer JJ, Haas TS, Tierney DM, Mueller FO. Sudden deaths in young competitive athletes: analysis of 1866 deaths in the United States, 1980-2006. *Circulation*. 2009;119:1085-92. <http://dx.doi.org/10.1161/CIRCULATIONAHA.108.804617>.
- 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.
- Corrado D, Basso C, Pavei A, Michieli P, Schiavon M, Thiene G. Trends in sudden cardiovascular death in young competitive athletes after implementation of a preparticipation screening program. *JAMA*. 2006;296:1593-601. <http://dx.doi.org/10.1001/jama.296.13.1593>.
- Maron BJ, Chaitman BR, Ackerman MJ, Bayés de Luna A, Corrado D, Crosson JE, Deal BJ, Driscoll DJ, Estes NA 3rd, Araújo CG, Liang DH, Mitten MJ, Myerburg RJ, Pelliccia A, Thompson PD, Towbin JA, Van Camp SP; for the Working Groups of the American Heart Association Committee on Exercise, Cardiac Rehabilitation, and Prevention; Councils on Clinical Cardiology and Cardiovascular Disease in the Young. Recommendations for physical activity and recreational sports participation for young patients with genetic cardiovascular diseases. *Circulation*. 2004;109:2807-16. <http://dx.doi.org/10.1161/01.CIR.0000128363.85581.E1>.
- Maron BJ, Estes NA 3rd. Commotio cordis. *N Engl J Med*. 2010;362:917-27. <http://dx.doi.org/10.1056/NEJMra0910111>.
- Maron BJ, Barry JA, Poole RS. Pilots, hypertrophic cardiomyopathy, and issues of aviation and public safety. *Am J Cardiol*. 2004;93:441-4. <http://dx.doi.org/10.1016/j.amjcard.2003.10.038>.
- Harris KM, Sponsel A, Hutter AM Jr., Maron BJ. Brief communication: cardiovascular screening practices of major North American professional sports teams. *Ann Intern Med*. 2006;145:507-11.
- Maron BJ. Contemporary insights and strategies for risk stratification and prevention of sudden death in hypertrophic cardiomyopathy [published correction appears in *Circulation*. 2010;122:e7]. *Circulation*. 2010;121:445-56. <http://dx.doi.org/10.1161/CIRCULATIONAHA.109.878579>.
- Maron BJ, Spirito P, Ackerman MJ, Casey SA, Semsarian C, Estes NA 3rd, Shannon KM, Ashley EA, Day SM, Pacileo G, Formisano F, Devoto E, Anastakis A, Bos JM, Woo A, Autore C, Pass RH, Boriani G, Garberich RF, Almquist AK, Russell MW, Boni L, Berger S, Maron MS, Link MS. Prevention of sudden cardiac death with implantable cardioverter-defibrillators in children and adolescents with hypertrophic cardiomyopathy. *J Am Coll Cardiol*. 2013;61:1527-35. <http://dx.doi.org/10.1016/j.jacc.2013.01.037>.
- Maron BJ, Spirito P, Shen WK, Haas TS, Formisano F, Link MS, Epstein AE, Almquist AK, Daubert JP, Lawrenz T, Boriani G, Estes NA 3rd, Favale S, Piccinino M, Winters SL, Santini M, Betocchi S, Arribas F, Sherrid MV, Buja G, Semsarian C, Bruzzi P. Implantable cardioverter-defibrillators and prevention of sudden cardiac death in hypertrophic cardiomyopathy [published correction appears in *JAMA*. 2007;298:1516]. *JAMA*. 2007;298:405-12. <http://dx.doi.org/10.1001/jama.298.4.405>.
- Bhonsale A, James CA, Tichnell C, Murray B, Gagarin D, Philips B, Dalal D, Tedford R, Russell SD, Abraham T, Tandri H, Judge DP, Calkins H. Incidence and predictors of implantable cardioverter-defibrillator therapy in patients with arrhythmic right ventricular dysplasia/cardiomyopathy undergoing implantable cardioverter-defibrillator implantation for primary prevention. *J Am Coll Cardiol*. 2011;58:1485-96. <http://dx.doi.org/10.1016/j.jacc.2011.06.043>.
- 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.
- Maron BJ, Ommen SR, Semsarian C, Spirito P, Olivetto I, Maron MS. Hypertrophic cardiomyopathy: present and future, with translation into contemporary cardiovascular medicine [published correction appears in *J Am Coll Cardiol*. 2014;64:1188]. *J Am Coll Cardiol*. 2014;64:83-99. <http://dx.doi.org/10.1016/j.jacc.2014.05.003>.
- 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. <http://dx.doi.org/10.1056/NEJM199811263392211>.

KEY WORDS ACC/AHA Scientific Statements, athletes, cardiovascular abnormalities, genetics, hypertrophic cardiomyopathy, sudden death

APPENDIX

Task Forces and Authors

Preamble, Principles, and General Considerations	Barry J. Maron, MD, FACC, Co-Chair; Douglas P. Zipes, MD, FAHA, MACC, Co-Chair; Richard J. Kovacs, MD, FAHA, FACC, Co-Chair
Task Force 1: Classification of Sport: Dynamic, Static and Impact	Benjamin D. Levine, MD, FAHA, FACC, Chair; Aaron L. Baggish, MD, FACC; Richard J. Kovacs, MD, FAHA, FACC; Mark S. Link, MD, FACC; Martin S. Maron, MD, FACC; Jere H. Mitchell, MD, FACC
Task Force 2: Preparticipation Screening for Cardiovascular Disease in Competitive Athletes	Barry J. Maron, MD, FACC, Chair; Benjamin D. Levine, MD, FAHA, FACC; Reginald L. Washington, MD, FAHA; Aaron L. Baggish, MD, FACC; Richard J. Kovacs, MD, FAHA, FACC; Martin S. Maron, MD, FACC
Task Force 3: Hypertrophic Cardiomyopathy, Arrhythmogenic Right Ventricular Cardiomyopathy and Other Cardiomyopathies, and Myocarditis	Barry J. Maron, MD, FACC, Chair; James E. Udelson, MD, FAHA, FACC; Robert O. Bonow, MD, MS, FAHA, MACC; Rick Nishimura, MD, FAHA, MACC; Michael J. Ackerman, MD, PhD, FACC; N.A. Mark Estes III, MD, FACC; Leslie T. Cooper, Jr, MD, FAHA, FACC; Mark S. Link, MD, FACC; Martin S. Maron, MD, FACC
Task Force 4: Congenital Heart Disease	George F. Van Hare, MD, FACC, Chair; Michael J. Ackerman, MD, PhD, FACC; Juli-anne K. Evangelista, DNP, APRN, CPNP-AC, FACC; Richard J. Kovacs, MD, FAHA, FACC; Robert J. Myerburg, MD, FACC; Keri M. Shafer, MD; Carole A. Warnes, MD, FACC; Reginald L. Washington, MD, FAHA
Task Force 5: Valvular Heart Disease	Robert O. Bonow, MD, MS, FAHA, MACC, Chair; Rick Nishimura, MD, FAHA, MACC; Paul D. Thompson, MD, FAHA, FACC; James E. Udelson, MD, FAHA, FACC
Task Force 6: Hypertension	Henry R. Black, MD, FAHA, Chair; Domenic Sica, MD; Keith Ferdinand, MD, FAHA, FACC; William B. White, MD
Task Force 7: Aortic Diseases, Including Marfan Syndrome	Alan C. Braverman, MD, FACC, Chair; Kevin M. Harris, MD, FACC; Richard J. Kovacs, MD, FAHA, FACC; Barry J. Maron, MD, FACC
Task Force 8: Coronary Artery Disease	Paul D. Thompson, MD, FAHA, FACC, Chair; Robert J. Myerburg, MD, FACC; Benjamin D. Levine, MD, FAHA, FACC; James E. Udelson, MD, FAHA, FACC; Richard J. Kovacs, MD, FAHA, FACC
Task Force 9: Arrhythmias and Conduction Defects	Douglas P. Zipes, MD, FAHA, MACC, Chair; Mark S. Link, MD, FACC; Michael J. Ackerman, MD, PhD, FACC; Richard J. Kovacs, MD, FAHA, FACC; Robert J. Myerburg, MD, FACC; N.A. Mark Estes III, MD, FACC
Task Force 10: The Cardiac Channelopathies	Michael J. Ackerman, MD, PhD, FACC, Chair; Douglas P. Zipes, MD, FAHA, MACC; Richard J. Kovacs, MD, FAHA, FACC; Barry J. Maron, MD, FACC
Task Force 11: Drugs and Performance Enhancing Substances	N.A. Mark Estes III, MD, FACC, Chair; Richard J. Kovacs, MD, FAHA, FACC; Aaron L. Baggish, MD, FACC; Robert J. Myerburg, MD, FACC
Task Force 12: Emergency Action Plans, Resuscitation, CPR, and AEDs	Mark S. Link, MD, FACC, Chair; Robert J. Myerburg, MD, FACC; N.A. Mark Estes III, MD, FACC
Task Force 13: Commotio Cordis	Mark S. Link, MD, FACC, Chair; N.A. Mark Estes III, MD, FACC; Barry J. Maron, MD, FACC
Task Force 14: Sickle Cell Trait	Barry J. Maron, MD, FACC, Chair; Kevin M. Harris, MD, FACC; Paul D. Thompson, MD, FAHA, FACC; E. Randy Eichner, MD; Martin H. Steinberg, MD
Task Force 15: Legal Aspects of Medical Eligibility and Disqualification Recommendations	Matthew J. Mitten, JD, Chair; Douglas P. Zipes, MD, FAHA, MACC; Barry J. Maron, MD, FACC; William J. Bryant, JD

AHA/ACC SCIENTIFIC STATEMENT

Eligibility and Disqualification Recommendations for Competitive Athletes With Cardiovascular Abnormalities: Task Force 1: Classification of Sports: Dynamic, Static, and Impact

A Scientific Statement From the American Heart Association and American College of Cardiology

Benjamin D. Levine, MD, FAHA,
FACC, *Chair**

Aaron L. Baggish, MD, FACC*

Richard J. Kovacs, MD, FAHA, FACC*

Mark S. Link, MD, FACC*

Martin S. Maron, MD, FACC*
Jere H. Mitchell, MD, FACC*

The “classification of sports” section has been a part of each iteration of the recommendations for participation in sports and provides a framework by which athletes with heart disease can be prescribed or proscribed specific sports for participation (1-3). For the 36th Bethesda Conference, an earlier version of the **Figure** was constructed that characterized sports by their strength component, expressed as the relative intensity of static muscle contractions (percentage of a maximal voluntary contraction), and their

endurance component, reflected by the relative intensity of dynamic exercise (regular contraction of large muscle groups) or percentage of maximal aerobic power ($\dot{V}O_{2\max}$) (3). The rationale for a classification scheme applicable to the competitive athlete with cardiac disease is based on the well-described hemodynamics of each different type of exercise (static versus dynamic) (3,4), as well as the apparent cardiac adaptation of athletes who compete in these sports (5), which reflects the chronic load on the

*On behalf of the American Heart Association Electrocardiography and Arrhythmias Committee of the Council on Clinical Cardiology, Council on Cardiovascular Disease in the Young, Council on Cardiovascular and Stroke Nursing, Council on Functional Genomics and Translational Biology, and the American College of Cardiology.

[illegible]

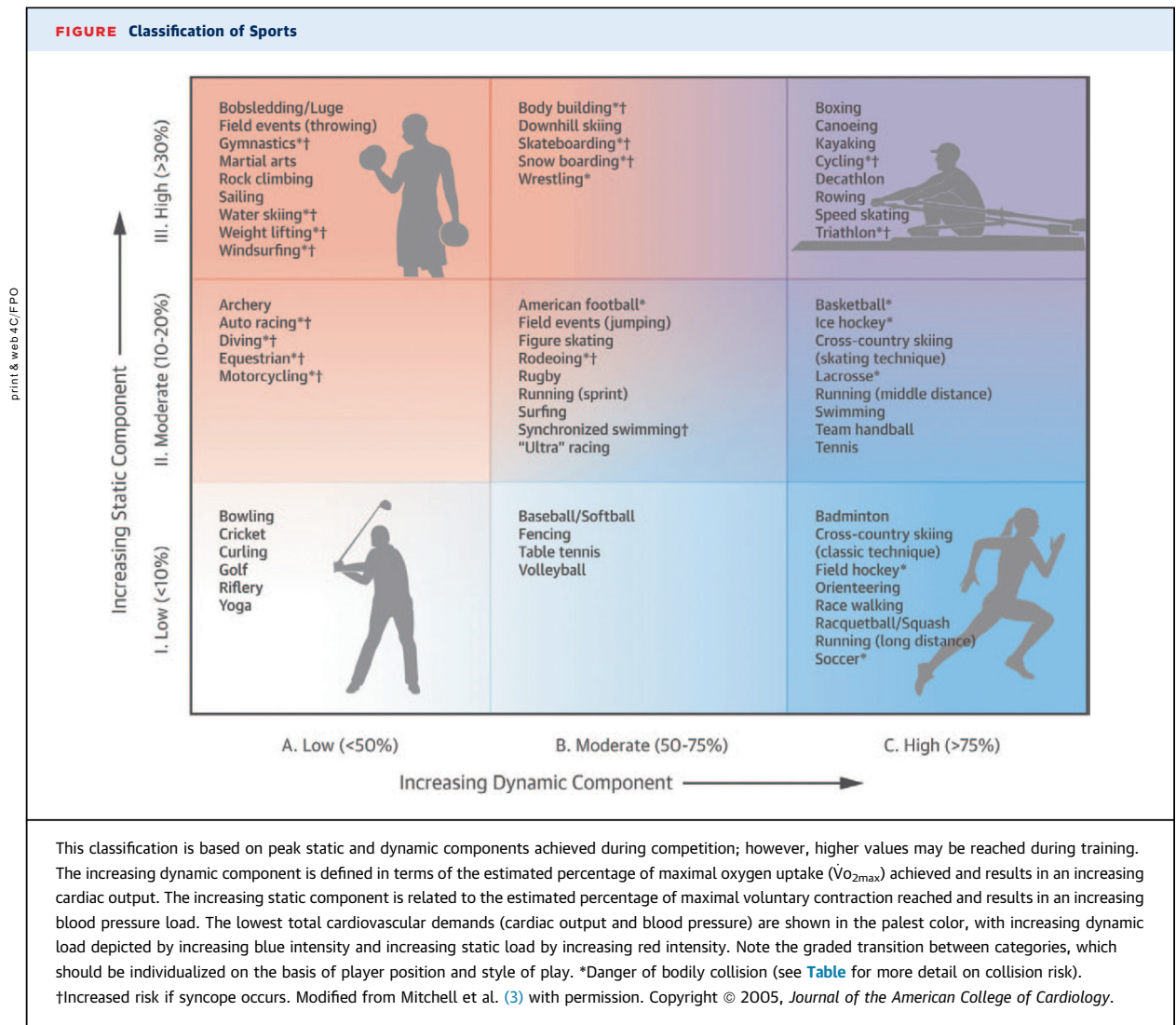
This statement was approved by the American Heart Association Science Advisory and Coordinating Committee on June 24, 2015, and the American Heart Association Executive Committee on July 22, 2015, and by the American College of Cardiology Board of Trustees and Executive Committee on June 3, 2015.

The American College of Cardiology requests that this document be cited as follows: Levine BD; Baggett AL, Kovacs RJ, Link MS, Maron MS, Mitchell JH; on behalf of the American Heart Association Electrocardiography and Arrhythmias Committee of the Council on Clinical Cardiology, Council on Cardiovascular Disease in the Young, Council on Cardiovascular and Stroke Nursing, Council on Functional Genomics and Translational Biology, and the American College of Cardiology. Eligibility and disqualification recommendations for competitive athletes with cardiovascular abnormalities: Task Force 1: classification of sports: dynamic, static, and impact: a scientific statement from the American Heart Association and American College of Cardiology. *J Am Coll Cardiol* 2015;xx:xxx-xxxx.

This article has been copublished in *Circulation*.

Copies: This document is available on the World Wide Web sites of the American Heart Association (<http://my.americanheart.org>) and the American College of Cardiology (www.acc.org). For copies of this document, please contact Elsevier Inc. Reprint Department via fax (212-633-3820) or e-mail (reprints@elsevier.com).

Permissions: Multiple copies, modification, alteration, enhancement, and/or distribution of this document are not permitted without the express permission of the American College of Cardiology. Requests may be completed online via the Elsevier site (<http://www.elsevier.com/about/policies/author-agreement/obtaining-permission>).



cardiovascular system. The underlying principle is that specific cardiovascular conditions may be more or less susceptible to complications (primarily ischemia, heart failure, or vascular compromise) based on unique characteristics of each lesion and the load placed on the heart during athletic competition.

Static contractions stimulate mechanical and metabolic afferents in skeletal muscle, which leads to large, sustained changes in blood pressure via the exercise pressor reflex (6-8). The larger the muscle mass involved, the greater the intensity of contraction, and the greater the rise in blood pressure (9); incorporation of a Valsalva maneuver during contractions will acutely and transiently increase transmural arterial pressure markedly in blood vessels outside of the chest, although left ventricular (LV) afterload does not appear to increase (10) because of a balanced rise in intracardiac and

intrathoracic pressure inside the chest. Dynamic exercise increases the demand for blood flow and cardiac output in proportion to the metabolic demand ($\dot{V}O_2$): for every 1 L/min increase in oxygen uptake, there is an obligate requirement for a 5 to 6 L/min increase in cardiac output (4,11) as a function of the Fick equation. This increase is independent of age, sex, or fitness (4,12,13).

Both dynamic and static exercise result in an increase in myocardial oxygen demand: heart rate, wall tension (before and after the contraction, which determines preload and afterload), and contractile state of the LV (14). During 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) (15) and a decrease in end-systolic volume (increased contractile state); for athletes, the most important factor is the increase in

end-diastolic volume (16). In high-intensity static exercise, a smaller increase occurs in heart rate, and little change occurs in end-diastolic and end-systolic volumes of the LV; however, arterial pressure and contractile state of the ventricle are increased. Thus, dynamic exercise primarily causes a volume load on the LV, whereas static exercise causes a pressure load. Virtually all sports require a combination of both types of effort, although when both are high, such as in rowing sports, the rise in blood pressure may be dramatic (17), and the cardiac adaptation is among the most prominent of all sports (18).

CLASSIFICATION OF SPORTS

On the basis of these considerations, the following matrix was developed (Figure). This Figure has been modified only slightly from the initial derivation published in the 36th Bethesda Conference, mostly to emphasize a more graded increase in effort/cardiovascular load between categories as opposed to a hard, discrete distinction.

Each sport is categorized 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 a significant risk because of 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 as IIC (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 would preclude a sport that produces a high pressure load on the LV may be advised to avoid sports classified as IIIA, IIIB, and IIC. It should be emphasized that in terms of the classification of sports matrix presented in the Figure, 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 appropriate (low static/high dynamic), then so are classes IA and IB (low static/low and moderate dynamic). Sports in each category are listed in alphabetical order to make them easier to find.

Although this scheme has been very useful in guiding practitioners and allowing recommendations for sports participation, there are a number of key limitations that must be acknowledged to use this approach to guide recommendations for individual athletes:

- The scheme as described is simplistic and is only a rough guide. It must be acknowledged that within each

sport, different position players may have quite different cardiovascular loads, for example, wide receiver or offensive lineman in American football, goalie versus midfielders or forwards in soccer, 50 m versus 400 m distances in swimming, and short-track versus long track speed skating. This differential load may even be manifest at the lowest-intensity sports such as yoga, which also can be practiced at much higher intensities. Therefore, practitioners should be prepared to individualize the classification scheme based on individual athletes and how they play their specific sport and position.

- Even within individual sports, the cardiovascular load may be quite different at different times during the competition. As such, it is recommended that the highest level achieved during competition be used for exercise prescription, even if this level is achieved relatively infrequently.
- The types and intensities of exercise required for training may be different from those achieved during a competition. Therefore, cardiovascular loads experienced during training, including high-intensity interval efforts, and during a game must be considered.
- These guidelines are intended for competitive sports and their required training regimen but may not apply to participation in sports at a recreational level. Moreover, many higher-class activities (such as cycling and running) can be performed by patients with cardiovascular disease after they have received counseling about intensity restriction and competition avoidance as part of healthy secondary prevention.
- Environmental conditions may alter the cardiovascular load for a given sport substantially. Increasing altitude alters oxygen availability and acutely increases the heart rate and cardiac output for any given absolute work rate (19). In patients with underlying coronary heart disease, it may also reduce the myocardial workload required to cause ischemia (20) and increase the risk of sudden death (21), although even short-term acclimatization appears to reduce this risk significantly (21). Heat is also a substantial stressor; because humans thermoregulate by sending blood to the skin, a large extra amount of cardiac output is required to maintain body temperature (22), and this could increase the dynamic classification of some sports (especially “hot yoga”). For patients with limited capability to augment cardiac output, thermal stress may be particularly problematic (23). The psychological and emotional demands of sports, particularly during high-stakes competitions, are also relevant and may increase heart rate substantially and unpredictably.

THE EFFECT OF IMPACT AND CONSIDERATIONS FOR ANTICOAGULATION

Athletes with cardiovascular disease who are taking anticoagulant drugs (vitamin K antagonists, direct thrombin or factor Xa inhibitors) must also consider the risk for impact during practice or competition. An impact that occurs while taking anticoagulation medication increases the risk of severe injury, especially for intracranial hemorrhage. Human-human or human-object impacts occur in many sports. Indeed, there are some sports in which impact is a key component of the game, such as American football and ice hockey. Conversely, there are some sports in which impact is extremely unlikely to occur, such as golf or track and field. For other sports, the risk and occurrence of impact are related to the age and competitiveness of the athletes. In these sports, such as basketball and soccer, the older the person and the more competitive the play, the more likely these people will undergo impacts. The [Table](#) divides sports according to the age of the athlete and the relative risk for impact.

Intracranial hemorrhage risk is possibly best ascertained by concussion incidence in sports; however, concussion incidences are certainly an underrepresentation of severe head injuries. Many head injuries do not result in concussion but nevertheless could put the person at a higher risk of intracranial bleeds if the person has been undergoing treatment with an anticoagulant agent. In high school athletes, concussion incidence is highest in American football ($\approx 23/10,000$ exposures), followed by ice hockey, lacrosse, soccer, basketball, and wrestling ([24,25](#)). Concussion risk is much higher in competition than in practice, with most concussions occurring as a result of player-player contact (70% of the concussions) or player-surface contact (17%) ([24,25](#)). Severe injuries not limited to head injury (defined as injuries that resulted in >21 lost days of sports participation) show a similar frequency distribution, with American football being most common ($\approx 20/10,000$ exposures) ([26](#)).

TABLE	Sports According to Risk of Impact and Educational Background	
	Junior High School	High School/College
Impact expected	American football Ice hockey Lacrosse Wrestling Karate/judo Fencing Boxing	American football Soccer Ice hockey Lacrosse Basketball Wrestling Karate/judo Downhill skiing Squash Fencing Boxing
Impact may occur	Soccer Basketball Field hockey Downhill skiing Equestrian Squash Cycling	Field hockey Equestrian Cycling Baseball/softball Gymnastics Figure skating
Impact not expected	Baseball/softball Cricket Golf Riflery Gymnastics Volleyball Swimming Track and field Tennis Figure skating Cross-country skiing Rowing Sailing Archery Weightlifting Badminton	Cricket Golf Riflery Volleyball Swimming Track and field Tennis Cross-country skiing Rowing Sailing Archery Weightlifting Badminton

Recommendations

- 1. The risk of bleeding with athletes receiving vitamin K antagonists or direct thrombin or factor Xa inhibitors is increased in sports in which impacts may occur, and athletes should be cautioned to avoid these sports (Class IIb; Level of Evidence C).**
- 2. Athletes taking vitamin K antagonists or direct thrombin or factor Xa inhibitors should not participate in sports with impact expected, because the risk of intracranial hemorrhage is increased (Class III; Level of Evidence C).**

DISCLOSURES

Writing Group Disclosures

Writing Group Member	Employment	Research Grant	Other Research Support	Speakers Bureau/Honoraria	Expert Witness	Ownership Interest	Consultant/Advisory Board	Other
Benjamin D. Levine	University of Texas Southwestern Medical Center; Texas Health Presbyterian Hospital Dallas Institute for Exercise and Environmental Medicine	None	None	None	None	None	None	None
Aaron L. Baggish	Massachusetts General Hospital	None	None	None	None	None	None	None
Richard J. Kovacs	Indiana University	None	None	None	None	None	None	None
Mark S. Link	Tufts University	None	None	None	None	None	None	None
Martin S. Maron	Tufts Medical Center	None	None	None	None	None	None	None
Jere H. Mitchell	University of Texas Southwestern Medical Center	None	None	None	None	None	None	None

This table represents the relationships of writing group members that may be perceived as actual or reasonably perceived conflicts of interest as reported on the Disclosure Questionnaire, which all members of the writing group are required to complete and submit. A relationship is considered to be "significant" if (a) the person receives \$10,000 or more during any 12-month period, or 5% or more of the person's gross income; or (b) the person owns 5% or more of the voting stock or share of the entity, or owns \$10,000 or more of the fair market value of the entity. A relationship is considered to be "modest" if it is less than "significant" under the preceding definition.

Reviewer Disclosures

Reviewer	Employment	Research Grant	Other Research Support	Speakers Bureau/Honoraria	Expert Witness	Ownership Interest	Consultant/Advisory Board	Other
Michael S. Emery	Greenville Health System	None	None	None	None	None	None	None
Michael J. Joyner	Mayo Clinic and Foundation, Rochester	None	None	None	None	None	None	None
Matthew V. Park	NorthWest Children's Heart Care, Pediatrix Medical Group	None	None	None	None	None	None	None

This table represents the relationships of reviewers that may be perceived as actual or reasonably perceived conflicts of interest as reported on the Disclosure Questionnaire, which all reviewers are required to complete and submit. A relationship is considered to be "significant" if (a) the person receives \$10,000 or more during any 12-month period, or 5% or more of the person's gross income; or (b) the person owns 5% or more of the voting stock or share of the entity, or owns \$10,000 or more of the fair market value of the entity. A relationship is considered to be "modest" if it is less than "significant" under the preceding definition.

REFERENCES

- 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.
- Mitchell JH, Haskell WL, Raven PB. Classification of sports. *J Am Coll Cardiol*. 1994;24:864-6.
- Mitchell JH, Haskell W, Snell P, Van Camp SP. Task Force 8: classification of sports. *J Am Coll Cardiol*. 2005;45:1364-7. <http://dx.doi.org/10.1016/j.jacc.2005.02.015>.
- Levine BD. Exercise physiology for the clinician. In: Thompson PD, editor. *Exercise and Sports Cardiology*. New York, NY: McGraw Hill, 2000:3-28.
- Baggish AL, Wood MJ. Athlete's heart and cardiovascular care of the athlete: scientific and clinical update. *Circulation*. 2011;123:2723-35. <http://dx.doi.org/10.1161/CIRCULATIONAHA.110.981571>.
- Mitchell JH. J.B. Wolfe memorial lecture: neural control of the circulation during exercise. *Med Sci Sports Exerc*. 1990;22:141-54.
- Mitchell JH, Victor RG. Neural control of the cardiovascular system: insights from muscle sympathetic nerve recordings in humans. *Med Sci Sports Exerc*. 1996;28 suppl:S60-9.
- Raven PB. Neural control of the circulation during exercise themed issue. *Exp Physiol*. 2006;91:25-6.
- Mitchell JH, Schibye B, Payne FC 3rd, Saltin B. Response of arterial blood pressure to static exercise in relation to muscle mass, force development, and electromyographic activity. *Circ Res*. 1981;48 pt 2: 170-5.
- Haykowsky M, Taylor D, Teo K, Quinney A, Humen D. Left ventricular wall stress during leg-press exercise performed with a brief Valsalva maneuver. *Chest*. 2001;119:150-4.
- Rowell LB. Neural control of muscle blood flow: importance during dynamic exercise. *Clin Exp Pharmacol Physiol*. 1997;24:117-25.
- Fu Q, Levine BD. Cardiovascular response to exercise in women. *Med Sci Sports Exerc*. 2005;37: 1433-5.
- McGuire DK, Levine BD, Williamson JW, Snell PG, Blomqvist CG, Saltin B, Mitchell JH. A 30-year follow-up of the Dallas Bedrest and Training Study, I: effect of age on the cardiovascular response to exercise. *Circulation*. 2001;104:1350-7.
- Mitchell JH, Hefner LL, Monroe RG. Performance of the left ventricle. *Am J Med*. 1972;53:481-94.
- Levine BD, Lane LD, Buckley JC, Friedman DB, Blomqvist CG. Left ventricular pressure-volume and Frank-Starling relations in endurance athletes: implications for orthostatic tolerance and exercise performance. *Circulation*. 1991;84:1016-23.
- Levine BD. $\dot{V}_{O_{2max}}$: what do we know, and what do we still need to know? *J Physiol*. 2008;586:25-34. <http://dx.doi.org/10.1113/jphysiol.2007.147629>.
- Clifford PS, Hanel B, Secher NH. Arterial blood pressure response to rowing. *Med Sci Sports Exerc*. 1994;26:715-9.
- 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. <http://dx.doi.org/10.1056/NEJM199101313240504>.
- Bärtsch P, Saltin B, Dvorak J, Federation Internationale de Football Association. Consensus statement on playing football at different altitude. *Scand J*

Med Sci Sports. 2008;18 suppl 1:96-9. <http://dx.doi.org/10.1111/j.1600-0838.2008.00837.x>.

20. Levine BD, Zuckerman JH, deFilippi CR. Effect of high-altitude exposure in the elderly: the Tenth Mountain Division study. *Circulation*. 1997;96:1224-32.

21. Lo MY, Daniels JD, Levine BD, Burtcher M. Sleeping altitude and sudden cardiac death. *Am Heart J*. 2013;166:71-5. <http://dx.doi.org/10.1016/j.ahj.2013.04.003>.

22. Crandall CG, González-Alonso J. Cardiovascular function in the heat-stressed human. *Acta Physiol (Oxf)*. 2010;199:407-23. <http://dx.doi.org/10.1111/j.1748-1716.2010.02119.x>.

23. Arbab-Zadeh A, Crandall CG, Levine BD. Thermo-regulation in patients with cardiac disease. *J Cardiopulm Rehabil*. 2002;22:38-9.

24. Marar M, McIlvain NM, Fields SK, Comstock RD. Epidemiology of concussions among United States high school athletes in 20 sports. *Am J Sports Med*. 2012;40:747-55. <http://dx.doi.org/10.1177/0363546511435626>.

25. Harmon KG, Drezner J, Gammons M, Guskiewicz K, Halstead M, Herring S, Kutcher J, Pana A, Putukian M, Roberts W, American Medical Society for Sports Medicine. American Medical Society for Sports Medicine position statement: concussion in sport. *Clin J Sport*

Med. 2013;23:1-18. <http://dx.doi.org/10.1097/JSM.0b013e31827f5f93>.

26. Darrow CJ, Collins CL, Yard EE, Comstock RD. Epidemiology of severe injuries among United States high school athletes: 2005-2007. *Am J Sports Med*. 2009;37:1798-805. <http://dx.doi.org/10.1177/0363546509333015>.

KEY WORDS ACC/AHA Scientific Statements, athletes, blood pressure, cardiac output, cardiovascular abnormalities, classification, oxygen saturation, sport

AHA/ACC SCIENTIFIC STATEMENT

Eligibility and Disqualification Recommendations for Competitive Athletes With Cardiovascular Abnormalities: Task Force 2: Preparticipation Screening for Cardiovascular Disease in Competitive Athletes

A Scientific Statement From the American Heart Association and American College of Cardiology

Barry J. Maron, MD, FACC, *Chair**

Benjamin D. Levine, MD, FAHA, FACC*

Richard J. Kovacs, MD, FAHA, FACC*

Reginald L. Washington, MD, FAHA*

Martin S. Maron, MD, FACC*

Aaron L. Baggish, MD, FACC*

The central purpose of preparticipation screening of trained competitive athletes is to identify or raise suspicion of those cardiovascular abnormalities and diseases that are potentially responsible for sudden unexpected death on the athletic field (1-14). When such athletes are recognized, they are exposed to eligibility and disqualification decisions that become the responsibility of the practicing physician

(4,15-17) and are a subject of this document. There is general (although not universal) (12) agreement with the principle that screening to detect important diseases and potentially prevent sudden death is justified and potentially beneficial (1-3,5-9,18).

There are many pathways and strategies by which competitive athletes with cardiovascular disease may be recognized: 1) comprehensive evaluation by a

*On behalf of the American Heart Association Electrocardiography and Arrhythmias Committee of the Council on Clinical Cardiology, Council on Cardiovascular Disease in the Young, Council on Cardiovascular and Stroke Nursing, Council on Functional Genomics and Translational Biology, and the American College of Cardiology.

The American Heart Association and the American College of Cardiology make every effort to avoid any actual or potential conflicts of interest that may arise as a result of an outside relationship or a personal, professional, or business interest of a member of the writing panel. Specifically, all members of the writing group are required to complete and submit a Disclosure Questionnaire showing all such relationships that might be perceived as real or potential conflicts of interest. The Preamble and other Task Force reports for these proceedings are available online at www.onlinejacc.org (J Am Coll Cardiol 2015;xx:000-000; 000-000; 000-000; 000-000; 000-000; 000-000; 000-000; 000-000; 000-000; 000-000; 000-000; 000-000; 000-000; 000-000; 000-000; 000-000; 000-000; 000-000; 000-000).

This statement was approved by the American Heart Association Science Advisory and Coordinating Committee on June 24, 2015, and the American Heart Association Executive Committee on July 22, 2015, and by the American College of Cardiology Board of Trustees and Executive Committee on June 3, 2015.

The American College of Cardiology requests that this document be cited as follows: Maron BJ, Levine BD, Washington RL, Baggish AL, Kovacs RJ, Maron MS; on behalf of the American Heart Association Electrocardiography and Arrhythmias Committee of the Council on Clinical Cardiology, Council on Cardiovascular Disease in the Young, Council on Cardiovascular and Stroke Nursing, Council on Functional Genomics and Translational Biology, and the American College of Cardiology. Eligibility and disqualification recommendations for competitive athletes with cardiovascular abnormalities: Task Force 2: preparticipation screening for cardiovascular disease in competitive athletes: a scientific statement from the American Heart Association and American College of Cardiology. J Am Coll Cardiol 2015;xx:●●●-●●●.

This article has been copublished in *Circulation*.

Copies: This document is available on the World Wide Web sites of the American Heart Association (<http://my.americanheart.org>) and the American College of Cardiology (www.acc.org). For copies of this document, please contact Elsevier Inc. Reprint Department via fax (212-633-3820) or e-mail (reprints@elsevier.com).

Permissions: Multiple copies, modification, alteration, enhancement, and/or distribution of this document are not permitted without the express permission of the American College of Cardiology. Requests may be completed online via the Elsevier site (<http://www.elsevier.com/about/policies/author-agreement/obtaining-permission>).

primary care physician; 2) systematic screening of families with known genetic diseases after diagnosis in a relative; 3) incidental and fortuitous findings on clinical examination or imaging, detected during evaluation for another medical problem; 4) systematic screening of large populations, such as high school and college-aged athletes, for the purpose of determining eligibility for competitive sports, with or without diagnostic testing; and 5) symptoms associated or unassociated with sports. It is likely that a large number (or even most) athletes with cardiovascular disease come to clinical attention based on the circumstances described in items 1 through 3, rather than with formal preparticipation screening.

GENERAL CONSIDERATIONS

Currently, broad-based cardiovascular screening is practiced systematically in athletes at all levels of performance (not confined to the elite) in only 3 countries: in the United States, with personal/family history and physical examination (but without ECGs) (1-3,19,20), and in both Italy (4-6,9) and Israel (7), with 12-lead ECGs in addition to history and physical examination. In many European countries, screening of athletes is largely limited to those performing at the elite level (e.g., in international, Olympic, or professional sports) (21). The potential benefit of such initiatives is the identification of a small number of people with potentially lethal genetic or congenital cardiovascular diseases (e.g., hypertrophic cardiomyopathy) so that 1) they may be withdrawn from competitive sports to decrease their personal risk and generally make the athletic field a safer environment, and 2) in the process, some high-risk people may be recognized who may be candidates for disease-modifying medical or surgical intervention, or for prevention of sudden death with implantable defibrillators. In 1973, the Japanese School Health Law mandated cardiovascular screening with modified ECG and history/physical examination for thousands of children in the first, seventh, and tenth grades (22,23). Few disease-related data have emerged from this initiative, although a variety of generally minor cardiovascular abnormalities or arrhythmias (unassociated with underlying organic heart disease) were identified in only 2% to 3% of children (23).

DEBATE AND CONTROVERSY

Within the context of these potential benefits, there has nevertheless been substantial discussion surrounding the most appropriate and efficacious strategy for screening, including national federally sponsored and mandated cardiovascular screening. For example, Italian investigators have intensely promoted screening with a routine 12-lead ECG (as well as history and physical examination) based on a unique >30-year program mandated by Italian law and

supported by sports medicine physicians dedicated full-time to the program (4-6,9). Since 1997, Israel has maintained a similar mandatory ECG-based initiative and national sports law (7). For >50 years, it has been customary practice in the United States to routinely screen high school and college-aged athletes with history and physical examination (but without noninvasive testing) (1-3,19,20). In contrast, Denmark has pointedly rejected systematic screening for cardiovascular disease in both athletes and any other segment of the population as being unjustified given the low event rate (12,13). Other than Japan (22,23), no country has systematically attempted broad-based cardiovascular screening in general healthy populations (not limited to athletes), with or without ECGs.

UNIVERSAL SCREENING: ECGs VERSUS HISTORY AND PHYSICAL EXAMINATION

Preparticipation screening for cardiovascular disease with personal/family history and physical examination has been the customary practice for all high school and college-aged competitive athletes in the United States for decades, independent of their performance level. This process is guided by the 14-point history and physical examination elements proposed by the American Heart Association (AHA) (1). The AHA recommendations acknowledge that athletes and others with underlying (but undiagnosed) cardiovascular abnormalities may well manifest clinical warning signs (e.g., chest pain, excessive exertional dyspnea, or syncope) identifiable by careful and systematic history. Because most diseases responsible for sudden death in the young are genetic/familial, a thorough family history may raise suspicion of the disorder. An organic heart murmur can alert the examining physician to valvular or other abnormalities, including left ventricular outflow tract obstruction.

A controversy persists as to whether an ECG (in addition to history and physical examination) is a superior strategy to history/physical examination alone for detecting potentially lethal cardiovascular disease, particularly when taking into account the important issues of false-negative and false-positive results, as well as cost and resource availability (1). Indeed, studies comparing these 2 strategies have failed to demonstrate a mortality benefit for ECG screening (18).

The debate between those who strongly promote routine ECGs and those opposed to ECGs as a routine screening tool is not fully resolved as yet, although a substantial literature consisting largely of editorials and viewpoint commentaries is accumulating rapidly. Nevertheless, several points are indisputable. First, the 12-lead ECG, although a mainstay of hospital-based cardiovascular practice for decades, is an unproven diagnostic tool for reliable detection of cardiovascular disease in generally

healthy populations (1). Second, outcome data on athlete screening and mortality have been driven primarily by only 1 database, from the Veneto region of Italy (9% of the national population) as part of its long-term screening program (6,9). This ambitious Italian initiative has been shown to be successful in identifying some at-risk athletes with potentially lethal cardiovascular disease (primarily right ventricular cardiomyopathy, which appears to be endemic in this area of Italy), resulting in their mandatory withdrawal from sports. In addition, a sharp decrease in mortality rate over a 30-year period was demonstrated, which these investigators attributed to incorporation of the 12-lead ECG into the screening program in the early 1980s.

Third, the Italian data showing that ECG screening reduces mortality in athletes have yet to be replicated elsewhere, and evidence from the United States (18) and Israel (7) appears to dispute or diminish the value of the ECG in reducing athlete mortality. For example, contemporary mortality rates in US athletes from Minnesota, where screening is limited to history and physical examination, do not differ from those in the Veneto region of Italy, where the ECG is used routinely (18); furthermore, athlete mortality rates in Israel were not different before and after legislation for mandatory ECGs (7). The fact that it has been difficult to consistently show a reduction in athlete mortality directly attributable to routine ECGs is an observation that may be driven by the generally low event rates in competitive athletes with cardiovascular disease (1-3,6,10,11,18,24-26).

RELEVANCE OF SUDDEN DEATH INCIDENCE TO SCREENING

Indeed, the low frequency with which sudden deaths occur in the competitive athlete population negatively impacts the justification for broad-based screening in large populations of young people, as well as the weight that can be afforded to this issue as a public health problem. In this regard, there is now overwhelming evidence that these events are relatively uncommon, albeit exceedingly tragic in each case. Most data place these cardiovascular sudden deaths in the range of approximately 1 in 80,000 to 1 in 200 000 participants per year, much less common in relative terms than motor vehicle accidents (by 5,000-fold), suicide, drugs, homicide, or cancer in the same age group and similar in frequency to that of fatal lightning strikes (1,11,25). In a college (National Collegiate Athletic Association) athlete population, drugs and suicide combined accounted for a similar number of deaths as confirmed cardiac disease (24), although a non-forensic-based analysis reported a higher incidence for sudden death (27).

Notably, the absolute number of sudden deaths attributable to documented cardiovascular disease in

competitive athletes is small in populations for which forensic data are reported. For example, the 33-year US Sudden Death in Athletes Registry has reported a maximum of 75 such deaths in any given year nationally (10), and the Veneto database reports 55 sudden deaths in 26 years, or only ≈ 2 per year (6). In other populations, the average number of confirmed cardiovascular deaths annually is much less, for instance, <1 in Minnesota high school athletes (11) or ≈ 4 in college (National Collegiate Athletic Association) athletes (24). Notably, false-negative screening results are a major concern, in which the system fails to identify the cardiac diseases for which it is in fact established. Indeed, a substantial proportion of athletes ($\approx 30\%$ to 40%) may die suddenly of cardiovascular abnormalities that would not necessarily be reliably detected by screening even with ECGs (1,11,24,25).

UNIVERSAL ECG SCREENING

On 3 occasions (1996, 2007, and 2014), AHA consensus expert panels evaluated and decided not to support mandatory national athlete screening in the United States, particularly with routine use of ECGs (1-3). Indeed, sudden cardiovascular deaths in athletes are rare (albeit tragic) events, insufficient in number to be judged as a major public health problem or to justify a change in national healthcare policy. The most frequently cited obstacles to mandatory national screening of trained athletes are as follows: 1) the large number of athletes to be screened nationally on an annual basis (i.e., ≈ 10 to 12 million); 2) the low incidence of events (1,8,10,11,18,24-26); 3) the substantial number of expected false-negative and false-positive results, in the range of 5% to 20% depending on the specific ECG criteria used (1-3,28-32); 4) cost-efficacy considerations, that is, the extensive resources and expenses required versus few events in absolute numbers; 5) liability issues that unavoidably impact physicians with the sole responsibility to disqualify athletes from competition and enforce that decision; 6) the lack of resources or physicians dedicated to performing examinations and interpreting ECGs, in contrast to the long-standing sports medicine program in Italy (4-6,9); 7) the influence of observer variability, technical considerations, and the impact of ethnicity/race on the interpretation of ECGs, which is particularly important for multicultural athlete populations such as in the United States; 8) the need for repetitive (i.e., annual) ECG screening during adolescence, given the possibility of developing phenotypic evidence of cardiomyopathies during this time period or later (33); 9) the logistical challenges and costs related to second-tier confirmatory screening with imaging and other testing, should primary evaluations raise the suspicion of cardiac disease; and 10) recognition that even with testing, screening cannot be expected to

identify all athletes with important cardiovascular abnormalities, and a significant false-negative rate may occur (34).

NONUNIVERSAL SCREENING FOR ATHLETES

Screening programs on a smaller, nonnational basis have been implemented in some high schools, colleges, and local communities that use ECGs (or echocardiograms) with varying expertise, quality control, and results for identifying important cardiac disease. Consistently, the AHA has not opposed ECG-based screening initiatives (often performed by volunteers) in smaller venues; however, for such screening initiatives, the AHA has prudently advised adequate quality control with due consideration for the prominent limitations of the process (including false-negative and false-positive test results), so that the risks and benefits can be understood and are acceptable to all participants, communities, and organizations (1-3).

There are certain known and anticipated limitations in the use of ECGs in population screening, including but not limited to false-positive and false-negative test results, technical and interpretation issues, “gray zone” ambiguous diagnoses, and cost and logistics involved in arranging second-tier diagnostic testing, all of which promote anxiety, uncertainty, and legal considerations (1,12,25,34).

SCREENING AND RACE

Sudden deaths attributable to cardiovascular disease have been reported in athletes of both sexes and a variety of races, although they are much less common in females (by 1:9) (10,14). Preparticipation screening is warranted with the same frequency and criteria, independent of sex and across racial lines. In particular, although hypertrophic cardiomyopathy unrecognized during life is a frequent cause of sudden death in African-Americans on the athletic field and a major impetus for screening in the black community (1,14,35), there is no evidence to justify different or separate screening strategies based on race. However, it is becoming increasingly apparent that ethnic/racial differences in ECG patterns may significantly impact the definition of normality (30,36-39) and therefore potentially the outcome of the screening process for minorities.

ETHICAL CONSIDERATIONS: WHO SHOULD BE SCREENED?

Unfortunately, often overlooked in the ECG screening debate is the potentially troublesome ethical dilemma created by confining (or proposing to limit) screening for potentially lethal diseases to those who choose engagement in competitive sports, while in the process excluding those who are not athletes. The degree to which people engaged in competitive athletics are at greater risk

(given unsuspected underlying heart disease) is not completely resolved. It is likely that the absolute number of sudden deaths is highest in nonathletes because that segment of the population is much larger in size. The AHA maintains the position (1) that theoretically there is no compelling reason to confine screening for cardiovascular disease to young competitive athletes, and exclude non-athletes.

Recommendations

The guidelines presented here are those of the AHA/American College of Cardiology 2014 initiative (1).

1. It is recommended that the AHA's 14-point screening guidelines and those of other societies, such as the American Academy of Pediatrics' Preparticipation Physical Evaluation, be used by examiners as part of a comprehensive history taking and physical examination to detect or raise suspicion of genetic/congenital cardiovascular abnormalities (*Class I; Level of Evidence C*).
2. It is recommended that standardization of the questionnaire forms used as guides for examiners of high school and college athletes in the United States be pursued (*Class I; Level of Evidence C*).
3. Screening with 12-lead ECGs (or echocardiograms) in association with comprehensive history-taking and physical examination to identify or raise suspicion of genetic/congenital and other cardiovascular abnormalities may be considered in relatively small cohorts of young healthy people 12 to 25 years of age, not necessarily limited to competitive athletes (e.g., in high schools, colleges/universities or local communities). Close physician involvement and sufficient quality control is mandatory. If undertaken, such initiatives should recognize the known and anticipated limitations of the 12-lead ECG as a population screening test, including the expected frequency of false-positive and false-negative test results, as well as the cost required to support these initiatives over time (*Class IIb; Level of Evidence C*).
4. Mandatory and universal mass screening with 12-lead ECGs in large general populations of young healthy people 12 to 25 years of age (including on a national basis in the United States) to identify genetic/congenital and other cardiovascular abnormalities is not recommended for athletes and nonathletes alike (*Class III, no evidence of benefit; Level of Evidence C*).
5. Consideration for large-scale, general population, and universal cardiovascular screening in the age group 12 to 25 years with history taking and physical examination alone is not recommended (including on a national basis in the United States) (*Class III, no evidence of benefit; Level of Evidence C*).

DISCLOSURES

Writing Group Disclosures

Writing Group Member	Employment	Research Grant	Other Research Support	Speakers Bureau/Honoraria	Expert Witness	Ownership Interest	Consultant/Advisory Board	Other
Barry J. Maron	Minneapolis Heart Institute Foundation	None	None	None	None	None	None	None
Aaron L. Baggish	Massachusetts General Hospital, Harvard Medical School	None	None	None	None	None	None	None
Richard J. Kovacs	Indiana University	None	None	None	None	None	None	None
Benjamin D. Levine	University of Texas Southwestern Medical Center	None	None	None	Texas Legislature*	None	None	None
Martin S. Maron	Tufts Medical Center	None	None	None	None	None	None	None
Reginald L. Washington	Rocky Mountain Hospital for Children	None	None	None	None	None	None	None

This table represents the relationships of writing group members that may be perceived as actual or reasonably perceived conflicts of interest as reported on the Disclosure Questionnaire, which all members of the writing group are required to complete and submit. A relationship is considered to be "significant" if (a) the person receives \$10,000 or more during any 12-month period, or 5% or more of the person's gross income; or (b) the person owns 5% or more of the voting stock or share of the entity, or owns \$10,000 or more of the fair market value of the entity. A relationship is considered to be "modest" if it is less than "significant" under the preceding definition.

*Modest.

Reviewer Disclosures

Reviewer	Employment	Research Grant	Other Research Support	Speakers Bureau/Honoraria	Expert Witness	Ownership Interest	Consultant/Advisory Board	Other
Robert M. Campbell	Children's Healthcare of Atlanta	None	None	None	None	None	None	None
Luciana D.N. Janot De Matos	Hospital Israelita Albert Einstein (Brazil)	None	None	None	None	None	None	None
Christine E. Lawless	Self-employed, Sports Cardiology Consultants	Bryan Health Foundation†	None	None	None	None	SCAA*	None

This table represents the relationships of reviewers that may be perceived as actual or reasonably perceived conflicts of interest as reported on the Disclosure Questionnaire, which all reviewers are required to complete and submit. A relationship is considered to be "significant" if (a) the person receives \$10,000 or more during any 12-month period, or 5% or more of the person's gross income; or (b) the person owns 5% or more of the voting stock or share of the entity, or owns \$10,000 or more of the fair market value of the entity. A relationship is considered to be "modest" if it is less than "significant" under the preceding definition.

*Modest.

†Significant.

REFERENCES

- Maron BJ, Friedman RA, Kligfield P, Levine BD, Viskin S, Chaitman BR, Okin P, Saul JP, Salberg L, Van Hare GF, Soliman EZ, Chen J, Matherene P, Bolling SF, Mitten MJ, Caplan A, Balady GJ, Thompson PS, on behalf of the American Heart Association Council on Clinical Cardiology, Advocacy Coordinating Committee, Council on Cardiovascular Disease in the Young, Council on Cardiovascular Surgery and Anesthesia, Council on Epidemiology and Prevention, Council on Functional Genomics and Translational Biology, Council on Quality of Care and Outcomes Research, and American College of Cardiology. Assessment of the 12-lead electrocardiogram as a screening test for detection of cardiovascular disease in healthy general populations of young people (12-25 years of age): a scientific statement from the American Heart Association and the American College of Cardiology. *J Am Coll Cardiol*. 2014;64:1479-514. <http://dx.doi.org/10.1016/j.jacc.2014.05.006>.
- Maron BJ, Thompson PD, Ackerman MJ, Balady G, Berger S, Cohen D, Dimeff R, Douglas PS, Glover DW, Hutter AM Jr., Krauss MD, Maron MS, Mitten MJ, Roberts WO, Puffer JC. Recommendations and considerations related to preparticipation screening for cardiovascular abnormalities in competitive athletes: update 2007: a scientific statement from the American Heart Association, Nutrition, Physical Activity, and Metabolism Council. *Circulation*. 2007;115:1643-55. <http://dx.doi.org/10.1161/CIRCULATIONAHA.107.181423>.
- Maron BJ, Thompson PD, Puffer JC, McGrew CA, Strong WB, Douglas PS, Clark LT, Mitten MJ, Crawford MH, Atkins DL, Driscoll DJ, Epstein AE. 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.
- Pelliccia A, Zipes DP, Maron BJ. 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. *J Am Coll Cardiol*. 2008;52:1990-6. <http://dx.doi.org/10.1016/j.jacc.2008.08.055>.
- Corrado D, Pelliccia A, Bjørnstad HH, Vanhees L, Biffi A, Borjesson M, Panhuyzen-Goedkoop N, Deligiannis A, Solberg E, Dugmore D, Mellwig KP, Assanelli D, Delise P, van-Buuren F, Anastakis A, Heidbuchel H, Hoffmann E, Fagard R, Priori SG, Basso C, Arbustini E, Blomstrom-Lundqvist C, McKenna WJ, Thiene G, Study Group of Sport Cardiology of the Working Group of Cardiac Rehabilitation and Exercise Physiology and the Working Group of Myocardial and Pericardial Diseases of the European Society of Cardiology. Cardiovascular pre-participation screening of young competitive athletes for prevention of sudden death: proposal for a common European protocol. Consensus Statement of the Study Group of Sport Cardiology of the Working Group of Cardiac Rehabilitation and Exercise Physiology and the Working Group of Myocardial and Pericardial Diseases of the European Society of Cardiology. *Eur Heart J*. 2005;26:516-24. <http://dx.doi.org/10.1093/eurheartj/ehi108>.
- Corrado D, Basso C, Pavei A, Michieli P, Schiavon M, Thiene G. Trends in sudden cardiovascular death in young competitive athletes after implementation of a

- preparticipation screening program. *JAMA*. 2006;296:1593-601. <http://dx.doi.org/10.1001/jama.296.13.1593>.
7. Steinvil A, Chundadze T, Zeltser D, Rogowski O, Halkin A, Galily Y, Perluk H, Viskin S. Mandatory electrocardiographic screening of athletes to reduce their risk for sudden death: proven fact or wishful thinking? *J Am Coll Cardiol*. 2011;57:1291-6. <http://dx.doi.org/10.1016/j.jacc.2010.10.037>.
 8. Maron BJ. Sudden death in young athletes. *N Engl J Med*. 2003;349:1064-75. <http://dx.doi.org/10.1056/NEJMr022783>.
 9. Corrado D, Basso C, Schiavon M, Thiene G. Screening for hypertrophic cardiomyopathy in young athletes. *N Engl J Med*. 1998;339:364-9. <http://dx.doi.org/10.1056/NEJM199808063390602>.
 10. Maron BJ, Doerer JJ, Haas TS, Tierney DM, Mueller FO. Sudden deaths in young competitive athletes: analysis of 1866 deaths in the United States, 1980-2006. *Circulation*. 2009;119:1085-92. <http://dx.doi.org/10.1161/CIRCULATIONAHA.108.804617>.
 11. Maron BJ, Haas TS, Ahluwalia A, Rutten-Ramos SC. Incidence of cardiovascular sudden deaths in Minnesota high school athletes. *Heart Rhythm*. 2013;10:374-7. <http://dx.doi.org/10.1016/j.hrthm.2012.11.024>.
 12. Holst AG, Winkel BG, Theilade J, Kristensen IB, Thomsen JL, Ottesen GL, Svendsen JH, Haunsø S, Prescott E, Tfelt-Hansen J. Incidence and etiology of sports-related sudden cardiac death in Denmark: implications for preparticipation screening. *Heart Rhythm*. 2010;7:1365-71. <http://dx.doi.org/10.1016/j.hrthm.2010.05.021>.
 13. Maron BJ. Diversity of views from Europe on national preparticipation screening for competitive athletes. *Heart Rhythm*. 2010;7:1372-3. <http://dx.doi.org/10.1016/j.hrthm.2010.06.001>.
 14. Maron BJ, Carney KP, Lever HM, Lewis JF, Barac I, Casey SA, Sherrid MV. Relationship of race to sudden cardiac death in competitive athletes with hypertrophic cardiomyopathy. *J Am Coll Cardiol*. 2003;41:974-80.
 15. 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.
 16. Maron BJ, Mitchell JH. 26th Bethesda Conference: recommendations for determining eligibility for competition in athletes with cardiovascular abnormalities: January 6-7, 1994 [published correction appears in *Med Sci Sports Exerc*. 1994;26:followup]. *J Am Coll Cardiol*. 1994;24:845-99.
 17. Maron BJ, Zipes DP. 36th Bethesda Conference: eligibility recommendations for competitive athletes with cardiovascular abnormalities. *J Am Coll Cardiol*. 2005;45:1313-75.
 18. Maron BJ, Haas TS, Doerer JJ, Thompson PD, Hodges JS. Comparison of U.S. and Italian experiences with sudden cardiac deaths in young competitive athletes and implications for preparticipation screening strategies. *Am J Cardiol*. 2009;104:276-80. <http://dx.doi.org/10.1016/j.amjcard.2009.03.037>.
 19. Glover DW, Glover DW, Maron BJ. Evolution in the process of screening United States high school student-athletes for cardiovascular disease. *Am J Cardiol*. 2007;100:1709-12. <http://dx.doi.org/10.1016/j.amjcard.2007.07.018>.
 20. Glover DW, Maron BJ. Profile of preparticipation cardiovascular screening for high school athletes. *JAMA*. 1998;279:1817-9.
 21. Corrado D, Schimied C, Basso C, Borjesson M, Schiavon M, Pelliccia A, Vanhees L, Thiene G. Risk of sports: do we need a pre-participation screening for competitive and leisure athletes? *Eur Heart J*. 2011;32:934-44. <http://dx.doi.org/10.1093/eurheartj/ehq482>.
 22. Tasaki H, Hamasaki Y, Ichimaru T. Mass screening for heart disease of school children in Saga city: 7-year follow up study. *Jpn Circ J*. 1987;51:1415-20.
 23. Haneda N, Mori C, Nishio T, Saito M, Kajino Y, Watanabe K, Kijima Y, Yamada K. Heart diseases discovered by mass screening in the schools of Shimane Prefecture over a period of 5 years. *Jpn Circ J*. 1986;50:1325-9.
 24. Maron BJ, Haas TS, Murphy CJ, Ahluwalia A, Rutten-Ramos S. Incidence and causes of sudden death in U.S. college athletes. *J Am Coll Cardiol*. 2014;63:1636-43. <http://dx.doi.org/10.1016/j.jacc.2014.01.041>.
 25. Maron BJ. Counterpoint: mandatory ECG screening of young competitive athletes. *Heart Rhythm*. 2012;9:1646-9. <http://dx.doi.org/10.1016/j.hrthm.2012.03.019>.
 26. Risgaard B, Winkel BG, Jabbari R, Glinge C, Ingemann-Hansen O, Thomsen JL, Ottesen GL, Haunsø S, Holst AG, Tfelt-Hansen J. Sports-related sudden cardiac death in a competitive and a noncompetitive athlete population aged 12 to 49 years: data from an unselected nationwide study in Denmark. *Heart Rhythm*. 2014;11:1673-81. <http://dx.doi.org/10.1016/j.hrthm.2014.05.026>.
 27. Harmon KG, Asif IM, Klossner D, Drezner JA. Incidence of sudden cardiac death in National Collegiate Athletic Association athletes. *Circulation*. 2011;123:1594-600. <http://dx.doi.org/10.1161/CIRCULATIONAHA.110.004622>.
 28. Baggish AL, Hutter AM Jr., Wang F, Yared K, Weiner RB, Kupperman E, Picard MH, Wood MJ. Cardiovascular screening in college athletes with and without electrocardiography: a cross-sectional study. *Ann Intern Med*. 2010;152:269-75. <http://dx.doi.org/10.7326/0003-4819-152-5-201003020-00004>.
 29. Malhotra R, West JJ, Dent J, Luna M, Kramer CM, Mounsey JP, Battle R, Saliba E, Rose B, Mistry D, MacKnight J, DiMarco JP, Mahapatra S. Cost and yield of adding electrocardiography to history and physical in screening Division I intercollegiate athletes: a 5-year experience. *Heart Rhythm*. 2011;8:721-7. <http://dx.doi.org/10.1016/j.hrthm.2010.12.024>.
 30. Magalski A, McCoy M, Zabel M, Magee LM, Goetze J, Main ML, Buntlen L, Reid KJ, Ramza BM. Cardiovascular screening with electrocardiography and echocardiography in collegiate athletes. *Am J Med*. 2011;124:511-8. <http://dx.doi.org/10.1016/j.amjmed.2011.01.009>.
 31. Pelliccia A, Maron BJ, Culasso F, Di Paolo FM, Spataro A, Biffi A, Caselli G, Piovano P. Clinical significance of abnormal electrocardiographic patterns in trained athletes. *Circulation*. 2000;102:278-84.
 32. Weiner RB, Hutter AM, Wang F, Kim JH, Wood MJ, Wang TJ, Picard MH, Baggish AL. Performance of the 2010 European Society of Cardiology criteria for ECG interpretation in athletes. *Heart*. 2011;97:1573-7. <http://dx.doi.org/10.1136/hrt.2011.227330>.
 33. Pelliccia A, Di Paolo FM, Quattrini FM, Basso C, Culasso F, Popoli G, De Luca R, Spataro A, Biffi A, Thiene G, Maron BJ. Outcomes in athletes with marked ECG repolarization abnormalities. *N Engl J Med*. 2008;358:152-61. <http://dx.doi.org/10.1056/NEJMoa060781>.
 34. Rowin EJ, Maron BJ, Appelbaum E, Link MS, Gibson CM, Lesser JR, Haas TS, Udelson JE, Manning WJ, Maron MS. Significance of false negative electrocardiograms in preparticipation screening of athletes for hypertrophic cardiomyopathy. *Am J Cardiol*. 2012;110:1027-32. <http://dx.doi.org/10.1016/j.amjcard.2012.05.035>.
 35. 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 4111 subjects in the CARDIA Study. *Circulation*. 1995;92:785-9.
 36. Papadakis M, Carre F, Kervio G, Rawlins J, Panoulas VF, Chandra N, Basavarajiah S, Carby L, Fonseca T, Sharma S. The prevalence, distribution, and clinical outcomes of electrocardiographic repolarization patterns in male athletes of African/Afro-Caribbean origin. *Eur Heart J*. 2011;32:2304-13. <http://dx.doi.org/10.1093/eurheartj/ehr140>.
 37. Magalski A, Maron BJ, Main ML, McCoy M, Florez A, Reid KJ, Epps HW, Bates J, Browne JE. Relation of race to electrocardiographic patterns in elite American football players. *J Am Coll Cardiol*. 2008;51:2250-5. <http://dx.doi.org/10.1016/j.jacc.2008.01.065>.
 38. Di Paolo FM, Schimied C, Zerguini YA, Junge A, Quattrini F, Culasso F, Dvorak J, Pelliccia A. The athlete's heart in adolescent Africans: an electrocardiographic and echocardiographic study. *J Am Coll Cardiol*. 2012;59:1029-36. <http://dx.doi.org/10.1016/j.jacc.2011.12.008>.
 39. Basavarajiah S, Boraita A, Whyte G, Wilson M, Carby L, Shah A, Sharma S. Ethnic differences in left ventricular remodeling in highly-trained athletes: relevance to differentiating physiologic left ventricular hypertrophy from hypertrophic cardiomyopathy. *J Am Coll Cardiol*. 2008;51:2256-62. <http://dx.doi.org/10.1016/j.jacc.2007.12.061>.

KEY WORDS ACC/AHA Scientific Statements, athletes, cardiovascular abnormalities, competitive athletes, preparticipation screening, sudden death

AHA/ACC SCIENTIFIC STATEMENT

Eligibility and Disqualification Recommendations for Competitive Athletes With Cardiovascular Abnormalities: Task Force 3: Hypertrophic Cardiomyopathy, Arrhythmogenic Right Ventricular Cardiomyopathy and Other Cardiomyopathies, and Myocarditis

A Scientific Statement From the American Heart Association and American College of Cardiology

Barry J. Maron, MD, FACC, *Chair**

James E. Udelson, MD, FAHA, FACC*

Robert O. Bonow, MD, MS,
FAHA, MACC*

Rick A. Nishimura, MD,
FAHA, MACC*

Michael J. Ackerman, MD, PhD*
N.A. Mark Estes III, MD, FACC*

Leslie T. Cooper JR, MD, FAHA,
FACC*

Mark S. Link, MD, FACC*

Martin S. Maron, MD, FACC*

Hypertrophic cardiomyopathy (HCM) (1,2) is a major focus of this document given that it is the single most common cause of sudden death in young competitive athletes in the United States, responsible for at least one-third of these events (3).

HYPERTROPHIC CARDIOMYOPATHY

HCM is the most frequent nontraumatic cause of sudden death in the young (1,2) and a common genetic heart disease, occurring in at least 1 in 500

*On behalf of the American Heart Association Electrocardiography and Arrhythmias Committee of the Council on Clinical Cardiology, Council on Cardiovascular Disease in the Young, Council on Cardiovascular and Stroke Nursing, Council on Functional Genomics and Translational Biology, and the American College of Cardiology.

[illegible]

This statement was approved by the American Heart Association Science Advisory and Coordinating Committee on June 24, 2015, and the American Heart Association Executive Committee on July 22, 2015, and by the American College of Cardiology Board of Trustees and Executive Committee on June 3, 2015.

The American College of Cardiology requests that this document be cited as follows: Maron BJ, Udelson JE, Bonow RO, Nishimura RA.

Ackerman MJ, Estes NAM 3rd, Cooper LT Jr, Link MS, Maron MS; on behalf of the American Heart Association Electrocardiography and Arrhythmias Committee of the Council on Clinical Cardiology, Council on Cardiovascular Disease in the Young, Council on Cardiovascular and Stroke Nursing, Council on Functional Genomics and Translational Biology, and the American College of Cardiology. Eligibility and disqualification recommendations for competitive athletes with cardiovascular abnormalities: Task Force 3: hypertrophic cardiomyopathy, arrhythmogenic right ventricular cardiomyopathy and other cardiomyopathies, and myocarditis: a scientific statement from the American Heart Association and American College of Cardiology. *J Am Coll Cardiol* 2015;xx:●●●-●●●.

This article has been copublished in *Circulation*.

Copies: This document is available on the World Wide Web sites of the American Heart Association (my.americanheart.org) and the American College of Cardiology (www.acc.org). For copies of this document, please contact Elsevier Inc. Reprint Department via fax (212-633-3820) or e-mail (reprints@elsevier.com).

Permissions: Multiple copies, modification, alteration, enhancement, and/or distribution of this document are not permitted without the express permission of the American College of Cardiology. Requests may be completed online via the Elsevier site (<http://www.elsevier.com/about/policies/author-agreement/obtaining-permission>).

people in the general population (4). HCM is a clinically and genetically heterogeneous disease, associated with >1,500 mutations in ≥11 major genes (and a variety of other susceptibility genes with lesser evidence for pathogenicity), encoding proteins of the cardiac sarcomere, adjacent Z disk, and calcium handling (5).

Although HCM is associated with substantial diversity in morphological expression (6), clinical diagnosis usually occurs with recognition of the characteristic disease phenotype, that is, left ventricular (LV) hypertrophy without chamber dilatation in the absence of another cardiac or systemic disease capable of producing the magnitude of hypertrophy evident (1,6). Neither systolic anterior motion of the mitral valve, hyperdynamic LV function, or identification of pathogenic sarcomere mutations is obligatory for the clinical diagnosis of HCM (2). Atrial fibrillation is a common cause of morbidity in HCM, occurring in ≈20% of patients, although usually after 30 years of age (1,2). Notably, the clinical presentation and course are diverse, with unexpected sudden death in the young the most visible disease complication.

SUDDEN DEATH RISK

A major impetus in HCM has been the identification of those patients at increased risk for sudden death. Indeed, a risk-stratification algorithm has been largely effective in identifying those people at highest risk who are eligible for primary prevention of sudden death with an implantable cardioverter-defibrillator (ICD) (7–10), thereby markedly reducing HCM-related mortality to 0.5% per year (7). Sudden death events are attributable to potentially lethal ventricular tachyarrhythmias (ventricular tachycardia/ventricular fibrillation) and usually occur in the presence of ≥1 the major risk markers (appropriate ICD interventions of 4% per year in patients implanted for primary prevention) (7–10). Some HCM patients may nevertheless die suddenly in the absence of all conventional risk factors (0.6% per year in non-ICD populations) (7).

Indeed, in the presence of underlying (and often unsuspected) HCM, participation in high-intensity competitive sports may itself promote ventricular tachycardia/ventricular fibrillation and act as a potent (yet modifiable) independent risk factor, even in the absence of conventional risk markers intrinsic to the disease process (3,7,11,12). Notably, the underlying electrophysiological substrate in HCM is unpredictable (1,2,7–10) and potentially subject to instability by interaction with physiological stresses inherent in athletic training and competition, including alternations in hydration, blood volume, and electrolytes, as well as the catecholamine surge.

Given these principles, it is difficult to apply conventional risk-stratification strategies to make reliable eligibility decisions specifically for aspiring competitive athletes with

HCM. The estimation of risk level based on phenotypic expression (e.g., specific LV wall thickness or LV outflow tract gradient) or other aspects of the clinical profile is a highly problematic endeavor. Such considerations are influenced by the morphological diversity of HCM and the unpredictable instability of the myocardial substrate, as well as the additive risk created by intense training and competition in susceptible patients with HCM (3). Therefore, in HCM, the most common cause of sudden death in young athletes (1–3), engagement in intense competitive sports is itself an acknowledged modifiable risk factor (1–3).

These observations necessitate conservative and prudent recommendations regarding sports eligibility applied in a homogeneous fashion across the broad HCM disease spectrum. This may unavoidably result in recommendations for disqualification in some athletes with HCM probably at low risk and unlikely to ever experience sudden death, who could potentially compete and train safely. Notably, the present disqualification/eligibility guidelines for competitive athletes with HCM do not differ measurably from those previously stated in the 36th Bethesda Conference (11), because alternative new data or insights have not emerged sufficient to substantially alter the recommendations.

On the other hand, the present American Heart Association/American College of Cardiology recommendations do not strictly exclude in absolute terms fully informed athletes from participating in competitive athletic programs as long as such a decision is ultimately made in concert with their physician and third-party interests (e.g., high schools and colleges). Although this expert consensus report serves as a prudent guideline regarding sports eligibility or disqualification, there will always be tolerance in the system for some degree of flexibility, individual responsibility, and choice in making these decisions for individual student athlete-patients.

Genotype Positive-Phenotype Negative

An increasing number of HCM family members are recognized with documented pathogenic (disease-causing) sarcomere mutations, but in the absence of a clinical HCM phenotype (i.e., LV hypertrophy) (5,13). Such patients have been identified at a broad range of ages, although they are most commonly adolescents and young adults, and some wish to engage in competitive sports.

Spontaneous conversion to LV hypertrophy in this subset appears to occur most often in adolescence between 12 and 20 years of age (1,13) but has also been observed in midlife and beyond (14,15). Nevertheless, such changes are unpredictable, and some genetically affected people will probably never develop the HCM phenotype. Spontaneous morphological conversions are not usually accompanied by cardiac symptoms, disease progression, or events (1,2,13–15). However, once LV hypertrophy

evolves, that person may theoretically be subject to an unstable HCM electrophysiological substrate.

With negative or ambiguous genetic test results, potentially affected relatives can nevertheless be suspected clinically by the presence of several echocardiographic or cardiovascular magnetic resonance (CMR) findings in the nonhypertrophied myocardium, that is, blood-filled crypts, elongated mitral valve leaflets, diastolic dysfunction, and myocardial scarring (5,16–19). At present, the risk for sudden death in gene-positive-phenotype-negative family members appears to be extremely low and likely no different from the risk in the general population of the same age without heart disease (5,20). CMR imaging is also an important consideration in family members who are gene positive and judged to be phenotype negative based on echocardiography, because areas of segmental LV hypertrophy may be detected only by CMR, particularly in the anterolateral free wall and apex (6,21).

Recommendations

1. **Participation in competitive athletics for asymptomatic, genotype-positive HCM patients without evidence of LV hypertrophy by 2-dimensional echocardiography and CMR is reasonable, particularly in the absence of a family history of HCM-related sudden death (Class IIa; Level of Evidence C).**
2. **Athletes with a probable or unequivocal clinical expression and diagnosis of HCM (ie, with the disease phenotype of LV hypertrophy) should not participate in most competitive sports, with the exception of those of low intensity (class IA sports) (see “Classification of Sport” [22]). This recommendation is independent of age, sex, magnitude of LV hypertrophy, particular sarcomere mutation, presence or absence of LV outflow obstruction (at rest or with physiological exercise), absence of prior cardiac symptoms, presence or absence of late gadolinium enhancement (fibrosis) on CMR, and whether major interventions such as surgical myectomy or alcohol ablation have been performed previously (Class III; Level of Evidence C).**
3. **Pharmacological agents (e.g., β -blockers) to control cardiac-related symptoms or ventricular tachyarrhythmias should not be administered for the sole purpose of permitting participation in high-intensity sports. Notably, such drugs may also be inconsistent with maximal physical performance in most sports (Class III; Level of Evidence C).**
4. **Prophylactic ICDs should not be placed in athlete-patients with HCM for the sole or primary purpose of permitting participation in high-intensity sports competition because of the possibility of device-related complications. ICD indications for competitive athletes with HCM should not differ from those in nonathlete patients with HCM (Class III; Level of Evidence B).**

Other recommendations for sports participation in patients with HCM and ICDs can be found in the Task Force 9 report on “Arrhythmias and Conduction Defects” (23).

LV NONCOMPACTION

LV noncompaction (LVNC) is an uncommon and recently recognized cardiac disease with sporadic or familial occurrence (24). Its true incidence and prevalence are not known, in part because of difficulty in making the diagnosis and lack of agreement on criteria, as well as its heterogeneous clinical spectrum and usual requirement of CMR for reliable diagnosis. Furthermore, its clinical presentation and implications differ with respect to genetic pathogenesis, race/ethnic origin, presence in isolation or in association with other diseases, or depending on the presence or absence of right ventricular involvement (25).

The natural history of LVNC remains incompletely resolved because of its relatively recent recognition with a short available follow-up period (26–34). The clinical expression of LVNC is variable, even within families: with or without symptoms, heart failure, atrial and ventricular arrhythmias or preexcitatory pathways, thromboembolic events, or sudden death (35). While LVNC patients with heart failure and systolic dysfunction, thromboembolic events, and sudden cardiac death have been reported (26,31,33,34), many uncomplicated cases are less likely to be recognized or appear in the literature (34). Risk for adverse consequences, including mortality, presently appear to be largely associated with LV systolic dysfunction or ventricular tachyarrhythmias (34).

Few competitive athletes with LVNC have been reported clinically, and therefore, the consequences of LVNC in this specific population are unknown. Furthermore, to date, forensic registries of sudden deaths in young athletes do not include LVNC as a cause (3), although the diagnosis may still be widely underappreciated in the routine medical examiner autopsy setting. Therefore, given the lack of long-term follow-up studies and other obstacles, it is not yet possible to reliably apply risk-stratification strategies to new patients (or athletes) with LVNC. This is not unlike the situation with other uncommon myocardial diseases for which few data concerning sudden death risk during competitive sports are available (e.g., dilated cardiomyopathy [DCM] or infiltrative diseases). Therefore, the complete natural history of noncompacted ventricular myocardium remains unresolved.

A variety of inheritance patterns have been reported (ie, autosomal dominant, autosomal recessive, and X-linked) (24,27). Mutations in genes encoding sarcomeric proteins, which previously have been implicated in the

pathogenesis of HCM and DCM, have also been identified in patients with isolated LVNC (24,27). These observations suggest that LVNC shares genetic overlap with other cardiomyopathies, and indeed, some individual patients have been reported with morphological features consistent with both HCM and LVNC (32).

LVNC is thought to be caused by the intrauterine arrest of the compaction process of the primordial embryonic myocardium. Diagnosis is considered in the presence of a 2-layered LV chamber that consists of noncompacted trabeculations with intertrabecular recesses layered on top of the typical compacted myocardium, with or without systolic dysfunction. The trabeculated layer is predominantly confined to the distal and mid portions of the LV chamber, sparing the base. Currently, there are no universally accepted criteria or guidelines for the morphological diagnosis of LVNC, although a ratio of noncompacted to compacted myocardium $>2.1:1$ at end systole (echocardiography) or $>2.3:1$ in end diastole (CMR) have been proposed (30,32,36). It is uncertain how athletic training may alter those definitions (28,29) or the frequency of LVNC-appearing morphology in a normal athlete population. CMR is generally superior to echocardiography for identification of regions of non-compacted myocardium and for more definitive diagnosis of LVNC.

Recommendations

1. **Until more clinical information is available, participation in competitive sports may be considered for asymptomatic patients with a diagnosis of LVNC and normal systolic function, without important ventricular tachyarrhythmias on ambulatory monitoring or exercise testing, and specifically with no prior history of unexplained syncope (Class IIb; Level of Evidence C).**
2. **Athletes with an unequivocal diagnosis of LVNC and impaired systolic function or important atrial or ventricular tachyarrhythmias on ambulatory monitoring or exercise testing (or with a history of syncope) should not participate in competitive sports, with the possible exception of low-intensity class 1A sports, at least until more clinical information is available (Class III; Level of Evidence C).**

OTHER MYOCARDIAL DISEASES

A number of other uncommon diseases of the myocardium deserve consideration as potential causes of sudden death in athletes. These include DCM (attributable to a variety of causes, including genetic), primary non-hypertrophied restrictive cardiomyopathy, and systemic infiltrative diseases with secondary cardiac involvement, such as sarcoidosis. Few data are available at present

regarding the relative risks of athletic training and competition in athletes with these myocardial diseases.

It is important to differentiate physiological LV enlargement caused by systematic training from pathological DCM. Long-term aerobic athletic training can lead to cardiac morphological changes, including increased LV cavity dimension and calculated mass. Increased cavity size can produce a higher stroke volume, and thus, the ejection fraction at rest may be in the low-normal to mildly reduced range. Up to 15% of trained athletes will have substantial enlargement of the LV cavity, with end-diastolic dimensions up to 70 mm in men and 66 mm in women (37,38). Ejection fraction in trained athletes has been shown to be as low as 45% (37). Whether newer imaging techniques such as myocardial Doppler tissue imaging, strain imaging, or contrast-CMR scanning can differentiate patients with borderline LV enlargement and low-normal or mildly reduced ejection fraction from DCM is unresolved.

It is unclear whether asymptomatic patients with DCM are at risk for sudden death during competitive athletics, because ventricular tachyarrhythmias are most common in patients with more advanced disease, that is, with cardiac symptoms and lower ejection fraction.

Recommendations

1. **Symptomatic athletes with DCM, primary non-hypertrophied restrictive cardiomyopathy, and infiltrative cardiac myopathies should not participate in most competitive sports, with the possible exception of low-intensity (class 1A sports) in selected cases, at least until more information is available (Class III; Level of Evidence C).**

MYOCARDITIS

General Considerations

Myocarditis commonly presents with disproportionate dyspnea on exertion, chest pain, and arrhythmias. It can also present as an acute myocardial infarction-like syndrome with sudden death in the presence of normal epicardial coronary arteries (39-44). The contribution of myocarditis to cardiovascular sudden death varies significantly with age, causing cardiovascular sudden death in $\approx 2\%$ of infants, 5% of children, and 4% to 7.5% of athletes (3,40). Higher rates of myocarditis are occasionally reported in postmortem studies from general populations younger than 35 to 40 years of age (41). Most cardiovascular sudden deaths attributable to myocarditis occur in males (42), and in some cases, myocarditis results in sudden death without antecedent symptoms or macroscopic cardiac abnormalities (40,42,43).

The data linking myocarditis to sudden death are strong and include autopsy studies and experimental myocarditis

models. For example, strenuous physical exertion was associated with sudden death in a cohort of U.S. military recruits, with the most frequent underlying cause being myocarditis (44). Case series of sudden death in athletes have established myocarditis as a significant risk in this specific group (3). In a murine model of coxsackie B3 myocarditis, 60 minutes of swimming daily increased viral titers, worsened cardiomyopathy, and increased the likelihood of death (45). In a chronic autoimmune myocarditis model, humoral and cellular immunity directed against heart tissues increased with treadmill exercise (46). Unlike heart failure, the risk of sudden death caused by myocarditis does not appear to correlate with the severity of myocardial inflammation (40). Sudden death has been observed occasionally after myopericarditis in association with normal LV function (47-49).

The pathogenesis of myocarditis consists of 3 overlapping phases: acute injury, often caused by a virus; the host innate and acquired immunologic response; and finally, recovery or a transition to scar and DCM. There is rarely a clear distinction between these phases clinically. The initial injury may cause an acute DCM with contractile impairment mediated by cytokines generated by the local inflammatory process. Several months later, the same dilated ventricle may have poor contractility caused by diffuse scar, with little or no inflammation. The transition from acute myocarditis to chronic DCM probably occurs over months, with substantial individual variability (50).

In clinical practice, myocarditis is often suspected but infrequently confirmed by endomyocardial biopsy, which creates a need for noninvasive diagnostic criteria to guide recommendations for athletic participation. For the purposes of this document, probable acute myocarditis is diagnosed when both of the following criteria are met:

1. A clinical syndrome that includes acute heart failure, angina-type chest pain, or myopericarditis of <3 months' duration.
2. An otherwise unexplained elevation in serum troponin; electrocardiographic features of cardiac ischemia; otherwise unexplained high-degree AV block or arrhythmias; wall motion abnormalities; pericardial effusion on echocardiography or CMR imaging. Additional CMR findings that suggest myocarditis in the acute clinical setting include characteristic alterations in tissue signal on T2- or T1-weighted images and the presence of late gadolinium enhancement (LGE).

CMR features that may be used to diagnose probable myocarditis include a regional increase in water content visible on T2-weighted images, an increase in regional contrast-enhanced T1-weighted epicardial or mid-myocardial signal obtained within a few minutes of the gadolinium bolus ("hyperemia" or "early-enhancement"

sequences), and epicardial or midmyocardial LGE (51). A regional and reversible increase in wall thickness that indicates myocardial edema is a supportive finding of acute myocarditis. Myocardial fibrosis, the late sequelae of myocarditis characteristic of DCM, may be indistinguishable from active myocarditis on LGE sequences. The sensitivity of CMR for myocarditis also decreases a few weeks after the initial illness (51).

Although acute myocarditis is associated with the characteristic findings of myocardial injury described in the diagnostic criteria above, there is no sensitive or specific test that can determine when the inflammatory process ends. DCM associated with acute myocarditis often resolves over 6 to 12 months. Athletes in whom the findings of acute inflammation have resolved may still have a risk of arrhythmias related to the resultant myocardial scar. The presence of LGE may convey a heightened risk for arrhythmias (52). The interval between initial assessment and retesting before resumption of sports will vary depending on the severity of the initial illness. A reasonable minimum interval for retesting based on experimental models is 3 to 6 months. The recommendations presented here recognize these gaps in knowledge and the need for additional clinical research to refine risk stratification for sudden death after acute myocarditis.

A diagnosis of myocarditis by biopsy is usually not required to guide clinical management, but a biopsy may be considered in select cases according to current professional society recommendations from the American Heart Association, American College of Cardiology, and European Society of Cardiology (53). Confirmation of myocarditis by endomyocardial biopsy creates a definitive diagnosis.

Recommendations

1. **Before returning to competitive sports, athletes who initially present with an acute clinical syndrome consistent with myocarditis should undergo a resting echocardiogram, 24-hour Holter monitoring, and an exercise ECG no less than 3 to 6 months after the initial illness (Class I; Level of Evidence C).**
2. **It is reasonable that athletes resume training and competition if all of the following criteria are met (Class IIa; Level of Evidence C):**
 - a. Ventricular systolic function has returned to the normal range.
 - b. Serum markers of myocardial injury, inflammation, and heart failure have normalized.
 - c. Clinically relevant arrhythmias such as frequent or complex repetitive forms of ventricular or supraventricular ectopic activity are absent on Holter monitor and graded exercise ECGs.

At present, it is unresolved whether resolution of myocarditis-related LGE should be required to permit return to competitive sports.

- 3. Athletes with probable or definite myocarditis should not participate in competitive sports while active inflammation is present. This recommendation is independent of age, gender, and LV function (Class III; Level of Evidence C).**

ARRHYTHMOGENIC RIGHT VENTRICULAR CARDIOMYOPATHY

Arrhythmogenic right ventricular cardiomyopathy (ARVC) is a cause of sudden death in young people and athletes, particularly in the northeastern (Veneto) region of Italy (54), but is seemingly less common in the United States (3). ARVC is characterized by a broad phenotypic spectrum and characteristically by loss of myocytes in the right ventricular myocardium, with fatty or fibrofatty replacement, which results in segmental or diffuse wall thinning, but there is also frequent involvement of the LV and an association with myocarditis (55). Genetics studies have demonstrated that ARVC is a desmosomal cardiomyopathy that results from genetically defective cell-adhesion proteins such as plakoglobin, plakophilin-2, desmoplakin, desmocollin-2, and desmoglein-2 (56,57).

Clinical diagnosis can be challenging but relies largely on familial occurrence, left bundle-branch pattern ventricular tachyarrhythmias, ECG findings of T-wave inversion in precordial leads V₁ through V₃, and epsilon waves, as well as right ventricular dilation or segmental wall motion abnormalities, aneurysm formation, or fatty deposition in the right ventricular wall identified with CMR imaging if substantial and unequivocal (or by biopsy tissue analysis). Diagnostic criteria for ARVC have been revised and updated and now include quantitative variables (58).

These criteria include global or regional structural dysfunction, as documented by echocardiography or CMR, biopsy abnormalities, ECG repolarization or depolarization abnormalities, arrhythmias, and family history. Each of these criteria is separated into major and minor criteria based on the severity of the finding. Patients meet an ARVC diagnosis if they possess 2 major, or 1 major and 2 minor, or 4 minor criteria. Borderline patients are those with 1 major and 1 minor criterion or 3 minor criteria. Patients with possible ARVC have 1 major criterion or 2 minor criteria. Athletes with borderline or possible ARVC, as well as those who are genotype positive-phenotype negative, should receive continued follow-up, because ARVC may progress phenotypically, and become more clinically apparent with time.

There is evidence in the experimental murine model that exercise increases the penetrance and arrhythmic risk in mutational carriers of ARVC (59). More recently,

these data have been confirmed in genetically positive patients (60), which is particularly relevant to the athlete, raising concern not only with regard to competitive sports but also regarding participation in moderate to extreme recreational physical activities.

Ventricular tachyarrhythmias and sudden death in ARVC commonly occur during exertion, including competitive sports (55,60,61), and frequent endurance exercise increases the risk for ventricular tachycardia/ventricular fibrillation and heart failure (60). However, risk factors for sudden cardiac death in ARVC are not as well defined as in HCM (1,2,7,8). There is general agreement that a prior history of sudden cardiac death, sustained ventricular tachycardia, or syncope represent the most important prognostic factors and define many high-risk patients who are most appropriately treated with a primary prevention ICD (62-64).

Recommendations

- 1. Athletes with a definite diagnosis of ARVC should not participate in most competitive sports, with the possible exception of low-intensity class 1A sports (Class III; Level of Evidence C).**
- 2. Athletes with a borderline diagnosis of ARVC should not participate in most competitive sports, with the possible exception of low-intensity class 1A sports (Class III; Level of Evidence C).**
- 3. Athletes with a possible diagnosis of ARVC should not participate in most competitive sports, with the possible exception of low-intensity class 1A sports (Class III; Level of Evidence C).**
- 4. Prophylactic ICD placement in athlete-patients with ARVC for the sole or primary purpose of permitting participation in high-intensity sports competition is not recommended because of the possibility of device-related complications (Class III; Level of Evidence C).**

Other recommendations for sports participation in patients with ARVC and ICDs can be found in the Task Force 9 report on "Arrhythmias and Conduction Defects" (23).

PERICARDITIS

The causes of pericarditis/myopericarditis are varied and are either infectious or noninfectious. The natural history is incompletely resolved, although long-term prognosis is generally favorable. The diagnosis of acute pericarditis is typically based on clinical criteria: chest pain, pericardial rub, ST-segment elevation, or new/worsening pericardial effusion. This syndrome may be considered part of the clinical spectrum of myocarditis. Recurrences are a significant consideration, and follow-up surveillance with echocardiography or CMR is recommended to exclude pericardial thickening or restriction consistent with restrictive pericarditis (50).

Recommendations

1. Athletes with pericarditis, regardless of its pathogenesis, should not participate in competitive sports during the acute phase. Such athletes can return to full activity when there is complete absence of evidence for active disease, including effusion by echocardiography, and when serum

markers of inflammation have normalized. For pericarditis associated with evidence of myocardial involvement, eligibility should also be based on the course of myocarditis. Chronic pericardial disease that results in constriction disqualifies the person from all competitive sports (Class III; Level of Evidence C).

DISCLOSURES

Writing Group Disclosures

Writing Group Member	Employment	Research Grant	Other Research Support	Speakers Bureau/Honoraria	Expert Witness	Ownership Interest	Consultant/Advisory Board	Other
Barry J. Maron	Minneapolis Heart Institute Foundation	None	None	None	None	None	None	None
Michael J. Ackerman	Mayo Clinic	NIH (R01 grants) [†]	None	None	None	None	Boston Scientific*; Gilead Sciences*; Medtronic*; St. Jude Medical*	Transgenomic [†]
Robert O. Bonow	Northwestern University	None	None	None	None	None	None	None
Leslie T. Cooper, Jr	Mayo Clinic	None	None	None	None	None	None	None
N.A. Mark Estes III	Tufts Medical Center	None	None	None	None	None	Medtronic*; St. Jude Medical [†] ; Boston Scientific [†]	None
Mark S. Link	Tufts Medical Center	None	None	None	None	None	None	None
Martin S. Maron	Tufts Medical Center	None	None	None	None	None	None	None
Rick A. Nishimura	Mayo Clinic	None	None	None	None	None	None	None
James E. Udelson	Tufts Medical Center	Gilead (DSMB member for a clinical trial)*	None	None	None	None	None	None

This table represents the relationships of writing group members that may be perceived as actual or reasonably perceived conflicts of interest as reported on the Disclosure Questionnaire, which all members of the writing group are required to complete and submit. A relationship is considered to be "significant" if (a) the person receives \$10,000 or more during any 12-month period, or 5% or more of the person's gross income; or (b) the person owns 5% or more of the voting stock or share of the entity, or owns \$10,000 or more of the fair market value of the entity. A relationship is considered to be "modest" if it is less than "significant" under the preceding definition.

*Modest.

[†]Significant.

Reviewer Disclosures

Reviewer	Employment	Research Grant	Other Research Support	Speakers Bureau/Honoraria	Expert Witness	Ownership Interest	Consultant/Advisory Board	Other
Linda J. Addonizio	Columbia University	None	None	None	None	None	None	None
Eugene DePasquale	University of California, Los Angeles	None	None	None	None	None	None	None
Michelle A. Grenier	University of Mississippi	None	None	None	None	None	None	None

This table represents the relationships of reviewers that may be perceived as actual or reasonably perceived conflicts of interest as reported on the Disclosure Questionnaire, which all reviewers are required to complete and submit. A relationship is considered to be "significant" if (a) the person receives \$10,000 or more during any 12-month period, or 5% or more of the person's gross income; or (b) the person owns 5% or more of the voting stock or share of the entity, or owns \$10,000 or more of the fair market value of the entity. A relationship is considered to be "modest" if it is less than "significant" under the preceding definition.

REFERENCES

- Maron BJ, Maron MS. Hypertrophic cardiomyopathy. *Lancet*. 2013;381:242-55. [http://dx.doi.org/10.1016/S0140-6736\(12\)60397-3](http://dx.doi.org/10.1016/S0140-6736(12)60397-3).
- Maron BJ, Ommen SR, Semsarian C, Spirito P, Olivetto I, Maron MS. Hypertrophic cardiomyopathy: present and future, with translation into contemporary cardiovascular medicine [published correction appears in *J Am Coll Cardiol*. 2014;64:1188]. *J Am Coll Cardiol*. 2014;64:83-99. <http://dx.doi.org/10.1016/j.jacc.2014.05.003>.
- Maron BJ, Doerer JJ, Haas TS, Tierney DM, Mueller FO. Sudden deaths in young competitive athletes: analysis of 1866 deaths in the United States, 1980-2006. *Circulation*. 2009;119:1085-92. <http://dx.doi.org/10.1161/CIRCULATIONAHA.108.804617>.
- 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 4111 subjects in the CARDIA Study: Coronary Artery Risk Development in (Young) Adults. *Circulation*. 1995;92:785-9.
- Maron BJ, Maron MS, Semsarian C. Genetics of hypertrophic cardiomyopathy after 20 years: clinical perspectives. *J Am Coll Cardiol*. 2012;60:705-15. <http://dx.doi.org/10.1016/j.jacc.2012.02.068>.
- Maron MS, Maron BJ, Harrigan C, Buross J, Gibson CM, Olivetto I, Biller L, Lesser JR, Udelson JE, Manning WJ, Appelbaum E. Hypertrophic cardiomyopathy phenotype revisited after 50 years with cardiovascular magnetic resonance. *J Am Coll Cardiol*. 2009;54:220-8. <http://dx.doi.org/10.1016/j.jacc.2009.05.006>.
- Maron BJ, Rowin EJ, Casey SA, Link MS, Lesser JR, Chan RH, Garberich RF, Udelson JE, Maron MS. Hypertrophic cardiomyopathy in adulthood associated with low cardiovascular mortality with contemporary management strategies. *J Am Coll Cardiol*. 2015;65:1915-28. <http://dx.doi.org/10.1016/j.jacc.2015.02.061>.
- Maron BJ, Spirito P, Shen WK, Haas TS, Formisano F, Link MS, Epstein AE, Almquist AK, Daubert JP, Lawrence T, Boriani G, Estes NA 3rd, Favale S, Piccinino M, Winters SL, Santini M, Betocchi S, Arribas F, Sherrid MV, Buja G, Semsarian C, Bruzzi P. Implantable cardioverter-defibrillators and prevention of sudden cardiac death in hypertrophic cardiomyopathy [published correction appears in *JAMA*. 2007;298:1516]. *JAMA*. 2007;298:405-12. <http://dx.doi.org/10.1001/jama.298.4.405>.
- Schinkel AF, Vriesendorp PA, Sijbrands EJ, Jordaens LJ, ten Cate FJ, Michels M. Outcome and complications after implantable cardioverter defibrillator therapy in hypertrophic cardiomyopathy: systematic review and meta-analysis. *Circ Heart Fail*. 2012;5:552-9. <http://dx.doi.org/10.1161/CIRCHEARTFAILURE.112.969626>.
- Maron BJ, Spirito P, Ackerman MJ, Casey SA, Semsarian C, Estes NA 3rd, Shannon KM, Ashley EA, Day SM, Pacileo G, Formisano F, Devoto E, Anastasakis A, Bos JM, Woo A, Autore C, Pass RH, Boriani G, Garberich RF, Almquist AK, Russell MW, Boni L, Berger S, Maron MS, Link MS. Prevention of sudden cardiac death with implantable cardioverter-defibrillators in children and adolescents with hypertrophic cardiomyopathy. *J Am Coll Cardiol*. 2013;61:1527-35. <http://dx.doi.org/10.1016/j.jacc.2013.01.037>.
- Maron BJ, Ackerman MJ, Nishimura RA, Pyeritz RE, Towbin JA, Udelson JE. Task Force 4: HCM and other cardiomyopathies, mitral valve prolapse, myocarditis and Marfan syndrome. *J Am Coll Cardiol*. 2005;45:1340-5.
- Gersh BJ, Maron BJ, Bonow RO, Dearani JA, Fifer MA, Link MS, Naidu SS, Nishimura RA, Ommen SR, Rakowski H, Seidman CE, Towbin JA, Udelson JE, Yancy CW. 2011 ACCF/AHA guideline for the diagnosis and treatment of hypertrophic cardiomyopathy: executive summary: a report of the American College of Cardiology Foundation/American Heart Association Task Force on Practice Guidelines. *J Am Coll Cardiol*. 2011;58:2703-38. <http://dx.doi.org/10.1016/j.jacc.2011.10.825>.
- Maron BJ, Yeates L, Semsarian C. Clinical challenges of genotype positive (+)-phenotype negative (-) family members in hypertrophic cardiomyopathy. *Am J Cardiol*. 2011;107:604-8. <http://dx.doi.org/10.1016/j.amjcard.2010.10.022>.
- Maron BJ, Niimura H, Casey SA, Soper MK, Wright GB, Seidman JG, Seidman CE. Development of left ventricular hypertrophy in adults in hypertrophic cardiomyopathy caused by cardiac myosin-binding protein C gene mutations. *J Am Coll Cardiol*. 2001;38:315-21.
- Maron BJ, Haas TS, Kitner C, Lesser JR. Onset of apical hypertrophic cardiomyopathy in adulthood. *Am J Cardiol*. 2011;108:1783-7. <http://dx.doi.org/10.1016/j.amjcard.2011.07.048>.
- Ho CY, Sweitzer NK, McDonough B, Maron BJ, Casey SA, Seidman JM, Seidman CE, Solomon SD. Assessment of diastolic function with Doppler tissue imaging to predict genotype in preclinical hypertrophic cardiomyopathy. *Circulation*. 2002;105:2992-7.
- Maron MS, Rowin EJ, Lin D, Appelbaum E, Chan RH, Gibson CM, Lesser JR, Lindberg J, Haas TS, Udelson JE, Manning WJ, Maron BJ. Prevalence and clinical profile of myocardial crypts in hypertrophic cardiomyopathy. *Circ Cardiovasc Imaging*. 2012;5:441-7. <http://dx.doi.org/10.1161/CIRCIMAGING.112.972760>.
- Rowin EJ, Maron MS, Lesser JR, Maron BJ. CMR with late gadolinium enhancement in genotype positive-phenotype negative hypertrophic cardiomyopathy. *J Am Coll Cardiol Img*. 2012;5:119-22. <http://dx.doi.org/10.1016/j.jcmg.2011.08.020>.
- Maron MS, Olivetto I, Harrigan C, Appelbaum E, Gibson CM, Lesser JR, Haas TS, Udelson JE, Manning WJ, Maron BJ. Mitral valve abnormalities identified by cardiovascular magnetic resonance represent a primary phenotypic expression of hypertrophic cardiomyopathy. *Circulation*. 2011;124:40-7. <http://dx.doi.org/10.1161/CIRCULATIONAHA.110.985812>.
- Christiaans I, Lekan dit Deprez RH, van Langen IM, Wilde AA. Ventricular fibrillation in MYH7-related hypertrophic cardiomyopathy before onset of ventricular hypertrophy. *Heart Rhythm*. 2009;6:1366-9. <http://dx.doi.org/10.1016/j.hrthm.2009.04.029>.
- Rickers C, Wilke NM, Jerosch-Herold M, Casey SA, Panse P, Panse N, Weil J, Zenovich AG, Maron BJ. Utility of cardiac magnetic resonance imaging in the diagnosis of hypertrophic cardiomyopathy. *Circulation*. 2005;112:855-61. <http://dx.doi.org/10.1161/CIRCULATIONAHA.104.507723>.
- Levine BD, Baggish AL, Kovacs RJ, Link MS, Maron MS, Mitchell JH, on behalf of the American Heart Association Electrocardiography and Arrhythmias Committee of the Council on Clinical Cardiology, Council on Cardiovascular Disease in the Young, Council on Cardiovascular and Stroke Nursing, Council on Functional Genomics and Translational Biology, and the American College of Cardiology. Eligibility and disqualification recommendations for competitive athletes with cardiovascular abnormalities: Task Force 1: classification of sport: dynamic, static, and impact: a scientific statement from the American Heart Association and American College of Cardiology. *J Am Coll Cardiol*. 2015 In Press. <http://dx.doi.org/10.1016/j.jacc.2015.09.033>.
- Zipes DP, Link MS, Ackerman MJ, Kovacs RJ, Myerburg RJ, Estes NAM 3rd, on behalf of the American Heart Association Electrocardiography and Arrhythmias Committee of the Council on Clinical Cardiology, Council on Cardiovascular Disease in the Young, Council on Cardiovascular and Stroke Nursing, Council on Functional Genomics and Translational Biology, and the American College of Cardiology. Eligibility and disqualification recommendations for competitive athletes with cardiovascular abnormalities: Task Force 9: arrhythmias and conduction defects: a scientific statement from the American Heart Association and American College of Cardiology. *J Am Coll Cardiol*. 2015 In Press. <http://dx.doi.org/10.1016/j.jacc.2015.09.041>.
- Maron BJ, Towbin JA, Thiene G, Antzevitch C, Corrado D, Arnett D, Moss AJ, Seidman CE, Young JB. Contemporary definitions and classification of the cardiomyopathies: an American Heart Association Scientific Statement from the Council on Clinical Cardiology, Heart Failure and Transplantation Committee; Quality of Care and Outcomes Research and Functional Genomics and Translational Biology Interdisciplinary Working Groups; and Council on Epidemiology and Prevention. *Circulation*. 2006;113:1807-16. <http://dx.doi.org/10.1161/CIRCULATIONAHA.106.174287>.
- Stacey RB, Andersen M, Haag J, Hall ME, McLeod G, Upadhyaya B, Hundley WG, Thohan V. Right ventricular morphology and systolic function in left ventricular noncompaction cardiomyopathy. *Am J Cardiol*. 2014;113:1018-23. <http://dx.doi.org/10.1016/j.amjcard.2013.12.008>.
- 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.
- Klaassen S, Probst S, Oechslin E, Gerull B, Krings G, Schuler P, Greutmann M, Hurlimann D, Yegitbasi M, Pons L, Gramlich M, Drenckhahn JD, Heuser A, Berger F, Jenni R, Thierfelder L. Mutations in sarcomere protein genes in left ventricular noncompaction.

- Circulation. 2008;117:2893-901. <http://dx.doi.org/10.1161/CIRCULATIONAHA.107.746164>.
28. Gati S, Chandra N, Bennett RL, Reed M, Kervio G, Panoulas VF, Ghani S, Sheikh N, Zaidi A, Wilson M, Wilson M, Papadakis M, Carré F, Sharma S. Increased left ventricular trabeculation in highly trained athletes: do we need more stringent criteria for the diagnosis of left ventricular non-compaction in athletes [published correction appears in Heart. 2013;99:506]? Heart. 2013;99:401-8. <http://dx.doi.org/10.1136/heartjnl-2012-303418>.
29. Ganga HV, Thompson PD. Sports participation in non-compaction cardiomyopathy: a systematic review. Br J Sports Med. 2014;48:1466-71. <http://dx.doi.org/10.1136/bjsports-2012-091855>.
30. Thavendiranathan P, Dahiya A, Phelan D, Desai MY, Tang WH. Isolated left ventricular non-compaction controversies in diagnostic criteria, adverse outcomes and management. Heart. 2013;99:681-9. <http://dx.doi.org/10.1136/heartjnl-2012-302816>.
31. Caliskan K, Michels M, Geleijnse ML, van Domburg RT, van der Boon R, Balk AH, Simoons ML. Frequency of asymptomatic disease among family members with noncompaction cardiomyopathy. Am J Cardiol. 2012;110:1512-7. <http://dx.doi.org/10.1016/j.amjcard.2012.07.009>.
32. Kelley-Hedgpeeth A, Towbin JA, Maron MS. Images in cardiovascular medicine: overlapping phenotypes: left ventricular noncompaction and hypertrophic cardiomyopathy. Circulation. 2009;119:e588-9. <http://dx.doi.org/10.1161/CIRCULATIONAHA.108.829564>.
33. Greutmann M, Mah ML, Silversides CK, Klaassen S, Attenhofer Jost CH, Jenni R, Oechslin EN. Predictors of adverse outcome in adolescents and adults with isolated left ventricular noncompaction. Am J Cardiol. 2012;109:276-81. <http://dx.doi.org/10.1016/j.amjcard.2011.08.043>.
34. Brescia ST, Rossano JW, Pignatelli R, Jefferies JL, Price JF, Decker JA, Denfield SW, Dreyer WJ, Smith O, Towbin JA, Kim JJ. Mortality and sudden death in pediatric left ventricular noncompaction in a tertiary referral center. Circulation. 2013;127:2202-8. <http://dx.doi.org/10.1161/CIRCULATIONAHA.113.002511>.
35. Paterick TE, Tajik AJ. Left ventricular non-compaction: a diagnostically challenging cardiomyopathy. Circ J. 2012;76:1556-62.
36. Jenni R, Oechslin E, Schneider J, Attenhofer Jost C, Kaufmann PA. Echocardiographic and pathoanatomical characteristics of isolated left ventricular non-compaction: a step towards classification as a distinct cardiomyopathy. Heart. 2001;86:666-71.
37. Pelliccia A, Culasso F, Di Paolo FM, Maron BJ. Physiologic left ventricular cavity dilatation in elite athletes. Ann Intern Med. 1999;130:23-31.
38. Pelliccia A, Maron BJ, Culasso F, Spataro A, Caselli G. Athlete's heart in women: echocardiographic characterization of highly trained elite female athletes. JAMA. 1996;276:211-5.
39. Karjalainen J, Heikkilä 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. <http://dx.doi.org/10.1053/ehj.1998.1444>.
40. Weber MA, Ashworth MT, Risdon RA, Malone M, Burch M, Sebire NJ. Clinicopathological features of paediatric deaths due to myocarditis: an autopsy series. Arch Dis Child. 2008;93:594-8. <http://dx.doi.org/10.1136/adc.2007.128686>.
41. Morentin B, Suárez-Mier MP, Aguilera B, Bodegas A. Myocardial disease mortality in children and young adults: a population-based observational study [in Spanish]. Rev Esp Cardiol. 2006;59:238-46.
42. Kitulwatté ID, Kim PJ, Pollanen MS. Sudden death related myocarditis: a study of 56 cases. Forensic Sci Med Pathol. 2010;6:13-9. <http://dx.doi.org/10.1007/s12024-009-9125-5>.
43. Corrado D, Basso C, Thiene G. Sudden cardiac death in young people with apparently normal heart. Cardiovasc Res. 2001;50:399-408.
44. 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.
45. Kiel RJ, Smith FE, Chason J, Khatib R, Reyes MP. Coxsackievirus B3 myocarditis in C3H/HeJ mice: description of an inbred model and the effect of exercise on virulence. Eur J Epidemiol. 1989;5:348-50.
46. Hosenpud JD, Campbell SM, Niles NR, Lee J, Mendelson D, Hart MV. Exercise induced augmentation of cellular and humoral autoimmunity associated with increased cardiac dilatation in experimental autoimmune myocarditis. Cardiovasc Res. 1987;21:217-22.
47. Machado S, Roubille F, Gahide G, Vernhet-Kovacsik H, Cornillet L, Cung TT, Sportouch-Dukhan C, Raczká F, Pasquière JL, Gervasoni R, Macia JC, Cransac F, Davy JM, Plot C, Leclercq F. Can troponin elevation predict worse prognosis in patients with acute pericarditis? Ann Cardiol Angeiol (Paris). 2010;59:1-7. <http://dx.doi.org/10.1016/j.jancard.2009.07.009>.
48. Imazio M, Cooper LT. Management of myopericarditis. Expert Rev Cardiovasc Ther. 2013;11:193-201. <http://dx.doi.org/10.1586/erc.12.184>.
49. Imazio M, Brucato A, Barbieri A, Ferroni F, Maestroni S, Ligabue G, Chinaglia A, Cumetti D, Della Casa G, Bonomi F, Mantovani F, Di Corato P, Lugli R, Faletti R, Leuzzi S, Bonamini R, Modena MG, Belli R. Good prognosis for pericarditis with and without myocardial involvement: results from a multicenter, prospective cohort study. Circulation. 2013;128:42-9. <http://dx.doi.org/10.1161/CIRCULATIONAHA.113.001531>.
50. Cooper LT Jr. Myocarditis. N Engl J Med. 2009;360:1526-38. <http://dx.doi.org/10.1056/NEJMra0800028>.
51. Friedrich MG, Sechtem U, Schulz-Menger J, Holmvang G, Alakija P, Cooper LT, White JA, Abdel-Aty H, Gutberlet M, Prasad S, Aletras A, Laissy JP, Paterson I, Filipchuk NG, Kumar A, Pauschinger M, Liu P, International Consensus Group on Cardiovascular Magnetic Resonance in Myocarditis. Cardiovascular magnetic resonance in myocarditis: a JACC White Paper. J Am Coll Cardiol. 2009;53:1475-87. <http://dx.doi.org/10.1016/j.jacc.2009.02.007>.
52. Grün S, Schumm J, Greulich S, Wagner A, Schneider S, Bruder O, Kispert EM, Hill S, Ong P, Klingel K, Kandolf R, Sechtem U, Mahrholdt H. Long-term follow-up of biopsy-proven viral myocarditis: predictors of mortality and incomplete recovery. J Am Coll Cardiol. 2012;59:1604-15. <http://dx.doi.org/10.1016/j.jacc.2012.01.007>.
53. Cooper LT, Baughman KL, Feldman AM, Frustaci A, Jessup M, Kuhl U, Levine GN, Narula J, Starling RC, Towbin J, Virmani R. The role of endomyocardial biopsy in the management of cardiovascular disease: a scientific statement from the American Heart Association, the American College of Cardiology, and the European Society of Cardiology. Circulation. 2007;116:2216-33. <http://dx.doi.org/10.1161/CIRCULATIONAHA.107.186093>.
54. Corrado D, Basso C, Pavei A, Michieli P, Schiavon M, Thiene G. Trends in sudden cardiovascular death in young competitive athletes after implementation of a preparticipation screening program. JAMA. 2006;296:1593-601. <http://dx.doi.org/10.1001/jama.296.13.1593>.
55. Basso C, Corrado D, Marcus FI, Nava A, Thiene G. Arrhythmogenic right ventricular cardiomyopathy. Lancet. 2009;373:1289-300. [http://dx.doi.org/10.1016/S0140-6736\(09\)60256-7](http://dx.doi.org/10.1016/S0140-6736(09)60256-7).
56. Corrado D, Thiene G. Arrhythmogenic right ventricular cardiomyopathy/dysplasia: clinical impact of molecular genetic studies. Circulation. 2006;113:1634-7. <http://dx.doi.org/10.1161/CIRCULATIONAHA.105.616490>.
57. Sen-Chowdhry S, Syrris P, McKenna WJ. Role of genetic analysis in the management of patients with arrhythmogenic right ventricular dysplasia/cardiomyopathy. J Am Coll Cardiol. 2007;50:1813-21. <http://dx.doi.org/10.1016/j.jacc.2007.08.008>.
58. Marcus FI, McKenna WJ, Sherrill D, Basso C, Bauce B, Bluemke DA, Calkins H, Corrado D, Cox MG, Daubert JP, Fontaine G, Gear K, Hauer R, Nava A, Picard MH, Protonotarios N, Saffitz JE, Sanborn DM, Steinberg JS, Tandri H, Thiene G, Towbin JA, Tsatsopoulou A, Wichter T, Zareba W. Diagnosis of arrhythmogenic right ventricular cardiomyopathy/dysplasia: proposed modification of the Task Force Criteria. Eur Heart J. 2010;31:806-14. <http://dx.doi.org/10.1093/eurheartj/ehq025>.
59. Kirchhof P, Fabritz L, Zwiener M, Witt H, Schäfers M, Zellerhoff S, Paul M, Athai T, Hiller KH, Baba HA, Breithardt G, Ruiz P, Wichter T, Levkau B. Age- and training-dependent development of arrhythmogenic right ventricular cardiomyopathy in heterozygous plakoglobin-deficient mice. Circulation. 2006;114:1799-806. <http://dx.doi.org/10.1161/CIRCULATIONAHA.106.624502>.
60. James CA, Bhonsale A, Tichnell C, Murray B, Russell SD, Tandri H, Tedford RJ, Judge DP, Calkins H. Exercise increases age-related penetrance and arrhythmic risk in arrhythmogenic right ventricular dysplasia/cardiomyopathy-associated desmosomal mutation carriers. J Am Coll Cardiol. 2013;62:1290-7. <http://dx.doi.org/10.1016/j.jacc.2013.06.033>.
61. Thiene G, Nava A, Corrado D, Rossi L, Pennelli N. Right ventricular cardiomyopathy and sudden death in young people. N Engl J Med. 1988;318:129-33. <http://dx.doi.org/10.1056/NEJM198801213180301>.
62. Corrado D, Leoni L, Link MS, Della Bella P, Gaita F, Curnis A, Salerno JU, Ilgidbashian D, Raviele A, Disertori M, Zanotto G, Verlato R, Vergara G, Delise P, Turrini P, Basso C, Naccarella F, Maddalena F, Estes NA 3rd, Buja G, Thiene G. Implantable cardioverter-defibrillator therapy for prevention of sudden death in patients with arrhythmogenic right

ventricular cardiomyopathy/dysplasia. *Circulation*. 2003;108:3084–91. <http://dx.doi.org/10.1161/01.CIR.0000103130.33451.D2>.

63. Wichter T, Paul M, Wollmann C, Acil T, Gerdes P, Ashraf O, Tjan TD, Soeparwata R, Block M, Borggreffe M, Scheld HH, Breithardt G, Böcker D. Implantable cardioverter/defibrillator therapy in arrhythmogenic right ventricular cardiomyopathy: single-center experience of long-term follow-up and complications in 60 patients.

Circulation. 2004;109:1503–8. <http://dx.doi.org/10.1161/01.CIR.0000121738.88273.43>.

64. Corrado D, Calkins H, Link MS, Leoni L, Favale S, Bevilacqua M, Basso C, Ward D, Boriani G, Ricci R, Piccini JP, Dalal D, Santini M, Buja G, Iliceto S, Estes NA 3rd, Wichter T, McKenna WJ, Thiene G, Marcus FI. Prophylactic implantable defibrillator in patients with arrhythmogenic right ventricular cardiomyopathy/dysplasia and no prior ventricular fibrillation or sustained ventricular tachycardia.

Circulation. 2010;122:1144–52. <http://dx.doi.org/10.1161/CIRCULATIONAHA.109.913871>.

KEY WORDS ACC/AHA Scientific Statements, athletes, arrhythmogenic right ventricular cardiomyopathy, cardiomyopathy, cardiovascular abnormalities, cardiovascular magnetic resonance, hypertrophic cardiomyopathy, myocarditis, sudden death

AHA/ACC SCIENTIFIC STATEMENT

Eligibility and Disqualification Recommendations for Competitive Athletes With Cardiovascular Abnormalities: Task Force 4: Congenital Heart Disease

A Scientific Statement From the American Heart Association and American College of Cardiology

George F. Van Hare, MD, FACC,
*Chair**

Michael J. Ackerman, MD, PhD,
 FACC*

Juli-anne K. Evangelista, DNP,
 APRN, CPNP-AC, FACC*

Richard J. Kovacs, MD, FAHA, FACC*
 Robert J. Myerburg, MD, FACC*

Keri M. Shafer, MD*

Carole A. Warnes, MD, FACC*

Reginald L. Washington, MD, FAHA*

Congenital heart disease (CHD) is the most common form of serious birth defect, occurring in 8 per 1,000 live births (1). The past several decades have seen dramatic improvements in survival with palliative or corrective heart surgery, such that there are now more adult patients than pediatric patients alive with CHD. Although restriction from competitive athletics may well be indicated for some, the great majority of patients can and should engage in some form of physical activity and should avoid a sedentary lifestyle. Clinicians should encourage their patients

to engage in healthy physical activities, bearing in mind specific features in some patients, such as residual obstruction, pulmonary vascular disease, low systemic ventricular function, and preexisting arrhythmias in the presence of implanted cardiac rhythm devices such as pacemakers and implantable cardioverter-defibrillators. In addition, the physiological effects of athletic activities at high altitude should be considered for patients with elevated pulmonary vascular resistance. These issues are covered elsewhere in this document. Fortunately,

*On behalf of the American Heart Association Electrocardiography and Arrhythmias Committee of the Council on Clinical Cardiology, Council on Cardiovascular Disease in the Young, Council on Cardiovascular and Stroke Nursing, Council on Functional Genomics and Translational Biology, and the American College of Cardiology.

The American Heart Association and the American College of Cardiology make every effort to avoid any actual or potential conflicts of interest that may arise as a result of an outside relationship or a personal, professional, or business interest of a member of the writing panel. Specifically, all members of the writing group are required to complete and submit a Disclosure Questionnaire showing all such relationships that might be perceived as real or potential conflicts of interest. The Preamble and other Task Force reports for these proceedings are available online at www.onlinejacc.org (J Am Coll Cardiol 2015;XX:000-000; 000-000; 000-000; 000-000; 000-000; 000-000; 000-000; 000-000; 000-000; 000-000; 000-000; 000-000; 000-000; 000-000; 000-000; 000-000; 000-000; 000-000; 000-000).

This statement was approved by the American Heart Association Science Advisory and Coordinating Committee on June 24, 2015, and the American Heart Association Executive Committee on July 22, 2015, and by the American College of Cardiology Board of Trustees and Executive Committee on June 3, 2015.

The American College of Cardiology requests that this document be cited as follows: Van Hare GF, Ackerman MJ, Evangelista JK, Kovacs RJ, Myerburg RJ, Shafer KM, Warnes CA, Washington RL; on behalf of the American Heart Association Electrocardiography and Arrhythmias Committee of the Council on Clinical Cardiology, Council on Cardiovascular Disease in the Young, Council on Cardiovascular and Stroke Nursing, Council on Functional Genomics and Translational Biology, and the American College of Cardiology. Eligibility and disqualification recommendations for competitive athletes with cardiovascular abnormalities: Task Force 4: congenital heart disease: a scientific statement from the American Heart Association and American College of Cardiology. J Am Coll Cardiol 2015;xx:●●●-●●●.

This article has been copublished in *Circulation*.

Copies: This document is available on the World Wide Web sites of the American Heart Association (<http://my.americanheart.org>) and the American College of Cardiology (www.acc.org). For copies of this document, please contact Elsevier Inc. Reprint Department via fax (212-633-3820) or e-mail (reprints@elsevier.com).

Permissions: Multiple copies, modification, alteration, enhancement, and/or distribution of this document are not permitted without the express permission of the American College of Cardiology. Requests may be completed online via the Elsevier site (<http://www.elsevier.com/about/policies/author-agreement/obtaining-permission>).

although repaired CHD is clearly associated with the development of arrhythmias such as atrial flutter and ventricular tachycardia, exercise does not appear to contribute to the risk.

The level of sports participation recommended includes consideration of both the training and the competitive aspects of the activity but must be individualized to the particular patient, taking into account the patient's functional status and history of surgery. Noninvasive testing, such as formal exercise testing, Holter monitoring, echocardiography, and cardiac magnetic resonance imaging studies, is also often useful.

TYPES OF CONGENITAL DEFECTS

Simple Shunting Lesions (Atrial Septal Defect, Ventricular Septal Defect, Patent Ductus Arteriosus), Treated and Untreated

Of the 8 most common subtypes of CHD, ventricular septal defect (VSD; 34%), atrial septal defect (ASD; 13%), and patent ductus arteriosus (PDA; 10%), respectively, are the most common (2). With rare exceptions, patients with hemodynamically insignificant CHD such as VSD, ASD, and PDA may participate competitively in all sports. There are no demonstrative data that children with hemodynamically insignificant VSD (open or after closure), ASD (open or after closure), or PDA (open or after closure) require exercise limitations or that these lesions are related to acknowledged episodes of sudden cardiac death (SCD) (3,4). Patients with associated pulmonary hypertension secondary to the above-mentioned lesions that is hemodynamically significant can develop acute symptoms, including reduced exercise capacity or, more importantly, arrhythmias, syncope, chest pain, or sudden death (5,6). For the purposes of this document, pulmonary hypertension is defined as a mean pulmonary artery pressure >25 mm Hg or a pulmonary vascular resistance index of >3 Wood units.

Patients with right-to-left shunting may become more cyanotic during exercise, at least in part because of changes in the ratio of systemic vascular resistance to pulmonary vascular resistance, which can result in increased hypoxemia. Therefore, full clinical assessment, including laboratory and exercise testing, should be considered before any physical activity, because this population represents a very high risk of sudden death (6). Additional precautions should be taken when these patients are exercising at altitude, because the pulmonary vascular resistance generally rises, thus increasing the degree of hypoxemia and cardiac workload.

Children with open or surgically closed VSDs have a normal exercise capacity despite a mild chronotropic

limitation in the latter. Some data suggest that aerobic capacity is reduced in patients with open or closed VSDs, as well as in patients with closed ASDs. Abnormal right ventricular (RV) and pulmonary pressure can also occur in those with isolated VSDs; however, these findings did not impact these exercise recommendations or identify any episodes of SCD (7).

ASD: Untreated

Recommendations

1. It is recommended that athletes with small defects (<6 mm), normal right-sided heart volume, and no pulmonary hypertension should be allowed to participate in all sports (*Class I; Level of Evidence C*).
2. It is recommended that athletes with a large ASD and no pulmonary hypertension should be allowed to participate in all sports (*Class I; Level of Evidence C*).
3. Athletes with an ASD and pulmonary hypertension may be considered for participation in low-intensity class IA sports (*Class I; Level of Evidence C*).
4. Athletes with associated pulmonary vascular obstructive disease who have cyanosis and a large right-to-left shunt should be restricted from participation in all competitive sports, with the possible exception of class IA sports (*Class III; Level of Evidence C*).

ASD: After Surgical Repair or Closure by Interventional Catheterization

Recommendations

1. Three to 6 months after operation or intervention, athletes without pulmonary hypertension, myocardial dysfunction, or arrhythmias may participate in all sports (*Class I; Level of Evidence C*).
2. After operation or intervention, patients with pulmonary hypertension, arrhythmias, or myocardial dysfunction may be considered for participation in low-intensity class IA sports (*Class IIb; Level of Evidence C*).

VSD: Untreated

Recommendations

1. An athlete with a small or restrictive VSD with normal heart size and no pulmonary hypertension can participate in all sports (*Class I; Level of Evidence C*).
2. An athlete with a large, hemodynamically significant VSD and pulmonary hypertension may consider participation in only low-intensity class IA sports (*Class IIb; Level of Evidence C*).

VSD: After Surgical Repair or Closure by
Interventional Catheterization

Recommendations

1. At 3 to 6 months after repair, asymptomatic athletes with no or a small residual defect and no evidence of pulmonary hypertension, ventricular or atrial tachyarrhythmia, or myocardial dysfunction can participate in all competitive sports (*Class I; Level of Evidence C*).
2. Athletes with persistent pulmonary hypertension should be allowed to participate in class IA sports only (*Class I; Level of Evidence B*).
3. Athletes with symptomatic atrial or ventricular tachyarrhythmias or second- or third-degree atrioventricular block should not participate in competitive sports until further evaluation by an electrophysiologist (*Class III; Level of Evidence C*).
4. Athletes with mild to moderate pulmonary hypertension or ventricular dysfunction should not participate in competitive sports, with the possible exception of low-intensity class IA sports (*Class III; Level of Evidence C*).

PDA: Untreated

Recommendations

1. Athletes with a small PDA, normal pulmonary artery pressure, and normal left-sided heart chamber dimension can participate in all competitive sports (*Class I; Level of Evidence C*).
2. Athletes with a moderate or large PDA and persistent pulmonary hypertension should be allowed to participate in class IA sports only (*Class I; Level of Evidence B*).
3. Athletes with a moderate or large PDA that causes left ventricular (LV) enlargement should not participate in competitive sports until surgical or interventional catheterization closure (*Class III; Level of Evidence C*).

PDA: Treated (After Surgical Repair or Closure by
Interventional Catheterization)

Recommendations

1. After recovery from catheter or surgical PDA closure, athletes with no evidence of pulmonary hypertension can participate in all competitive sports (*Class I; Level of Evidence C*).
2. Athletes with residual pulmonary artery hypertension should be restricted from participation in all competitive sports, with the possible exception of class IA sports (*Class I; Level of Evidence B*).

Pulmonary Valve Stenosis: Treated and Untreated

Mild valvar pulmonary stenosis (PS) is characterized by a systolic ejection murmur, a systolic ejection click that

varies with respiration, and a normal ECG. Decisions are based on estimated severity by use of Doppler-derived peak instantaneous gradients. A gradient <40 mm Hg indicates mild PS, 40 to 60 mm Hg indicates moderate PS, and >60 mm Hg indicates severe PS. Treatment can be by surgery or more commonly by balloon valvuloplasty. Adequate relief means a resolution of symptoms or a reduction in gradient to <40 mm Hg.

Recommendations

1. Athletes with mild PS and normal RV function can participate in all competitive sports. Annual reevaluation is also recommended (*Class I; Level of Evidence B*).
2. Athletes treated by operation or balloon valvuloplasty who have achieved adequate relief of PS (gradient <40 mm Hg by Doppler) can participate in all competitive sports (*Class I; Level of Evidence B*).
3. Athletes with moderate or severe PS can consider participation only in low-intensity class IA and IB sports (*Class IIb; Level of Evidence B*).
4. Athletes with severe pulmonary insufficiency as demonstrated by marked RV enlargement can consider participation in low-intensity class IA and IB sports (*Class IIb; Level of Evidence B*).

Aortic Valve Stenosis: Treated and Untreated

Assessment of fully grown athletes with aortic stenosis (AS) is discussed in the Task Force 5 report on valvular heart disease (8). The following discussion pertains to recommendations in children and adolescents. Patients with AS are differentiated between those with mild, moderate, and severe AS by physical examination, ECG, and Doppler echocardiography. In all cases, regardless of the degree of stenosis, patients with a history of fatigue, light-headedness, dizziness, syncope, chest pain, or pallor on exercise deserve a full evaluation. Annual reevaluation is required for all patients with AS, because the disease can progress. Patients with severe AS are at risk of sudden death, particularly with exercise (9).

Mild AS is defined as a mean Doppler gradient of <25 mm Hg or a peak instantaneous Doppler gradient <40 mm Hg. On evaluation, patients should have a normal ECG, normal exercise tolerance, and no history of exercise-related chest pain, syncope, or atrial or ventricular tachyarrhythmia. Moderate AS is defined as a mean Doppler gradient of 25 to 40 mm Hg or a peak instantaneous Doppler gradient of 40 to 70 mm Hg. Patients should have only mild or no LV hypertrophy by echocardiogram and an absence of LV strain pattern on ECG, as well as a normal maximum exercise stress test without

evidence of ischemia or tachyarrhythmia, with normal exercise duration and blood pressure response. Severe AS is defined as a mean Doppler gradient >40 mm Hg or a peak instantaneous Doppler gradient >70 mm Hg. Such patients may have symptoms such as exercise intolerance, chest pain, near-syncope, or syncope and likely will have LV hypertrophy with strain on ECG, as well as an abnormal blood pressure response to exercise. For cases in which symptoms or findings on ECG or exercise test appear more severe than expected for the estimated severity by Doppler, cardiac catheterization may be indicated.

Treatment may be by surgery or balloon aortic valvuloplasty. After treatment, patients may be left with residual valve gradient, aortic insufficiency, or both and may experience recurrence or progression, and thus, continued clinical follow-up is needed.

Recommendations

1. Athletes with mild AS can participate in all competitive sports (*Class I; Level of Evidence B*).
2. Athletes with severe AS can participate only in low-intensity class IA sports (*Class I; Level of Evidence B*).
3. Athletes with moderate AS may be considered for participation in low static or low to moderate dynamic sports (class IA, IB, and IIA) (*Class IIb; Level of Evidence B*).
4. Athletes with severe AS should be restricted from all competitive sports, with the possible exception of low-intensity (class IA) sports (*Class III; Level of Evidence B*).

AS After Surgery or Balloon Dilation

Recommendations

1. Athletes with residual AS may be considered for participation in sports according to the above recommendations based on severity (*Class IIb; Level of Evidence C*).
2. Athletes with significant (moderate or severe) aortic valve insufficiency may participate in sports according to the recommendation of Task Force 5 in this document (8).

Coarctation of the Aorta: Treated and Untreated

Coarctation may be discrete or in the form of a long segment and causes hypertension in the upper limbs and hypotension in the lower limbs. The severity is determined by a clinical examination that includes the arm/leg pressure gradient, exercise testing, echocardiographic studies, and magnetic resonance imaging. Coarctation is often considered part of a more general aortopathy with a medial abnormality, particularly when associated with

a bicuspid aortic valve. This renders the aorta more vulnerable to dilation, aneurysm formation, and dissection and rupture. There is a recognized association with cerebral aneurysms. Virtually all patients, except those with mild coarctation, will undergo intervention, in the form of either surgical repair or percutaneous balloon angioplasty and stenting.

Even after successful surgical repair or stent placement, residual abnormalities may persist. These include residual coarctation and aneurysm formation at the site of repair or stent. Because of the aortopathy, the ascending aorta may also dilate and even dissect and rupture. Systemic hypertension may persist and if not present at rest may also occur on exercise. Some patients may have residual LV hypertrophy, and many may have residual aortic valve disease when a concomitant bicuspid aortic valve is present. Lifetime follow-up is mandatory, and the potential for premature coronary artery disease has been reported.

Before a decision is made regarding exercise participation, a detailed evaluation should be conducted, which should include a physical examination, ECG, chest radiograph, exercise testing transthoracic echocardiographic evaluation of the aortic valve and aorta, and either magnetic resonance imaging or computed tomography angiography. Normal standards exist for peak systolic blood pressure on exercise testing, by age and sex (10,11). Magnetic resonance imaging or computed tomography imaging should be performed to evaluate the thoracic aorta in its entirety, because transthoracic echocardiographic imaging alone will not visualize the entire aorta, and both residual coarctation and aneurysm may be missed.

Coarctation of the Aorta: Untreated

Recommendations

1. Athletes with coarctation and without significant ascending aortic dilation (z score ≤ 3.0 ; a score of 3.0 equals 3 standard deviations from the mean for patient size) with a normal exercise test and a resting systolic blood pressure gradient <20 mm Hg between the upper and lower limbs and a peak systolic blood pressure not exceeding the 95th percentile of predicted with exercise can participate in all competitive sports (*Class I; Level of Evidence C*).
2. Athletes with a systolic blood pressure arm/leg gradient >20 mm Hg or exercise-induced hypertension (a peak systolic blood pressure exceeding the 95th percentile of predicted with exercise) or with significant ascending aortic dilation (z score >3.0) may be considered for participation only in low-intensity class IA sports (*Class IIb; Level of Evidence C*).

Coarctation of the Aorta: Treated by Surgery or Balloon and Stent

Recommendations

1. Athletes who are >3 months past surgical repair or stent placement with <20 mm Hg arm/leg blood pressure gradient at rest, as well as (1) a normal exercise test with no significant dilation of the ascending aorta (z score <3.0), (2) no aneurysm at the site of coarctation intervention, and (3) no significant concomitant aortic valve disease, may be considered for participation in competitive sports, but with the exception of high-intensity static exercise (classes IIIA, IIIB, and IIIC), as well as sports that pose a danger of bodily collision (*Class IIb; Level of Evidence C*).
2. Athletes with evidence of significant aortic dilation or aneurysm formation (not yet at a size to need surgical repair) may be considered for participation only in low-intensity (classes IA and IB) sports (*Class IIb; Level of Evidence C*).

Elevated Pulmonary Vascular Resistance in CHD

Patients with pulmonary vascular disease and CHD are at risk of sudden death during sports activity. In those with shunts (commonly septal defects or complex CHD), cyanosis is usually present at rest (Eisenmenger syndrome) and worsens with exercise. Most of these patients self-limit their activity, and they should not participate in competitive sports, with the exception of low-intensity (class IA) sports. The benefits of a regular exercise program, however, including improved walk distance, peak oxygen consumption, quality of life, and functional class, have been demonstrated, and thus, physical activity that does not require maximal effort should be encouraged. This usually comprises physical activity that allows the patient to speak a sentence comfortably (the “talk test”), and 6-minute walk tests will facilitate guidance in this regard.

Patients with suspected residual pulmonary hypertension who have undergone prior surgical repair or catheter intervention for shunt lesions should have a complete hemodynamic evaluation by cardiac catheterization before engaging in competitive athletics. Pulmonary arterial hypertension is usually defined as a mean pulmonary artery pressure of >25 mm Hg and a pulmonary arteriolar resistance >3 Wood units. Decisions proscribing exercise for patients with mild degrees of pulmonary hypertension are quite arbitrary, and no evidence-based scientific data exist. Similarly, no data exist with regard to appropriate exercise prescriptions for patients with mild and moderate pulmonary hypertension, which emphasizes the need to collect prospective data.

Patients and families should be cautioned, however, concerning the potential effect of high altitude on the existing abnormal cardiopulmonary physiology, because this may lead to important further elevations in pulmonary vascular resistance in such patients, with adverse effects.

Recommendations

1. Patients with mean pulmonary artery pressure of <25 mm Hg can participate in all competitive sports (*Class I; Level of Evidence B*).
2. Patients with moderate or severe pulmonary hypertension, with a mean pulmonary artery pressure >25 mm Hg, should be restricted from all competitive sports, with the possible exception of low-intensity (class IA) sports. Complete evaluation and exercise prescription (physician guidance on exercise training) should be obtained before athletic participation (*Class III; Level of Evidence B*).

Ventricular Dysfunction After CHD Surgery

It is not unusual for a patient to present with significant ventricular dysfunction early or late after surgery for CHD, and this dysfunction, of course, affects exercise performance. Assessment of ventricular function is more straightforward for patients with systemic LVs than for those with systemic RVs, but the use of cardiac magnetic resonance imaging has improved the assessment of RV function (12). In general, throughout this document, severe ventricular dysfunction is defined as an ejection fraction (EF) <40%, moderate dysfunction as EF 40% to 50%, and normal as EF >50%. It should be recognized that these definitions are somewhat arbitrary. Of course, the other characteristics of the patient's heart disease and repair should be considered as well, such as valvar stenosis and insufficiency.

Recommendations

1. Before participation in competitive sports, all athletes with ventricular dysfunction after CHD surgery should undergo evaluation that includes clinical assessment, ECG, imaging assessment of ventricular function, and exercise testing (*Class I; Level of Evidence B*).
2. Athletes with normal or near-normal systemic ventricular function (EF ≥50%) can participate in all sports (*Class I; Level of Evidence B*).
3. It is reasonable for athletes with mildly diminished ventricular function (EF 40%–50%) to participate in low- and medium-intensity static and dynamic sports (classes IA, IB, and IIA and IIB) (*Class IIb; Level of Evidence B*).

4. Athletes with moderately to severely diminished ventricular function (EF <40%) should be restricted from all competitive sports, with the possible exception of low-intensity (class IA) sports (Class III; Level of Evidence B).

Cyanotic CHD, Including Tetralogy of Fallot

Cyanotic Heart Disease: Unoperated or With Palliative Shunts

Patients with congenital defects resulting in chronic cyanosis can reach adolescence and adulthood but have significantly diminished exercise tolerance, which correlates with clinical outcomes (13-15). Iron deficiency further exacerbates exercise intolerance, whereas select treatments may improve exercise capacity in this population (16,17). Cardiopulmonary exercise testing shows that significant desaturation occurs in these patients with exercise, with performance and symptoms related to underlying anatomy (14,18), including those with palliative shunts, because of changes in the balance between pulmonary and systemic vascular resistance. Full clinical assessment, including laboratory and exercise testing, should be considered before any physical activity, because this population represents a very high risk of sudden death (19). Additional caution should be taken when these patients are exercising at altitude ("Elevated Pulmonary Vascular Resistance in CHD"). Unfortunately, data to address the safety of participation in competitive sports in this population are lacking.

Recommendations

- 1. In athletes with unrepaired cyanotic heart disease, a complete evaluation is recommended, which should involve exercise testing. An exercise prescription based on clinical status and underlying anatomy should be obtained before athletic participation (Class I; Level of Evidence C).**
- 2. Athletes with unrepaired cyanotic heart disease who are clinically stable and without clinical symptoms of heart failure may be considered for participation in only low-intensity class IA sports (Class IIb; Level of Evidence C).**

Postoperative Tetralogy of Fallot

Most patients with tetralogy of Fallot currently undergo initial repair in the first 2 years of life but often develop clinically significant pulmonary valve dysfunction in adolescence or adulthood. Clinical evaluation of patients before participation in competitive sports should include assessment of pulmonary valve function and assessment of factors associated with increased risk of sudden death in this population (15,19-21). In particular, attention should be paid to

careful assessment of LV function (13,22). Exercise testing is recommended to evaluate ability to augment cardiovascular function during increasing exercise intensity and for evidence of exercise-related ECG changes suggestive of arrhythmia or ischemia. Given its prognostic utility, cardiopulmonary exercise testing should be considered to fully evaluate patients before sports participation, particularly those with evidence of residual lesions on physical examination or imaging assessment (13,15). We strongly caution against participation in high-intensity competitive sports for those with severe biventricular dysfunction, atrial or ventricular arrhythmias, and significant abnormalities on exercise testing or abnormal hemodynamic assessment. Evaluation of lung function with pulmonary function tests may also be useful to assess for evidence of underlying disease and optimization before sports participation (23). For participation in moderate- and high-intensity sports, the patient should be asymptomatic at rest and with exercise, as well as free (or relatively free) of risk factors associated with sudden death, although individualized assessment is key for assessment of additional anatomic anomalies such as anomalous coronary arteries or residual outflow tract obstruction. Specific data regarding safety of long-term high-intensity exercise are needed in tetralogy of Fallot patients with preserved ventricular function with moderate to severe regurgitation, because a blunted stroke-volume response with high-intensity exercise has been reported in this population. One could extrapolate from these data that exercise performance in class III sports would be limited, although there is insufficient evidence to understand cardiovascular risk for these athletes (24). Given this, we recommend serial clinical evaluation with assessment of ventricular function during the period of sports participation.

Recommendations

- 1. Before participation in competitive sports, it is recommended that all athletes with repaired tetralogy of Fallot should undergo evaluation, including clinical assessment, ECG, imaging assessment of ventricular function, and exercise testing (Class I; Level of Evidence B).**
- 2. Athletes without significant ventricular dysfunction (EF >50%), arrhythmias, or outflow tract obstruction may be considered for participation in moderate- to high-intensity sports (class II to III). To meet these criteria, the athlete must be able to complete an exercise test without evidence of exercise-induced arrhythmias, hypotension, ischemia, or other concerning clinical symptoms (Class IIb; Level of Evidence B).**

3. Athletes with severe ventricular dysfunction (EF <40%), severe outflow tract obstruction, or recurrent or uncontrolled atrial or ventricular arrhythmias should be restricted from all competitive sports, with the possible exception of low-intensity (class IA) sports (Class III; Level of Evidence B).

Transposition of the Great Arteries: After Atrial Switch (Mustard or Senning Operation)

The atrial switch procedure was reported in 1959 and was performed frequently for transposition of the great arteries (TGA) from approximately the 1960s to the 1990s. Thus, the significant majority of patients with this anatomy are adults, because survival into the third and fourth decades occurs in most patients. Exercise tolerance is diminished in this population and correlates with clinical outcomes (13,15). Recent studies show this population may be at higher risk of sudden death than other CHD populations (19,20). The strongest predictors of sudden death are the presence of prior arrhythmia and severe systemic ventricular dysfunction, although prior VSD, age at repair, QRS duration, and heart failure symptoms may also be associated with an increased risk (19,25-29). The population with TGA with atrial switch likely has a unique response to exercise given reports that a high proportion of sudden death events occur during exertion (28). This adds complexity to the evaluation before sports participation, because the pathophysiology and prevention strategies for SCD in this population are not well understood. Unfortunately, evidence of exercise-induced arrhythmias on routine clinical testing has not been shown to reliably predict exercise-induced SCD events (28). Thus, careful evaluation of clinical status with special attention to clinical history of arrhythmias, patency and structure of the venous baffles, systemic ventricular function, coronary artery anatomy, and presence of additional obstructive lesions (e.g., PS) is recommended. Severe systemic ventricular function is defined as an EF <40%. Clinical evaluation should include cardiopulmonary exercise testing with continuous oximetry before sports participation. Restriction from high-intensity activities should be considered in the presence of severe systemic ventricular dysfunction, persistent arrhythmias, hypoxia, or inability to increase cardiac output, blood pressure, or heart rate with exertion. In the absence of these findings, moderate-intensity sports participation may be safe (30). However, the effect of long-term exercise training on the systemic RV is not known. Therefore, we recommend serial clinical evaluation during the period of sports participation, with assessment of ventricular function to evaluate the medium- and long-term effects of exercise participation.

Evaluation for and optimization of pulmonary dysfunction are recommended (31).

Recommendations

1. It is recommended that before participation in competitive sports, all athletes who have undergone the Senning and Mustard procedure should undergo an evaluation that includes clinical assessment, ECG, imaging assessment of ventricular function, and exercise testing (Class I; Level of Evidence B).
2. Participation in competitive sports in those athletes with a history of clinically significant arrhythmias or severe ventricular dysfunction may be considered on an individual basis based on clinical stability (Class IIb; Level of Evidence C).
3. Athletes without clinically significant arrhythmias, ventricular dysfunction, exercise intolerance, or exercise-induced ischemia may be considered for participation in low- and moderate-intensity competitive sports (classes IA, IB, IIA, and IIB) (Class IIb; Level of Evidence C).
4. Athletes with severe clinical systemic RV dysfunction, severe RV outflow tract obstruction, or recurrent or uncontrolled atrial or ventricular arrhythmias should be restricted from all competitive sports, with the possible exception of low-intensity (class IA) sports. (Class III; Level of Evidence C).

Congenitally Corrected TGA

Patients with congenitally corrected TGA (CCTGA) are often diagnosed in childhood, usually in the presence of additional defects, including PS, VSD, or systemic atrioventricular valve abnormalities (see appropriate sections for additional recommendations). In CCTGA, exercise tolerance is limited, and both exercise tolerance and ventricular function are predictive of adverse outcomes (13,15,32,33). Systemic atrioventricular valve dysfunction is not uncommon in this population and correlates with exercise performance (33). In a recent study, patients with CCTGA and additional defects were found to have a particularly high rate of sudden death (19). However, because of the small number of patients with this anatomy, it is difficult to determine the risk factors for this outcome, although systemic ventricular dysfunction and arrhythmias may correlate with these events. When evaluating patients before competitive sports participation, we recommend assessment of clinical stability with noninvasive imaging and cardiopulmonary exercise testing. Clinical assessment should include evaluation of systemic ventricular and atrioventricular valve function and coronary artery anatomy, as well as exclusion of outflow tract obstruction. One small study found that participation of patients with CCTGA in a 3-month

exercise training program of moderate to high intensity was not associated with clinical decline (30); however, the effect of long-term exercise training on the systemic RV is not known. Therefore, we recommend serial clinical evaluation during the period of sports participation, with assessment of ventricular function to evaluate the medium- and long-term effects of exercise participation.

Limited data are available to assess the risks associated with sports participation in those who have had a double-switch procedure that resulted in the redirection of pulmonary venous blood to the LV and aorta. However, assessment of the venous baffle and Rastelli or arterial switch integrity is required before consideration of sports participation.

Recommendations

1. It is recommended that before participation in competitive sports, all CCTGA athletes should undergo evaluation that includes clinical assessment, ECG, imaging assessment of ventricular function, and exercise testing (*Class I; Level of Evidence B*).
2. Participation in competitive sports in those CCTGA athletes with a history of clinically significant arrhythmias or severe ventricular dysfunction may be considered on an individual basis based on clinical stability (*Class IIb; Level of Evidence C*).
3. Athletes with CCTGA and without clinically significant arrhythmias, ventricular dysfunction, exercise intolerance, or exercise-induced ischemia may be considered for participation in low- and moderate-intensity competitive sports (class IA and IB) (*Class IIb; Level of Evidence C*).
4. Asymptomatic athletes with CCTGA and without abnormalities on clinical evaluation may be considered for participation in moderate- to high-intensity competitive sports (classes II and IIIB or IIIC) (*Class IIb; Level of Evidence C*).
5. Athletes with severe clinical systemic RV dysfunction, severe RV outflow tract obstruction, or recurrent or uncontrolled atrial or ventricular arrhythmias should be restricted from all competitive sports, with the possible exception of low-intensity (class IA) sports (*Class III; Level of Evidence C*).

TGA, After Arterial Switch Procedure

Significant numbers of patients have now undergone the arterial switch procedure over the past 3 decades, and thus, many are at an age when sports participation is desired. Coronary stenosis or obstruction is fortunately rare, and concerns are mainly focused on the possibility of supraventricular PS at the site of anastomosis, which is rarely significant. Patients with symptoms such as

syncope or exertional chest pain should have a careful assessment of their coronary artery status, because sudden death has been reported late after arterial switch repair (34). Exercise studies are not particularly sensitive in this group of patients, and coronary angiography or other modalities such as computed tomography angiography may be necessary in those with significant symptoms (35). The issue of surveillance of asymptomatic patients after the arterial switch procedure is controversial.

Recommendations

1. It is recommended that before participation in competitive sports, athletes who have undergone the arterial switch procedure for TGA should undergo evaluation that includes clinical assessment, ECG, imaging assessment of ventricular function, and exercise testing (*Class I; Level of Evidence B*).
2. It is reasonable for athletes with no cardiac symptoms, normal ventricular function, and no tachyarrhythmias after the arterial switch procedure for TGA to participate in all competitive sports (*Class IIb; Level of Evidence C*).
3. After the arterial switch procedure for TGA, athletes with more than mild hemodynamic abnormalities or ventricular dysfunction may be considered for participation in low and moderate static/low dynamic competitive sports (classes IA, IB, IC, and IIA), provided that exercise testing is normal (*Class IIb; Level of Evidence C*).
4. After the arterial switch procedure for TGA, athletes with evidence of coronary ischemia should be restricted from all competitive sports, with the possible exception of low-intensity (class IA) sports (*Class III; Level of Evidence B*).

Fontan Procedure

The Fontan operation, a complete redirection of systemic venous blood to the pulmonary arteries, is performed to palliate single-ventricle physiology. Patients with this circulation have significantly decreased exercise performance, and they are able to increase cardiac output during exercise through unique mechanisms (13,36,37). Limitation to exercise performance is multifactorial and correlates with morbidity and mortality (36,38). When patients are evaluated before sports participation, it is imperative to recognize that both the Fontan circulation and the underlying cardiac anatomy can be extremely variable among patients. As a result, thorough clinical assessment is recommended before sports participation. This clinical assessment should include evaluation for risk factors associated with sudden death (20,38,39). Additionally, comprehensive

cardiac imaging is recommended, as well as cardiopulmonary exercise testing with continuous oximetry. If there is significant exercise intolerance during a maximal effort test, as evidenced by such things as an inability to increase blood pressure or heart rate, systemic desaturation, or development of arrhythmias or other symptomatic limitations, the healthcare provider should strongly consider restriction from participation in moderate- and high-intensity competitive sports and training. If the recommended evaluation is unremarkable, participation in moderate-intensity and moderate-duration exercise can be considered. This recommendation is based on small studies that have shown evidence of improvement in some measures of fitness without evidence of clinical deterioration in those participating in moderate-intensity exercise training and resistance training (40,41). However, the safety of participation in high-intensity or high-duration sports is unknown. Additionally, evaluation and optimization of lung function before sports participation is recommended (42). All Fontan patients requiring chronic anticoagulation should be restricted from participation in contact sports.

Recommendations

- 1. It is recommended that before participation in competitive sports, all athletes who have undergone the Fontan procedure should undergo an evaluation that includes clinical assessment, ECG, imaging assessment of ventricular function, and exercise testing (Class I; Level of Evidence B).**
- 2. Athletes who have undergone the Fontan procedure and who have no symptomatic heart failure or significantly abnormal intravascular hemodynamics can participate only in low-intensity class IA sports (Class I; Level of Evidence C).**
- 3. Participation in other sports may be considered on an individual basis with regard for the athlete's ability to complete an exercise test without evidence of exercise-induced arrhythmias, hypotension, ischemia, or other concerning clinical symptoms (Class IIb; Level of Evidence C).**

Ebstein Anomaly of the Tricuspid Valve

The phenotypic spectrum of this malformation is extreme, ranging from minimal to profound tricuspid regurgitation and right-sided heart enlargement. If there is an atrial shunt, cyanosis may be present. A minority of patients with Ebstein anomaly will have preexcitation that could precipitate clinically important and symptomatic arrhythmias. Physical disability and increased risk for sudden death with exercise have been reported with severe cases. Risk stratification for exercise-related

arrhythmias remains imprecise for this anomaly. In patients for whom there is also evidence of Wolff-Parkinson-White syndrome or in whom a defibrillator has been implanted, the recommendations found in Task Force 9 (43) should be respected as well. Note that the recommendations below apply both before and after surgical plication and are based on the degree of valve regurgitation and existence of arrhythmias.

Recommendations

- 1. Patients with mild to moderate Ebstein anomaly (i.e., no cyanosis, normal RV size, tricuspid regurgitation that is moderate or less, and no evidence of atrial or ventricular arrhythmias) can be considered for participation in all sports (Class IIb; Level of Evidence C).**
- 2. Patients with Ebstein anomaly with severe tricuspid regurgitation but without evidence of arrhythmias on ambulatory electrocardiographic monitoring (except isolated premature contractions) may be considered for participation only in low-intensity class IA sports (Class IIb; Level of Evidence C).**

Congenital Coronary Anomalies

Anomalies of coronary arteries are second in frequency among identified structural causes of SCD in competitive athletes, accounting for ≈17% of such deaths in the United States (44). Anomalous origins of coronary arteries from the wrong sinus of Valsalva or from the pulmonary artery are estimated to be present in ≈1% of the overall population (45) but are proportionately far more common in athletes who die suddenly, as cited above. Although the vast majority of sudden deaths associated with coronary anomalies occur during or shortly after exercise (46), sudden death has been reported in the sedentary state (47).

The most common anomalous origin is the right coronary artery originating from the left sinus of Valsalva, but among athletes who have died suddenly, anomalous origin of the left main or left anterior descending coronary artery from the right sinus of Valsalva is far more prevalent. Furthermore, SCDs are most strongly associated with the pattern in which the anomalous left coronary artery passes between the aorta and main pulmonary artery. An anomalous origin of a coronary artery from the pulmonary artery is far less commonly observed in athletes who die suddenly and in fact often presents with myocardial infarction in infancy or early childhood. Nonetheless, some cases are not recognized until adolescence or adulthood and may be associated with sudden death in athletes, albeit rarely. Nonspecific electrocardiographic findings may be observed in adolescents with otherwise unrecognized anomalous coronary arteries arising from the pulmonary artery.

The ECG is an unreliable screening tool for suspecting or recognizing anomalous origin of coronary arteries before an event, and even stress tests are not uniformly positive among people with these anomalies (48). Clinical symptoms, such as exertional chest discomfort or dyspnea, may be helpful, but 2 reports suggest that 50% of SCDs associated with coronary artery anomalies were first events without prior symptoms (46,49). The best methods for identifying the anomaly include coronary angiography, computed tomography angiography, and magnetic resonance angiography. Although not uniformly successful, athletes undergoing echocardiographic studies for any reason should have careful attempts to identify the origins of the coronary arteries.

Surgical procedures are the only therapies available for correcting these anomalies (50), with return to intense athletic activities permitted after 3 months after the procedure with demonstration of the absence of ischemia on postoperative stress testing (51).

Recommendations

1. Athletes with anomalous origin of a coronary artery from the pulmonary artery can participate only in low-intensity class IA sports, whether or not they have had a prior myocardial infarction, and pending repair of the anomaly (*Class I; Level of Evidence C*).
2. Athletes with an anomalous origin of a right coronary artery from the left sinus of Valsalva should be evaluated by an exercise stress test. For those without either symptoms or a positive exercise stress test, permission to compete can be considered after adequate counseling of the athlete and/or the athlete's parents (in the case of a minor) as to risk and benefit, taking into consideration the uncertainty of accuracy of a negative stress test (*Class IIa; Level of Evidence C*).
3. After successful surgical repair of an anomalous origin from the wrong sinus, athletes may consider participation in all sports 3 months after surgery if the patient remains free of symptoms and an exercise stress test shows no evidence of ischemia or cardiac arrhythmias (*Class IIb; Level of Evidence C*).
4. After repair of anomalous origin of a coronary artery from the pulmonary artery, decisions regarding exercise restriction may be based on presence of sequelae such as myocardial infarction or ventricular dysfunction (*Class IIb; Level of Evidence C*).
5. Athletes with an anomalous origin of a left coronary artery from the right sinus of Valsalva, especially when the artery passes between the pulmonary artery and aorta, should be restricted from participation in all competitive sports, with the possible exception of class IA sports, before surgical repair. This recommendation applies whether the anomaly is identified as a consequence of symptoms or discovered incidentally (*Class III; Level of Evidence B*).
6. Nonoperated athletes with an anomalous origin of a right coronary artery from the left sinus of Valsalva who exhibit symptoms, arrhythmias, or signs of ischemia on exercise stress test should be restricted from participation in all competitive sports, with the possible exception of class IA sports, before a surgical repair (*Class III; Level of Evidence C*).

DISCLOSURES

Writing Group Disclosures

Writing Group Member	Employment	Research Grant	Other Research Support	Speakers Bureau/Honoraria	Expert Witness	Ownership Interest	Consultant/Advisory Board	Other
George F. Van Hare	Washington University	None	None	None	None	None	None	None
Michael J. Ackerman	Mayo Clinic	NIH (RO1 grants) [†]	None	None	None	None	Boston Scientific*; Gilead Sciences*; Medtronic*; St. Jude Medical*	Transgenomic [†]
Juli-anne K. Evangelista	Boston Children's Hospital	None	None	None	None	None	None	None
Richard J. Kovacs	Indiana University	None	None	None	None	None	None	None
Robert J. Myerburg	University of Miami	None	None	None	None	None	None	None
Keri M. Shafer	Boston Children's Hospital	None	None	None	None	None	None	None
Carole A. Warnes	Mayo Clinic-Rochester	None	None	None	None	None	None	None
Reginald L. Washington	Rocky Mountain Hospital for Children	None	None	None	None	None	None	None

This table represents the relationships of writing group members that may be perceived as actual or reasonably perceived conflicts of interest as reported on the Disclosure Questionnaire, which all members of the writing group are required to complete and submit. A relationship is considered to be "significant" if (a) the person receives \$10,000 or more during any 12-month period, or 5% or more of the person's gross income; or (b) the person owns 5% or more of the voting stock or share of the entity, or owns \$10,000 or more of the fair market value of the entity. A relationship is considered to be "modest" if it is less than "significant" under the preceding definition.

*Modest.

[†]Significant.

Reviewer Disclosures

Reviewer	Employment	Research Grant	Other Research Support	Speakers Bureau/Honoraria	Expert Witness	Ownership Interest	Consultant/Advisory Board	Other
Emile Bacha	New York Presbyterian Hospital-Columbia University College of Physicians and Surgeons	None	None	None	None	None	None	None
Julian I.E. Hoffman	University of California	None	None	None	None	None	None	None
Robert D.B. Jaquiss	Duke University School of Medicine	None	None	None	None	None	None	None
Silvana M. Lawrence	Baylor College of Medicine	None	None	None	None	None	None	Vice President, Science and Research, Championship Hearts Foundation [†]
John Moore	UCSD	None	None	None	None	None	None	None
Karen K. Stout	University of Washington	None	None	None	None	None	None	None

This table represents the relationships of reviewers that may be perceived as actual or reasonably perceived conflicts of interest as reported on the Disclosure Questionnaire, which all reviewers are required to complete and submit. A relationship is considered to be "significant" if (a) the person receives \$10,000 or more during any 12-month period, or 5% or more of the person's gross income; or (b) the person owns 5% or more of the voting stock or share of the entity, or owns \$10,000 or more of the fair market value of the entity. A relationship is considered to be "modest" if it is less than "significant" under the preceding definition.

[†]Significant.

REFERENCES

1. Dolk H, Loane MA, Abramsky L, de Walle H, Garne E. Birth prevalence of congenital heart disease. *Epidemiology*. 2010;21:275-7. <http://dx.doi.org/10.1097/EDE.0b013e3181c2979b>.
2. van der Linde D, Konings EE, Slager MA, Witsenburg M, Helbing WA, Takkenberg JJ, Roos-Hesselink JW. Birth prevalence of congenital heart disease worldwide: a systematic review and meta-analysis. *J Am Coll Cardiol*. 2011;58:2241-7. <http://dx.doi.org/10.1016/j.jacc.2011.08.025>.
3. Maron BJ, Zipes DP. Introduction: eligibility recommendations for competitive athletes with cardiovascular abnormalities-general considerations. *J Am Coll Cardiol*. 2005;45:1318-21. <http://dx.doi.org/10.1016/j.jacc.2005.02.006>.
4. Hirth A, Reybrouck T, Bjarnason-Wehrens B, Lawrenz W, Hoffmann A. Recommendations for participation in competitive and leisure sports in patients with congenital heart disease: a consensus document. *Eur J Cardiovasc Prev Rehabil*. 2006;13:293-9.
5. Kobayashi Y, Nakanishi N, Kosakai Y. Pre- and postoperative exercise capacity associated with hemodynamics in adult patients with atrial septal defect: a retrospective study. *Eur J Cardiothorac Surg*. 1997;11:1062-6.
6. Zipes DP, Wellens HJ. Sudden cardiac death. *Circulation*. 1998;98:2334-51.
7. Möller T, Brun H, Fredriksen PM, Holmstrøm H, Peersen K, Pettersen E, Grünig E, Mereles D, Thaulow E. Right ventricular systolic pressure response during exercise in adolescents born with atrial or ventricular septal defect. *Am J Cardiol*. 2010;105:1610-6. <http://dx.doi.org/10.1016/j.amjcard.2010.01.024>.
8. Bonow RO, Nishimura RA, Thompson PD, Udelson JE, on behalf of the American Heart Association Electrocardiography and Arrhythmias Committee of the Council on Clinical Cardiology, Council on Cardiovascular Disease in the Young, Council on Cardiovascular and Stroke Nursing, Council on Functional Genomics and Translational Biology, and the American College of Cardiology. Eligibility and disqualification recommendations for competitive athletes with cardiovascular abnormalities: Task Force 5: valvular heart disease: a scientific statement from the American Heart Association and American College of Cardiology. *J Am Coll Cardiol*. 2015 In Press. <http://dx.doi.org/10.1016/j.jacc.2015.09.037>.
9. Driscoll DJ, Edwards WD. Sudden unexpected death in children and adolescents. *J Am Coll Cardiol*. 1985;5 suppl:118B-21B.
10. Daida H, Allison TG, Squires RW, Miller TD, Gau GT. Peak exercise blood pressure stratified by age and gender in apparently healthy subjects. *Mayo Clin Proc*. 1996;71:445-52. [http://dx.doi.org/10.1016/S0025-6196\(11\)64085-8](http://dx.doi.org/10.1016/S0025-6196(11)64085-8).
11. Le VV, Mitiku T, Sungar G, Myers J, Froelicher V. The blood pressure response to dynamic exercise testing: a systematic review. *Prog Cardiovasc Dis*. 2008;51:135-60. <http://dx.doi.org/10.1016/j.pcad.2008.07.001>.
12. Blalock SE, Banka P, Geva T, Powell AJ, Zhou J, Prakash A. Interstudy variability in cardiac magnetic resonance imaging measurements of ventricular volume, mass, and ejection fraction in repaired tetralogy of Fallot: a prospective observational study. *J Magn Reson Imaging*. 2013;38:829-35. <http://dx.doi.org/10.1002/jmri.24050>.
13. Kempny A, Dimopoulos K, Uebing A, Moceri P, Swan L, Gatzoulis MA, Diller GP. Reference values for exercise limitations among adults with congenital heart disease: relation to activities of daily life: single centre experience and review of published data. *Eur Heart J*. 2012;33:1386-96. <http://dx.doi.org/10.1093/eurheartj/ehr461>.
14. Bruaene AVD, Meester PD, Voigt JU, Delcroix M, Pasquet A, Backer JD, Pauw MD, Naeije R, Vachiéry JL, Paelinck BP, Morissens M, Budts W. Worsening in oxygen saturation and exercise capacity predict adverse outcome in patients with Eisenmenger syndrome. *Int J Cardiol*. 2013;168:1386-92.
15. Inuzuka R, Diller GP, Borgia F, Benson L, Tay EL, Alonso-Gonzalez R, Silva M, Charalambides M, Swan L, Dimopoulos K, Gatzoulis MA. Comprehensive use of cardiopulmonary exercise testing identifies adults with congenital heart disease at increased mortality risk in the medium term. *Circulation*. 2012;125:250-9. <http://dx.doi.org/10.1161/CIRCULATIONAHA.111.058719>.
16. Galiè N, Beghetti M, Gatzoulis MA, Granton J, Berger RM, Lauer A, Chiossi E, Landzberg M, for the Bosentan Randomized Trial of Endothelin Antagonist Therapy-5 (BREATHE-5) Investigators. Bosentan therapy in patients with Eisenmenger syndrome: a multicenter, double-blind, randomized, placebo-controlled study. *Circulation*. 2006;114:48-54. <http://dx.doi.org/10.1161/CIRCULATIONAHA.106.630715>.
17. Tay EL, Peset A, Papaphylactou M, Inuzuka R, Alonso-Gonzalez R, Giannakoulas G, Tzifa A, Goletto S, Broberg C, Dimopoulos K, Gatzoulis MA. Replacement therapy for iron deficiency improves exercise capacity and quality of life in patients with cyanotic congenital heart disease and/or the Eisenmenger syndrome. *Int J Cardiol*. 2011;151:307-12. <http://dx.doi.org/10.1016/j.ijcard.2010.05.066>.
18. Müller J, Hess J, Hager A. Exercise performance and quality of life is more impaired in Eisenmenger syndrome than in complex cyanotic congenital heart disease with pulmonary stenosis. *Int J Cardiol*. 2011;150:177-81. <http://dx.doi.org/10.1016/j.ijcard.2010.04.005>.
19. Koyak Z, Harris L, de Groot JR, Silversides CK, Oechslin EN, Bouma BJ, Budts W, Zwiderman AH, Van Gelder IC, Mulder BJ. Sudden cardiac death in adult congenital heart disease. *Circulation*. 2012;126:1944-54. <http://dx.doi.org/10.1161/CIRCULATIONAHA.112.104786>.
20. Gallego P, Gonzalez AE, Sanchez-Recalde A, Peinado R, Polo L, Gomez-Rubin C, Lopez-Sendon JL, Oliver JM. Incidence and predictors of sudden cardiac arrest in adults with congenital heart defects repaired before adult life. *Am J Cardiol*. 2012;110:109-17. <http://dx.doi.org/10.1016/j.amjcard.2012.02.057>.
21. Chiu SN, Wu MH, Su MJ, Wang JK, Lin MT, Chang CC, Hsu HW, Shen CT, Thériault O, Chahine M. Coexisting mutations/polymorphisms of the long QT syndrome genes in patients with repaired tetralogy of Fallot are associated with the risks of life-threatening events. *Hum Genet*. 2012;131:1295-304. <http://dx.doi.org/10.1007/s00439-012-1156-4>.
22. Khairy P, Aboulhosn J, Gurvitz MZ, Opatowsky AR, Mongeon FP, Kay J, Valente AM, Earing MG, Lui G, Gersony DR, Cook S, Ting JG, Nickolaus MJ, Webb G, Landzberg MJ, Broberg CS, for the Alliance for Adult Research in Congenital Cardiology (AARCC). Arrhythmia burden in adults with surgically repaired tetralogy of Fallot: a multi-institutional study. *Circulation*. 2010;122:868-75. <http://dx.doi.org/10.1161/CIRCULATIONAHA.109.928481>.
23. Alonso-Gonzalez R, Borgia F, Diller GP, Inuzuka R, Kempny A, Martinez-Naharro A, Tutarel O, Marino P, Wustmann K, Charalambides M, Silva M, Swan L, Dimopoulos K, Gatzoulis MA. Abnormal lung function in adults with congenital heart disease: prevalence, relation to cardiac anatomy, and association with survival. *Circulation*. 2013;127:882-90. <http://dx.doi.org/10.1161/CIRCULATIONAHA.112.126755>.
24. Marcuccio E, Arora G, Quivers E, Yurchak MK, McCaffrey F. Noninvasive measurement of cardiac output during exercise in children with tetralogy of Fallot. *Pediatr Cardiol*. 2012;33:1165-70. <http://dx.doi.org/10.1007/s00246-012-0276-x>.
25. Lange R, Hörer J, Kostolny M, Cleuziou J, Vogt M, Busch R, Holper K, Meisner H, Hess J, Schreiber C. Presence of a ventricular septal defect and the Mustard operation are risk factors for late mortality after the atrial switch operation: thirty years of follow-up in 417 patients at a single center. *Circulation*. 2006;114:1905-13. <http://dx.doi.org/10.1161/CIRCULATIONAHA.105.606046>.
26. Roubertie F, Thambo JB, Bretonneau A, Iriart X, Laborde N, Baudet E, Roques X. Late outcome of 132 Senning procedures after 20 years of follow-up. *Ann Thorac Surg*. 2011;92:2206-13. <http://dx.doi.org/10.1016/j.athoracsurg.2011.06.024>.
27. Schwerzmann M, Salehian O, Harris L, Siu SC, Williams WG, Webb GD, Colman JM, Redington A, Silversides CK. Ventricular arrhythmias and sudden death in adults after a Mustard operation for transposition of the great arteries. *Eur Heart J*. 2009;30:1873-9. <http://dx.doi.org/10.1093/eurheartj/ehp179>.
28. Kammeraad JA, van Deuren CH, Sreeram N, Bink-Boelkens MT, Ottenkamp J, Helbing WA, Lam J, Sobotka-Plojhar MA, Daniels O, Balaji S. Predictors of sudden cardiac death after Mustard or Senning repair for transposition of the great arteries. *J Am Coll Cardiol*. 2004;44:1095-102. <http://dx.doi.org/10.1016/j.jacc.2004.05.073>.
29. Warnes CA. Transposition of the great arteries. *Circulation*. 2006;114:2699-709. <http://dx.doi.org/10.1161/CIRCULATIONAHA.105.592352>.
30. Winter MM, van der Bom T, de Vries LC, Balducci A, Bouma BJ, Pieper PG, van Dijk AP, van der Plas MN, Picchio FM, Mulder BJ. Exercise training improves exercise capacity in adult patients with a systemic right ventricle: a randomized clinical trial. *Eur Heart J*. 2012;33:1378-85. <http://dx.doi.org/10.1093/eurheartj/ehs396>.
31. Sterrett LE, Ebenroth ES, Montgomery GS, Schamberger MS, Hurwitz RA. Pulmonary limitation to exercise after repair of D-transposition of the great vessels: atrial baffle versus arterial switch. *Pediatr*

Cardiol. 2011;32:910-6. <http://dx.doi.org/10.1007/s00246-011-0013-x>.

32. Graham TP Jr., Bernard YD, Mellen BG, Celermajer D, Baumgartner H, Cetta F, Connolly HM, Davidson WR, Dellborg M, Foster E, Gersony WM, Gessner IH, Hurwitz RA, Kaemmerer H, Kugler JD, Murphy DJ, Noonan JA, Morris C, Perloff JK, Sanders SP, Sutherland JL. Long-term outcome in congenitally corrected transposition of the great arteries: a multi-institutional study. *J Am Coll Cardiol*. 2000;36:255-61.

33. Grewal J, Crean A, Garceau P, Wald R, Woo A, Rakowski H, Silversides CK. Subaortic right ventricular characteristics and relationship to exercise capacity in congenitally corrected transposition of the great arteries. *J Am Soc Echocardiogr*. 2012;25:1215-21. <http://dx.doi.org/10.1016/j.echo.2012.08.014>.

34. Gatlin S, Kalynych A, Sallee D, Campbell R. Detection of a coronary artery anomaly after a sudden cardiac arrest in a 17 year-old with D-transposition of the great arteries status post arterial switch operation: a case report. *Congenit Heart Dis*. 2011;6:384-8. <http://dx.doi.org/10.1111/j.1747-0803.2011.00491.x>.

35. Legendre A, Losay J, Touchot-Kone A, Serraf A, Belli E, Piot JD, Lambert V, Capderou A, Planche C. Coronary events after arterial switch operation for transposition of the great arteries. *Circulation*. 2003;108 suppl 1:II186-90.

36. Fernandes SM, Alexander ME, Graham DA, Khairy P, Clair M, Rodriguez E, Pearson DD, Landzberg MJ, Rhodes J. Exercise testing identifies patients at increased risk for morbidity and mortality following Fontan surgery. *Congenit Heart Dis*. 2011;6:294-303. <http://dx.doi.org/10.1111/j.1747-0803.2011.00500.x>.

37. Shafer KM, Garcia JA, Babb TG, Fixler DE, Ayers CR, Levine BD. The importance of the muscle and ventilatory blood pumps during exercise in patients without a subpulmonary ventricle (Fontan operation). *J Am Coll Cardiol*. 2012;60:2115-21. <http://dx.doi.org/10.1016/j.jacc.2012.08.970>.

38. Diller GP, Giardini A, Dimopoulos K, Gargiulo G, Müller J, Derrick G, Giannakoulas G, Khambadkone S, Lammers AE, Picchio FM, Gatzoulis MA, Hager A. Predictors of morbidity and mortality in contemporary Fontan patients: results from a multicenter study

including cardiopulmonary exercise testing in 321 patients. *Eur Heart J*. 2010;31:3073-83. <http://dx.doi.org/10.1093/eurheartj/ehq356>.

39. Khairy P, Fernandes SM, Mayer JE Jr., Friedman JK, Walsh EP, Lock JE, Landzberg MJ. Long-term survival, modes of death, and predictors of mortality in patients with Fontan surgery. *Circulation*. 2008;117:85-92. <http://dx.doi.org/10.1161/CIRCULATIONAHA.107.738559>.

40. Brassard P, Bédard E, Jobin J, Rodés-Cabau J, Poirier P. Exercise capacity and impact of exercise training in patients after a Fontan procedure: a review. *Can J Cardiol*. 2006;22:489-95.

41. Cordina RL, O'Meagher S, Karmali A, Rae CL, Liess C, Kemp GJ, Puranik R, Singh N, Celermajer DS. Resistance training improves cardiac output, exercise capacity and tolerance to positive airway pressure in Fontan physiology. *Int J Cardiol*. 2013;168:780-8. <http://dx.doi.org/10.1016/j.ijcard.2012.10.012>.

42. Matthews IL, Fredriksen PM, Bjørnstad PG, Thaulow E, Gronn M. Reduced pulmonary function in children with the Fontan circulation affects their exercise capacity. *Cardiol Young*. 2006;16:261-7. <http://dx.doi.org/10.1017/S1047951106000345>.

43. Zipes DP, Link MS, Ackerman MJ, Kovacs RJ, Myerburg RJ, Estes NAM 3rd, on behalf of the American Heart Association Electrocardiography and Arrhythmias Committee of the Council on Clinical Cardiology, Council on Cardiovascular Disease in the Young, Council on Cardiovascular and Stroke Nursing, Council on Functional Genomics and Translational Biology, and the American College of Cardiology. Eligibility and disqualification recommendations for competitive athletes with cardiovascular abnormalities: Task Force 9: arrhythmias and conduction defects: a scientific statement from the American Heart Association and American College of Cardiology. *J Am Coll Cardiol*. 2008;52:1890-947. <http://dx.doi.org/10.1016/j.jacc.2008.10.002>.

44. Maron BJ, Doerer JJ, Haas TS, Tierney DM, Mueller FO. Sudden deaths in young competitive athletes: analysis of 1866 deaths in the United States, 1980-2006. *Circulation*. 2009;119:1085-92. <http://dx.doi.org/10.1161/CIRCULATIONAHA.108.804617>.

45. Tuo G, Marasini M, Brunelli C, Zannini L, Balbi M. Incidence and clinical relevance of primary congenital

anomalies of the coronary arteries in children and adults. *Cardiol Young*. 2012;1-6.

46. 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.

47. De Rosa G, Piastra M, Pardeo M, Caresta E, Capelli A. Exercise-unrelated sudden death as the first event of anomalous origin of the left coronary artery from the right aortic sinus. *J Emerg Med*. 2005;29:437-41. <http://dx.doi.org/10.1016/j.jemermed.2005.07.001>.

48. Edwards CP, Yavari A, Sheppard MN, Sharma S. Anomalous coronary origin: the challenge in preventing exercise-related sudden cardiac death. *Br J Sports Med*. 2010;44:895-7. <http://dx.doi.org/10.1136/bjsm.2008.054387>.

49. Frommelt PC. Congenital coronary artery abnormalities predisposing to sudden cardiac death. *Pacing Clin Electrophysiol*. 2009;32 suppl 2:S63-6. <http://dx.doi.org/10.1111/j.1540-8159.2009.02387.x>.

50. Warnes CA, Williams RG, Bashore TM, Child JS, Connolly HM, Dearani JA, del Nido P, Fasules JW, Graham TP Jr., Hijazi ZM, Hunt SA, King ME, Landzberg MJ, Miner PD, Radford MJ, Walsh EP, Webb GD. ACC/AHA 2008 guidelines for the management of adults with congenital heart disease: a report of the American College of Cardiology/American Heart Association Task Force on Practice Guidelines (Writing Committee to Develop Guidelines on the Management of Adults With Congenital Heart Disease). *J Am Coll Cardiol*. 2015 In Press. <http://dx.doi.org/10.1016/j.jacc.2015.09.037>.

51. Graham TP Jr., Driscoll DJ, Gersony WM, Newburger JW, Rocchini A, Towbin JA. Task Force 2: congenital heart disease. *J Am Coll Cardiol*. 2005;45:1326-33. <http://dx.doi.org/10.1016/j.jacc.2005.02.009>.

KEY WORDS ACC/AHA Scientific Statements, athletes, cardiovascular abnormalities, congenital heart disease, coronary vessel anomalies, Fontan procedure, transposition of great arteries

A Scientific Statement From the American Heart Association and American College of Cardiology

James E. Udelson, MD, FAHA, FACC*

Permissions: Multiple copies, modification, alteration, enhancement, and/or distribution of this document are not permitted without the express permission of the American College of Cardiology. Requests may be completed online via the Elsevier site (<http://www.elsevier.com/about/policies/author-agreement/obtaining-permission>).

normal left ventricular (LV) systolic function. Stage C designates asymptomatic patients with severe valvular heart disease with evidence of preserved systolic function (stage C1) or LV dysfunction (C2), and stage D designates patients with symptomatic severe valvular heart disease with or without LV dysfunction. Eligibility for competitive sports is a pertinent issue for people with valvular heart disease in stages A, B, and C, whereas symptomatic patients in stage D are not candidates for competition and under most circumstances should be referred for valve replacement or repair. Athletic competition is also a relevant issue in asymptomatic patients who have undergone successful valve surgery.

AORTIC VALVE DISEASE

Aortic valve disease is usually caused by degenerative changes in a bicuspid or tricuspid aortic valve. Calcification of trileaflet aortic valves is an increasingly common cause of aortic stenosis (AS) in middle-aged and elderly people because of increased longevity in the United States and other developed nations. Bicuspid aortic valves occur in 1.5% to 2.0% of the population and thus are common findings in young athletes (2). In developed countries, there is a very low incidence of rheumatic aortic valve disease, but this pathogenesis predominates in athletes from developing nations. Additional causes of outflow obstruction can be nonvalvular and include subvalvular and supra-valvular AS, both produced by cardiomyopathies and congenital abnormalities in the left ventricle and ascending aorta, as discussed in other sections of this document. Primary diseases of the aorta are common causes of aortic valve regurgitation (AR) (1), and this pathogenesis should be considered in athletes presenting with AR.

Aortic Stenosis

AS is a well-known cause of exertion-related sudden cardiac death but is responsible for <4% of sudden deaths in young athletes (3). The severity of AS is best evaluated with the combination of the history, physical examination, and Doppler echocardiography. A history of decreasing exercise tolerance, exertional dyspnea, or exercise-induced angina in an athlete with a systolic murmur should raise the possibility of severe AS. A decreased volume and delayed upstroke of the carotid pulse, as well as a greater intensity and duration of the systolic murmur, also suggest clinically important AS.

Assessment of congenital AS in children and adolescents and recommendations for participation in athletics in these age groups are discussed in the Task Force 4 report (4) on congenital heart disease in this document. The following discussion pertains to recommendations in fully grown athletes in late adolescence and adulthood.

Doppler echocardiography is the standard method to assess AS (1), and its severity is graded as shown in Table 1.

Clinicians should combine features of the history, physical examination, and echocardiogram in evaluating the severity of AS, as well as integrating the various echocardiographic measures of jet velocity, mean gradient, and calculated valve area, because each has limitations. In young patients with abnormal aortic valves, it is also important to assess the size and morphology of the ascending aorta to exclude concomitant aortopathy, as discussed below. Athletes with mild or moderate AS (stage B) should be evaluated yearly, because the valve can narrow progressively. Exercise testing with electrocardiographic and blood pressure monitoring is useful in evaluating ostensibly asymptomatic athletes with AS because it may reveal unexpectedly low exercise tolerance, exercise hypotension, or electrocardiographic abnormalities that may alter the exercise recommendations. Doppler echocardiography can underestimate the severity of the aortic valve gradient, so further evaluation is warranted in athletes with Doppler evidence of mild or moderate AS who have symptoms or LV hypertrophy.

Evaluation

Athletes with bicuspid aortic valves without stenosis (stage A) should undergo yearly physical examinations for detection of new onset of heart murmurs. Athletes with mild to moderate AS (stage B) should have a yearly history, physical examination, and Doppler echocardiogram to evaluate disease severity. Exercise testing should be performed in athletes with mild and moderate AS to ensure that their effort tolerance is commensurate with the proposed athletic activity and that they do not develop exercise hypotension or electrocardiographic evidence of ischemia.

Recommendations

1. Athletes with AS should be evaluated yearly to determine whether sports participation can continue (Class I; Level of Evidence C).

TABLE 1 Severity of Aortic Stenosis by Doppler Echocardiography

Severity	Jet Velocity, m/s	Mean Gradient, mm Hg	Aortic Valve Area, cm ²
Mild	<3	<20	>1.5
Moderate	3–4	20–40	1–1.5
Severe	>4	>40	<1.0

Reprinted from Nishimura *et al.* (1). Copyright © 2014, American Heart Association, Inc.

2. Athletes with mild AS (stage B) and a normal maximal exercise response can participate in all sports (Class IIa; Level of Evidence C).
3. Athletes with moderate AS (stage B) can 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 and the training regimen demonstrates satisfactory exercise capacity without symptoms, ST-segment depression, or ventricular tachyarrhythmias, and with a normal blood pressure response (Class IIa; Level of Evidence C).
4. Asymptomatic athletes with severe AS (stage C) should not participate in competitive sports, with the possible exception of low-intensity (class IA) sports (Class III; Level of Evidence C).
5. Symptomatic patients with AS (stage D) should not participate in competitive sports (Class III; Level of Evidence C).

Aortic Regurgitation

The common causes of chronic AR include bicuspid aortic valve disease, congenital connective tissues disorders such as Marfan syndrome, rheumatic heart disease, and idiopathic or hypertensive dilation of the ascending aorta (1). Chronic AR is usually asymptomatic and well tolerated for years, but when severe, it produces a gradual increase in LV dimensions. The diagnosis during the asymptomatic stages of AR (stages B and C) is suggested on physical examination by a wide arterial pulse pressure, a diastolic murmur heard along the sternal border, or a systolic outflow murmur related to the increased forward stroke volume. Doppler echocardiography is useful in confirming the diagnosis and grading the severity of AR (1,5). AR produces both pressure and volume loading of the LV but is usually well tolerated for decades, with normal LV systolic performance despite the increased LV volume until the LV cannot tolerate further increases in the volume overload.

It is often difficult to differentiate the LV dilatation produced in athletes by exercise training from the dilatation produced by chronic severe AR in its early and advanced stages. Therefore, assessment of LV enlargement in highly trained athletes with known or suspected AR must take this issue into consideration. Progressively severe AR can result in LV volumes that exceed the normal physiological responses to athletic training, but there is overlap in LV volume encountered in normal athletes and patients with AR. Up to 45% of trained male athletes have LV end-diastolic dimension (LVEDD) >55 mm (6,7), but only 14% of even elite male athletes have LVEDD >60 mm, and LVEDD rarely exceeds 70 mm (6,7). LVEDD >55 mm occurs in <10% of elite women athletes

and is >60 mm in only 1% (8). Hence, athletes with severe AR and LVEDD exceeding these values have a high likelihood that severe AR is contributing to the LV dilation and should be evaluated carefully for decreasing exercise tolerance and absence of ventricular augmentation with exercise. Similarly, LV end-systolic dimension (LVESD) may also be increased with athletic training. Among elite athletes, the upper limit of LVESD is 49 mm for men and 38 mm for women (7). It may be helpful to normalize LVEDD and LVESD for body size (9), because larger athletes have larger ventricular volumes. Data indexed for body surface area and height in athletes are available for LVEDD (6,8) but not LVESD. The reported upper limit of LVEDD indexed for body surface area is 35.3 mm/m² for men and 40.8 mm/m² for women (8). Values for LVEDD and LVESD for elite athletes as reported by Pelliccia et al (7,8) are summarized in Table 2.

The LV ejection fraction response to exercise is also maintained in patients with chronic AR until there is severe LV dilation (10). An ejection fraction <50% at rest in an athlete with severe AR indicates LV decompensation. Serial assessment of the LVESD is valuable in assessing the progressive effects of severe AR in those with normal LV ejection fractions. In patients with severe AR, the 2014 American Heart Association/American College of Cardiology "Guideline for the Management of Patients With Valvular Heart Disease" (1) defines preserved systolic function (stage C1) as LV ejection fraction ≥50% and LVESD ≤50 mm or indexed LVESD ≤25 mL/m².

Evaluation

Athletes with AR should undergo a yearly history and physical examination with Doppler echocardiography. Exercise testing to at least the level of activity achieved in competition and the training regimen is helpful in confirming asymptomatic status and assessing blood pressure responses. The usefulness of assessing LV function with exercise in athletes has not been established. Patients with AR often have underlying bicuspid aortic valves. In these patients, it is important to also assess the morphology of the aortic root and ascending aorta to rule

TABLE 2 Left Ventricular Dimensions in Elite Athletes

	Men		Women	
	Mean±SD	Upper Limit	Mean±SD	Upper Limit
LVESD, mm*	38.2±3.2	49	32.9±2.9	38
LVEDD, mm*	58.8±3.4	70	52.2±3.2	60
LVEDD, mm†	54.2±2.0	66	48.9±3.8	66
LVEDD/BSA, mm/m ² †	28.1±2.3	35.3	29.8±2.5	40.8
LVEDD/height, mm/m†	30.1±2.1	36.8	29.3±1.9	35.9

BSA indicates body surface area; LVEDD, left ventricular end-diastolic dimension; and LVESD, left ventricular end-systolic dimension.

*Data from 114 Olympic athletes (89 men, 25 women) (7).

†Data from 1338 elite athletes (738 men, 600 women) (8).

out associated aortopathy. Recommendations for sports participation for athletes with bicuspid aortic valves and dilated aortas are provided in the Task Force 7 recommendations of this report (11).

Recommendations

- 1. Athletes with AR should be evaluated annually to determine whether sports participation can continue (Class I; Level of Evidence C).**
- 2. Exercise testing to at least the level of activity achieved in competition and the training regimen is helpful in confirming asymptomatic status in athletes with AR and assessing blood pressure responses (Class I; Level of Evidence C).**
- 3. Athletes with mild to moderate degrees of AR (stage B) with normal LV ejection fraction and no or mild LV dilatation can participate in all competitive sports if they have normal exercise tolerance on exercise testing (Class I; Level of Evidence C).**
- 4. Athletes with mild to moderate degrees of AR with normal LV ejection fraction and moderate LV dilatation (LVESD <50 mm [men], <40 mm [women], or <25 mm/m² [either sex]) can reasonably participate in all competitive sports if they have normal exercise tolerance on exercise testing (Class IIa; Level of Evidence C).**
- 5. It may be reasonable for athletes with severe AR, LV ejection fraction ≥50% (stage C1), and LVESD <50 mm (men), <40 mm (women), or <25 mm/m² (either sex) to participate in all competitive sports if they have normal exercise tolerance, and Doppler echocardiography indicates no progression of AR severity or severity of LV dilatation (Class IIb; Level of Evidence C).**
- 6. It may be reasonable for athletes with AR and aortic dimensions of 41 to 45 mm to participate in sports with low risk of bodily contact (Class IIb; Level of Evidence C).**
- 7. Athletes with severe AR and symptoms (stage D), LV systolic dysfunction with ejection fraction <50% (stage C2), LVESD >50 mm or >25 mm/m² (stage C2), or severe increase in LVEDD (>70 mm or ≥35.3 mm/m² [men], >65 mm or ≥40.8 mm/m² [women]) should not participate in competitive sports (Class III; Level of Evidence C).**

Bicuspid Aortic Valves

Bicuspid aortic valve is present in 1% to 2% of the population (1,2) and is the marker of connective tissue abnormalities that affect both the aortic valve and aorta. That patients with bicuspid aortic valves are at increased risk for AS and AR is well known, but these patients are also at increased risk for aortic enlargement and aortic

dissection, although the absolute risk for these events is quite small (1,12), and it is not known whether restriction of physical activity limits the risk or the rate of aortic enlargement or dissection.

Evaluation

Patients with bicuspid aortic valves should undergo echocardiography to evaluate both aortic valve function and the size of the aortic sinuses and ascending aorta. The Task Force 7 report (11) in this document contains recommendations for sports participation in athletes with bicuspid aortic valves and dilated aortas.

MITRAL VALVE DISEASE

Mitral Stenosis

The pathogenesis of mitral stenosis (MS) is almost always rheumatic. Most patients with significant MS will be sufficiently symptomatic during exercise that participation in competitive sports is not an issue, but patients with mild to moderate MS may be asymptomatic even with strenuous exercise. MS rarely causes sudden death; however, 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 (13). 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, nor is the effect of even periodic strenuous exercise on the likelihood of developing atrial fibrillation. When atrial fibrillation occurs, even patients with mild MS must receive anticoagulation therapy. The above 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, which occurs most commonly in the presence of atrial fibrillation, but there is no evidence that this potential complication is provoked by strenuous exercise.

Clues regarding the hemodynamic severity of MS may often be obtained from the history and physical examination, but accurate noninvasive assessment of severity requires 2-dimensional and Doppler echocardiography in the majority of patients. MS is categorized as severe when the mitral valve area is <1.5 cm², which corresponds to a mean transmitral gradient of 5 to 10 mm Hg at normal resting heart rates (1). The mean pressure gradient is highly dependent on the transvalvular flow and diastolic filling period and will vary greatly with increases in heart rate during exercise. A mean transmitral gradient >15 mm Hg or pulmonary artery wedge pressure >25 mm Hg during exercise is indicative or significant MS.

Evaluation

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 that approximates 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 (1).

Recommendations

- 1. Athletes with MS should be evaluated annually to determine whether sports participation can continue (Class I; Level of Evidence C).**
- 2. Exercise testing to at least the level of activity achieved in competition and the training regimen is useful in confirming asymptomatic status in patients with MS (Class I; Level of Evidence C).**
- 3. It is reasonable for athletes with mild MS (mitral valve area >2.0 cm², mean gradient <10 mm Hg at rest) in sinus rhythm to participate in all competitive sports (Class IIa; Level of Evidence C).**
- 4. Athletes with severe MS (mitral valve area <1.5 cm²) in either sinus rhythm or atrial fibrillation should not participate in competitive sports, with the possible exception of low-intensity (class IA) sports (Class III; Level of Evidence C).**
- 5. Patients with MS of any severity who are in atrial fibrillation or have a history of atrial fibrillation, who must receive anticoagulation therapy, should not engage in any competitive sports involving the risk of bodily contact (Class III; Level of Evidence C).**

Mitral Regurgitation

Mitral regurgitation (MR) has a variety of possible causes, the most common of which in an athletic population is mitral valve prolapse (myxomatous mitral valve disease). Other common causes are rheumatic heart disease, infective endocarditis, and connective tissue diseases (such as Marfan syndrome). Secondary forms of MR can develop in patients with coronary artery disease and dilated cardiomyopathy because of tethering of the mitral leaflets and restricted leaflet closure. The recommendations outlined in this section are for athletes with primary valvular MR rather than MR secondary to coronary artery disease or other conditions that cause LV dilation or systolic dysfunction.

MR is detected by the characteristic systolic murmur, confirmed and quantified by Doppler echocardiography (1,5). The severity of the MR is related to the magnitude

of the regurgitant volume, which results in LV dilation and increases in left atrial pressure and volume. The majority of people with mild or moderate MR are asymptomatic (stage B). 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, such as ejection fraction, tend to overestimate true myocardial performance (14). For purposes of this discussion, LV systolic dysfunction in subjects with MR is defined as LV ejection fraction $<60\%$ or LVESD >40 mm (1). As with AR, the distinction between LV dilation caused by athletic training versus that caused by severe MR is difficult when the LVEDD is <60 mm (or <40 mm/m²). However, LVEDD measurements >60 mm strongly suggest the presence of severe MR and perhaps the need for surgical mitral valve repair and thus warrant further investigation.

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 minute) or blood pressure with exercise may manifest marked increases in regurgitant volume and pulmonary capillary pressures.

Evaluation

Athletes with MR should undergo yearly physical examinations, Doppler echocardiograms, and exercise stress testing to at least the level of activity that approximates the exercise demands of the sport. 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, particularly in athletes with greater severity of MR (1). 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, and thus, the recommendations below should be tempered in patients with these mechanisms of MR.

Recommendations

- 1. Athletes with MR should be evaluated annually to determine whether sports participation can continue (Class I; Level of Evidence C).**
- 2. Exercise testing to at least the level of activity achieved in competition and the training regimen is useful in confirming asymptomatic status in patients with MR (Class I; Level of Evidence C).**

3. Athletes with mild to moderate MR who are in sinus rhythm with normal LV size and function and with normal pulmonary artery pressures (stage B) can participate in all competitive sports (*Class I; Level of Evidence C*).
4. It is reasonable for athletes with 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 [LVEDD <60 mm or <35 mm/m² in men or <40 mm/m² in women]) to participate in all competitive sports (stage B) (*Class IIa; Level of Evidence C*).
5. Athletes with severe 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 [LVEDD <60 mm or <35.3 mm/m² in men or <40 mm/m² in women]) can participate in low-intensity and some moderate-intensity sports (classes IA, IIA, and IB) (stage C1) (*Class IIb; Level of Evidence C*).
6. Athletes with MR and definite LV enlargement (LVEDD ≥65 mm or ≥35.3 mm/m² [men] or ≥40 mm/m² [women]), pulmonary hypertension, or any degree of LV systolic dysfunction at rest (LV ejection fraction <60% or LVESD >40 mm) should not participate in any competitive sports, with the possible exception of low-intensity class IA sports (*Class III; Level of Evidence C*).
7. Athletes with a history of atrial fibrillation who are receiving long-term anticoagulation should not engage in sports involving any risk of bodily contact (*Class III; Level of Evidence C*).

ATHLETIC PARTICIPATION AFTER CARDIAC VALVE SURGERY

Despite advances in cardiac surgery, the long-term mortality after valve replacement surgery is greater than that of a normal population of similar age. A transvalvular gradient of varying severity is present in most patients after valve replacement, which may be aggravated during exercise (1,15). Moreover, after implantation of a mechanical prosthesis, which is common in young patients

requiring valve replacement, chronic anticoagulation is required. These considerations are important in determining an athlete's suitability for competition after valve replacement. In patients who have undergone aortic valve repair or, more commonly, mitral valve repair, a different set of issues regarding the risks of physical trauma during athletic competition must be considered.

In assessing the athlete's capacity for physical activity after valve surgery, exercise stress testing to at least the level of activity performed in the competitive sport is valuable. In some cases, assessment of prosthetic valve function during exercise will also provide useful information.

Recommendations

1. It is reasonable for athletes with aortic or mitral bioprosthetic valves, not taking anticoagulant agents, who have normal valvular function and normal LV function to participate in low-intensity and some moderate-intensity competitive sports (classes IA, IB, IC, and IIA) (*Class IIa; Level of Evidence C*).
2. Athletes with aortic or mitral mechanical prosthetic valves taking anticoagulant agents with normal valvular function and normal LV function can reasonably participate in low-intensity competitive sports if there is low likelihood of bodily contact (classes IA, IB, and IIA) (*Class IIa; Level of Evidence C*).
3. It is reasonable for patients with MS who have undergone successful percutaneous mitral balloon valvotomy or surgical commissurotomy to participate in competitive sports based on the residual severity of the MS or MR and pulmonary artery pressures at rest and with exercise (*Class IIa; Level of Evidence C*).
4. Athletes who have undergone mitral valve repair for MR or surgical aortic valve repair, have no or mild residual AR or MR, and have normal LV systolic function may be considered for participation in sports at the discretion of the managing physician if there is low likelihood of bodily contact (classes IA, IB, and IIA) (*Class IIa; Level of Evidence C*).

DISCLOSURES

Writing Group Disclosures

Writing Group Member	Employment	Research Grant	Other Research Support	Speakers Bureau/Honoraria	Expert Witness	Ownership Interest	Consultant/Advisory Board	Other
Robert O. Bonow	Northwestern University	None	None	None	None	None	None	None
Rick A. Nishimura	Mayo Clinic	None	None	None	None	None	None	None
Paul D. Thompson	Hartford Hospital, Hartford, CT	None	None	None	None	None	None	None
James E. Udelson	Tufts Medical Center	None	None	None	None	None	None	None

This table represents the relationships of writing group members that may be perceived as actual or reasonably perceived conflicts of interest as reported on the Disclosure Questionnaire, which all members of the writing group are required to complete and submit. A relationship is considered to be "significant" if (a) the person receives \$10,000 or more during any 12-month period, or 5% or more of the person's gross income; or (b) the person owns 5% or more of the voting stock or share of the entity, or owns \$10,000 or more of the fair market value of the entity. A relationship is considered to be "modest" if it is less than "significant" under the preceding definition.

Reviewer Disclosures

Reviewer	Employment	Research Grant	Other Research Support	Speakers Bureau/Honoraria	Expert Witness	Ownership Interest	Consultant/Advisory Board	Other
Michael S. Emery	Greenville Health System	None	None	None	None	None	None	None
Geetha Raghuvier	Children's Mercy Hospital	None	None	None	None	None	None	None

This table represents the relationships of reviewers that may be perceived as actual or reasonably perceived conflicts of interest as reported on the Disclosure Questionnaire, which all reviewers are required to complete and submit. A relationship is considered to be "significant" if (a) the person receives \$10,000 or more during any 12-month period, or 5% or more of the person's gross income; or (b) the person owns 5% or more of the voting stock or share of the entity, or owns \$10,000 or more of the fair market value of the entity. A relationship is considered to be "modest" if it is less than "significant" under the preceding definition.

REFERENCES

- Nishimura RA, Otto CM, Bonow RO, Carabello BA, Erwin JP 3rd, Guyton RA, O'Gara PT, Ruiz CE, Skubas NJ, Sorajja P, Sundt TM 3rd, Thomas JD. 2014 AHA/ACC guideline 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 [published corrections appear in J Am Coll Cardiol. 2014;63:2489]. J Am Coll Cardiol. 2014;63:2438-88. <http://dx.doi.org/10.1016/j.jacc.2014.02.537>.
- Siu SC, Silversides CK. Bicuspid aortic valve disease. J Am Coll Cardiol. 2010;55:2789-800. <http://dx.doi.org/10.1016/j.jacc.2009.12.068>.
- Maron BJ. Sudden death in young athletes. N Engl J Med. 2003;349:1064-75. <http://dx.doi.org/10.1056/NEJMr022783>.
- Van Hare GF, Ackerman MJ, Evangelista JK, Kovacs RJ, Myerburg RJ, Shafer KM, Warnes CA, Washington RL, on behalf of the American Heart Association Electrocardiography and Arrhythmias Committee of the Council on Clinical Cardiology, Council on Cardiovascular Disease in the Young, Council on Cardiovascular and Stroke Nursing, Council on Functional Genomics and Translational Biology, and the American College of Cardiology. Eligibility and disqualification recommendations for competitive athletes with cardiovascular abnormalities: Task Force 4: congenital heart disease: a scientific statement from the American Heart Association and American College of Cardiology. J Am Coll Cardiol. 2015 In Press. <http://dx.doi.org/10.1016/j.jacc.2015.09.036>.
- Zoghbi WA, Enriquez-Sarano M, Foster E, Grayburn PA, Kraft CD, Levine RA, Nihoyannopoulos P, Otto CM, Quinones MA, Rakowski H, Stewart WJ, Waggoner A, Weissman NJ, American Society of Echocardiography. Recommendations for evaluation of the severity of native valvular regurgitation with two-dimensional and Doppler echocardiography. J Am Soc Echocardiogr. 2003;16:777-802. [http://dx.doi.org/10.1016/S0894-7317\(03\)00335-3](http://dx.doi.org/10.1016/S0894-7317(03)00335-3).
- Pelliccia A, Culasso F, Di Paolo FM, Maron BJ. Physiologic left ventricular cavity dilatation in elite athletes. Ann Intern Med. 1999;130:23-31.
- Pelliccia A, Kinoshita N, Pisicchio C, Quattrini F, Dipaolo FM, Ciardo R, Di Giacinto B, Guerra E, De Blasiis E, Casasco M, Culasso F, Maron BJ. Long-term clinical consequences of intense, uninterrupted endurance training in Olympic athletes. J Am Coll Cardiol. 2010;55:1619-25. <http://dx.doi.org/10.1016/j.jacc.2009.10.068>.
- Pelliccia A, Maron BJ, Culasso F, Spataro A, Caselli G. Athlete's heart in women: echocardiographic characterization of highly trained elite female athletes. JAMA. 1996;276:211-5.
- Dujardin KS, Enriquez-Sarano M, Schaff HV, Bailey KR, Seward JB, Tajik AJ. Mortality and morbidity of aortic regurgitation in clinical practice: a long-term follow-up study. Circulation. 1999;99:1851-7.
- Bonow RO, Lakatos E, Maron BJ, Epstein SE. Serial long-term assessment of the natural history of asymptomatic patients with chronic aortic regurgitation and normal left ventricular systolic function. Circulation. 1991;84:1625-35.
- Braverman AC, Harris KM, Kovacs RJ, Maron BJ, on behalf of the American Heart Association Electrocardiography and Arrhythmias Committee of the Council on Clinical Cardiology, Council on Cardiovascular Disease in the Young, Council on Cardiovascular and Stroke Nursing, Council on Functional Genomics and Translational Biology, and the

American College of Cardiology. Eligibility and disqualification recommendations for competitive athletes with cardiovascular abnormalities: Task Force 7: aortic diseases, including Marfan syndrome: a scientific statement from the American Heart Association and American College of Cardiology. *J Am Coll Cardiol*. 2015 In Press. <http://dx.doi.org/10.1016/j.jacc.2015.09.039>.

12. Michelena HI, Khanna AD, Mahoney D, Margaryan E, Topilsky Y, Suri RM, Eidem B,

Edwards WD, Sundt TM 3rd, Enriquez-Sarano M. Incidence of aortic complications in patients with bicuspid aortic valves. *JAMA*. 2011;306:1104-12. <http://dx.doi.org/10.1001/jama.2011.1286>.

13. Rahimtoola SH, Durairaj A, Mehra A, Nuno I. Current evaluation and management of patients with mitral stenosis. *Circulation*. 2002;106:1183-8.

14. Bonow RO. Chronic mitral regurgitation and aortic regurgitation: have indications for surgery changed?

J Am Coll Cardiol. 2013;61:693-701. <http://dx.doi.org/10.1016/j.jacc.2012.08.1025>.

15. Rahimtoola SH. Choice of prosthetic heart valve for adult patients. *J Am Coll Cardiol*. 2003;41:893-904.

KEY WORDS ACC/AHA Scientific Statements, athletes, cardiovascular abnormalities, valvular heart disease

having an elevated BP at or above certain levels measured by routine sphygmomanometry under appropriate conditions on at least 2 separate occasions separated by at least 1 week (3). However, BP measurements in the competitive athlete are typically obtained by different healthcare providers, which makes it particularly necessary that the testing conditions be standardized before the diagnosis of hypertension is made. People >18 years of age with a BP >140 mm Hg systolic and/or >90 mm Hg diastolic are considered to have hypertension (3). In children and adolescents, hypertension is defined as average systolic or diastolic BP levels greater than the 95th percentile for sex, age, and height; however, earlier physical maturation of the competitive athlete leaves open to question when an adult age criterion for hypertension should be applied to the adolescent (4). In determining the level of competitive athletic activity that a hypertensive person may engage in, it is also important to determine the degree of hypertension-related target-organ damage. Although hypertension has been associated with an increased risk for complex ventricular arrhythmias and sudden death, this cardiovascular risk factor per se has not been implicated in sudden death in young competitive athletes (5). For the general population, increased levels of noncompetitive recreational physical activity are generally regarded as beneficial. With physical activity, BP typically falls, the incidence of hypertension drops (6,7), and protection against stroke is afforded (8). Those who are hypertensive derive protection from both all-cause and cardiovascular mortality by maintaining higher levels of cardiorespiratory fitness (9).

ASSESSMENT OF BP

BP should be accurately measured in all people who wish to participate in competitive athletics before they begin training. BP should be measured by standard techniques, using the guidelines listed in the [Table](#). It is common in young athletes to have their BP measured with an inappropriately sized BP cuff because of their often larger (>33 cm) midarm circumference. In these people, BP measured this way is often spuriously increased and results in unnecessary referrals to clinicians for evaluation and consideration of antihypertensive therapy. Also, there are often discrepancies between in-office and out-of-office BP measurements. For example, elevations induced by anxiety related to the medical examination are seen in young people concerned about the potential negative consequences of the examination. Anxiety-related BP elevations may be marked by elevations in heart rate, which further complicates the interpretation of the physical examination findings. In such instances, it is advisable to obtain unbiased and more comprehensive information through the use of 24-hour ambulatory BP monitoring.

TABLE Guidelines for Clinic (or Office) BP Measurement

Posture	
BP 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 BP is recorded.	
Circumstances	
No caffeine should be ingested during the hour preceding the reading, and no smoking during the 30 min preceding the reading.	
A quiet, warm setting should be available for BP measurements.	
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 validated electronic (digital) device, a recently calibrated aneroid or mercury column sphygmomanometer.	
Technique	
Number of readings	
On each occasion, take at least 2 readings, separated by as much time as is practical. If readings vary by >10 mm Hg, take additional readings until 2 consecutive readings are within 10 mm Hg.	
If the arm pressure is elevated, take the measurement in 1 leg to rule out aortic coarctation (particularly in patients <30 y of age).	
Initially, take pressures in both arms; if the blood pressures differ, use the arm with the higher pressure.	
If the initial values are elevated, obtain 2 other sets of readings at least 1 wk apart.	
Performance	
Inflate the bladder quickly to a pressure 20 mm Hg above the systolic BP, as recognized by the disappearance of the radial pulse; deflate the bladder at 2 mm Hg/s.	
Record the Korotkoff phase I (appearance) and phase V (disappearance) sounds. If the Korotkoff sounds are weak, have the patient raise the arm, then open and close the hand 5-10 times, and then reinflate the bladder quickly.	

BP indicates blood pressure.

In some people, extremely high BPs may occur on a single measurement. In this type of patient, ambulatory BP monitoring would help to further stratify the athlete's risk of hypertension at present or in the future if borderline values were obtained. Ambulatory BP measurement in people with elevated exercise BP values improves the prediction of left ventricular hypertrophy (LVH) by echocardiography and development of sustained hypertension according to 1 study with an 8-year follow-up (10).

EVALUATION

All people who are diagnosed as hypertensive, whether competitive athletes or not, need a thorough but directed history and physical examination with a minimal number of laboratory tests. The history should be sure to determine whether the person has a family history of hypertension or cardiovascular disease, symptoms suggestive of a pheochromocytoma (paroxysmal hypertension, headache,

diaphoresis, and palpitations) or if he or she uses nonsteroidal anti-inflammatory agents or street drugs, especially cocaine or amphetamines. Use of nonsteroidal anti-inflammatory agents is particularly common among competitive athletes, who often have minor injuries for which these analgesic agents are beneficial and available without a prescription. Amphetamines are used to increase mental alertness and decrease fatigue (11). Participation in certain extracurricular activities, such as high-contact sports, may influence male participants to misuse prescription stimulants as performance enhancers either on or off the playing field. However, the use of these agents is not more common in competitive athletes than in the general population. Although anabolic steroid abuse is becoming increasingly uncommon in athletes in competitive sports, an analysis of existing evidence suggests that chronic anabolic steroid use does have a negative impact on lipoproteins and BP in athletes (12).

The physical examination should be used to look for clues to an identifiable cause of hypertension (so-called secondary hypertension) such as abdominal bruits, which may indicate the presence of renal artery stenosis and renovascular hypertension, or a cushingoid body habitus or abdominal striae suggesting adrenocortical hormonal excess. The laboratory tests should also be limited to assessing the presence of other cardiovascular risk factors such as dyslipidemias, glucose intolerance, and diabetes mellitus, and particularly chronic renal disease, a problem common among young black men and that is often asymptomatic until its later stages. All competitive athletes should have a lipid profile (total cholesterol, high-density lipoprotein cholesterol, and serum triglycerides) performed; fasting serum glucose, electrolytes, and hemoglobin measured; and urinary protein estimated by dipstick (3,13). Although it is usually recommended that a lipid profile and glucose determination should be obtained after at least a 9-hour fast, this may be logistically difficult. Having the blood drawn in the athletes in a fasting state may not be feasible in most circumstances, and it may be more reasonable to obtain the samples when convenient and only repeat the test in the fasting state when it is abnormal (13).

A 12-lead ECG is recommended but not mandated to ascertain the presence of LVH or conduction abnormalities, although the yield will be small. In those people with stage 2 hypertension (a systolic BP >160 mm Hg or a diastolic BP >100 mm Hg) or who have a suggestion of target-organ damage on history or physical examination, a screening echocardiogram is advisable to distinguish physiological hypertrophy attributable to physical exercise (athlete's heart) versus pathological LVH from hypertension. Athletes with normal (or physiological) hypertrophy have echocardiographic and other imaging evidence of increased posterior and septal wall thicknesses with normal cavity chamber size accompanied by normal rates of left ventricular filling

during diastole (14); in contrast, hypertrophy caused by hypertension, although having similar structural findings, has both impaired rates of left ventricular filling and slow isovolumic relaxation times (15). If needed, the pathophysiology of cardiac hypertrophy attributable to physiological causes versus pathophysiological causes (hypertension) can be discriminated with echocardiography using Doppler imaging or magnetic resonance imaging as a tertiary methodology. People with larger body size and blacks may have an increase in wall thicknesses on echocardiography, which should be correlated with ECG, clinical signs and symptoms, and family history before they are advised against participation in competitive sports. It is rare for physiological increased left ventricular wall thicknesses to exceed 13 mm and indicates the advisability of a referring the patient for further evaluation for hypertrophic cardiomyopathy with ECG, clinical assessment, and family history. Of note, LVH is more prevalent in blacks and is an independent predictor of diminished cardiovascular survival (16). The ECG is widely available, inexpensive, and has high specificity but poor sensitivity for detection of LVH; however, the combination of an abnormal ECG, any signs and symptoms of heart disease, and a positive family history for premature cardiac death warrants further evaluation. Cardiac stress testing is not warranted unless there are symptoms that occur with maximal exercise. The competitive athlete need not routinely require orthostatic BP determinations unless the athlete is symptomatic in the upright position in a volume replete state.

In an adolescent or young adult (i.e., <25 years of age) with stage 2 hypertension, it may be appropriate to refer this person for further evaluation and therapy to a cardiologist or hypertension specialist. The workup for secondary forms of hypertension and proper pharmacological management is often outside the scope of general pediatricians and family practitioners who might otherwise be seeing these athletes.

EFFECTS OF EXERCISE ON BP

Both systolic and diastolic BP rise during resistance (static or isometric) exercise, and strenuous aerobic or resistance exertion may precipitate myocardial infarction and sudden death in susceptible, untrained people. In the long term, both systolic and diastolic BPs are lower with aerobic (dynamic) exercise and remain lower for up to 24 hours (17). In a person with normal BP at rest, a rise in systolic BP to >200 mm Hg during an exercise treadmill test may suggest underlying hypertension. This person may benefit from further investigation, including 24-hour ambulatory BP monitoring, to document true sustained hypertension (18). A hypertensive responsive to exercise testing may also indicate an independent risk for cardiovascular events and mortality (19).

Recommendations

1. It is reasonable that the presence of stage 1 hypertension in the absence of target-organ damage should not limit the eligibility for any competitive sport. Once having begun a training program, the hypertensive athlete should have BP measured every 2 to 4 months (or more frequently, if indicated) to monitor the impact of exercise (*Class I; Level of Evidence B*).
2. Before people begin training for competitive athletics, it is reasonable that they undergo careful assessment of BP, and those with initially high levels (>140 mm Hg systolic or >90 mm Hg diastolic) should have comprehensive out-of-office measurements to exclude errors in diagnosis. Ambulatory BP monitoring with proper cuff and bladder size would be the most precise means of measurement (*Class I; Level of Evidence B*).
3. Those with prehypertension (BP of 120/80 mm Hg–139/89 mm Hg) should be encouraged to modify their lifestyles but should not be restricted from physical activity. Those with sustained hypertension should have screening echocardiography performed. Athletes with LVH beyond that seen with “athlete’s heart” should limit participation until BP is normalized by appropriate antihypertensive drug therapy (*Class IIa; Level of Evidence B*).
4. It is reasonable that athletes with stage 2 hypertension (a systolic BP >160 mm Hg or a diastolic BP >100 mm Hg), even without evidence of target-organ damage, should be restricted, particularly from high static sports, such as weight lifting, boxing, and wrestling, until hypertension is controlled by either lifestyle modification or drug therapy (*Class IIa; Level of Evidence B*).
5. When prescribing antihypertensive drugs, particularly diuretic agents, for competitive athletes, it is reasonable for clinicians to use drugs already registered with appropriate governing bodies and if necessary obtain a therapeutic exemption (*Class IIa; Level of Evidence B*).
6. When hypertension coexists with another cardiovascular disease, it is reasonable that eligibility for participation in competitive athletics is based on the type and severity of the associated condition (*Class IIa; Level of Evidence C*).

DISCLOSURES

Writing Group Disclosures

Writing Group Member	Employment	Research Grant	Other Research Support	Speakers Bureau/Honoraria	Expert Witness	Ownership Interest	Consultant/Advisory Board	Other
Henry R. Black	New York University (Retired)	None	None	None	None	None	None	None
Keith Ferdinand	Tulane University	Boehringer Ingelheim*	None	None	None	None	Amgen*; AstraZeneca*; Boehringer Ingelheim*; Sanofi	None
Domenic Sica	Virginia Commonwealth University	None	None	None	None	None	None	None
William B. White	University of Connecticut	None	None	None	None	None	None	None

This table represents the relationships of writing group members that may be perceived as actual or reasonably perceived conflicts of interest as reported on the Disclosure Questionnaire, which all members of the writing group are required to complete and submit. A relationship is considered to be “significant” if (a) the person receives \$10,000 or more during any 12-month period, or 5% or more of the person’s gross income; or (b) the person owns 5% or more of the voting stock or share of the entity, or owns \$10,000 or more of the fair market value of the entity. A relationship is considered to be “modest” if it is less than “significant” under the preceding definition.

*Modest.

Reviewer Disclosures

Reviewer	Employment	Research Grant	Other Research Support	Speakers Bureau/Honoraria	Expert Witness	Ownership Interest	Consultant/Advisory Board	Other
Lawrence Fine	NHLBI	None	None	None	None	None	None	None
Samuel S. Gidding	Nemours Foundation	GlaxoSmithKline†	None	None	None	None	None	None
Martha A. Gulati	Ohio State University	None	None	None	None	None	None	None
Christina Salazar	Lawrence Memorial Hospital	None	None	None	None	None	None	None
Richard A. Stein	New York University	None	None	None	None	None	None	None

This table represents the relationships of reviewers that may be perceived as actual or reasonably perceived conflicts of interest as reported on the Disclosure Questionnaire, which all reviewers are required to complete and submit. A relationship is considered to be “significant” if (a) the person receives \$10,000 or more during any 12-month period, or 5% or more of the person’s gross income; or (b) the person owns 5% or more of the voting stock or share of the entity, or owns \$10,000 or more of the fair market value of the entity. A relationship is considered to be “modest” if it is less than “significant” under the preceding definition.

†Significant.

REFERENCES

- Go AS, Mozaffarian D, Roger VL, Benjamin EJ, Berry JD, Borden WB, Bravata DM, Dai S, Ford ES, Fox CS, Franco S, Fullerton HJ, Gillespie C, Hailpern SM, Heit JA, Howard VJ, Huffman MD, Kissela BM, Kittner SJ, Lackland DT, Lichtman JH, Lisabeth LD, Magid D, Marcus GM, Marelli A, Matchar DB, McGuire DK, Mohler ER, Moy CS, Mussolino ME, Nichol G, Paynter NP, Schreiner PJ, Sorlie PD, Stein J, Turan TN, Virani SS, Wong ND, Woo D, Turner MB, on behalf of the American Heart Association Statistics Committee and Stroke Statistics Subcommittee. Heart disease and stroke statistics-2013 update: a report from the American Heart Association [published corrections appear in *Circulation*. 2013;127:e841 and *Circulation*. 2013;127:doi:10.1161/CIR.Obo13e31828124ad]. *Circulation*. 2013;127:e6-245. <http://dx.doi.org/10.1161/CIR.Obo13e31828124ad>.
- McNiece KL, Poffenbarger TS, Turner JL, Franco KD, Sorof JM, Portman RJ. Prevalence of hypertension and pre-hypertension among adolescents. *J Pediatr*. 2007;150:640-4.e1. <http://dx.doi.org/10.1016/j.jpeds.2007.01.052>.
- Chobanian AV, Bakris GL, Black HR, Cushman WC, Green LA, Izzo JL Jr., Jones DW, Materson BJ, Oparil S, Wright JT Jr., Roccella EJ, National High Blood Pressure Education Program Coordinating Committee. JNC 7: complete report: seventh report of the Joint National Committee on Prevention, Detection, Evaluation, and Treatment of High Blood Pressure. *Hypertension*. 2003;42:1206-52. <http://dx.doi.org/10.1161/01.HYP.0000107251.49515.c2>.
- Moyer VA, U.S. Preventive Services Task Force. Screening for primary hypertension in children and adolescents: U.S. Preventive Services Task Force recommendation statement. *Pediatrics*. 2013;132:907-14. <http://dx.doi.org/10.1542/peds.2013-2864>.
- Maron BJ. Sudden death in young athletes. *N Engl J Med*. 2003;349:1064-75. <http://dx.doi.org/10.1056/NEJMr022783>.
- 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.
- 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. <http://dx.doi.org/10.1161/01.HYP.0000107400.72456.19>.
- Lee CD, Folsom AR, Blair SN. Physical activity and stroke risk: a meta-analysis. *Stroke*. 2003;34:2475-81. <http://dx.doi.org/10.1161/01.STR.0000091843.02517.9D>.
- 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.
- Zanettini JO, Pisani Zanettini J, Zanettini MT, Fuchs FD. Correction of the hypertensive response in the treadmill testing by the work performance improves the prediction of hypertension by ambulatory blood pressure monitoring and incidence of cardiac abnormalities by echocardiography: results of an eight year follow-up study. *Int J Cardiol*. 2010;141:243-9. <http://dx.doi.org/10.1016/j.ijcard.2008.11.208>.
- Veliz P, Boyd C, McCabe SE. Adolescent athletic participation and nonmedical Adderall use: an exploratory analysis of a performance-enhancing drug. *J Stud Alcohol Drugs*. 2013;74:714-9.
- Achar S, Rostamian A, Narayan SM. Cardiac and metabolic effects of anabolic-androgenic steroid abuse on lipids, blood pressure, left ventricular dimensions, and rhythm. *Am J Cardiol*. 2010;106:893-901. <http://dx.doi.org/10.1016/j.amjcard.2010.05.013>.
- Glover DW, Maron BJ. Profile of preparticipation cardiovascular screening for high school athletes. *JAMA*. 1998;279:1817-9.
- Granger CB, Karimeddini MK, Smith VE, Shapiro HR, Katz AM, Riba AL. Rapid ventricular filling in left ventricular hypertrophy, I: physiologic hypertrophy. *J Am Coll Cardiol*. 1985;5:862-8.
- Smith VE, Schulman P, Karimeddini MK, White WB, Meeran MK, Katz AM. Rapid ventricular filling in left ventricular hypertrophy, II: pathologic hypertrophy. *J Am Coll Cardiol*. 1985;5:869-74.
- Kizer JR, Arnett DK, Bella JN, Parancas M, Rao DC, Province MA, Oberman A, Kitzman DW, Hopkins PN, Liu JE, Devereux RB. Differences in left ventricular structure between black and white hypertensive adults: the Hypertension Genetic Epidemiology Network study. *Hypertension*. 2004;43:1182-8. <http://dx.doi.org/10.1161/01.HYP.0000128738.94190.9f>.
- Pescatello LS, Fargo AE, Leach CN Jr., Scherzer HH. Short-term effect of dynamic exercise on arterial blood pressure. *Circulation*. 1991;83:1557-61.
- Pickering TG, White WB, American Society of Hypertension Writing Group. When and how to use self (home) and ambulatory blood pressure monitoring. *J Am Soc Hypertens*. 2008;2:119-24. <http://dx.doi.org/10.1016/j.jash.2008.04.002>.
- Adabag AS, Grandits GA, Prineas RJ, Crow RS, Bloomfield HE, Neaton JD, MRFIT Research Group. Relation of heart rate parameters during exercise test to sudden death and all-cause mortality in asymptomatic men. *Am J Cardiol*. 2008;101:1437-43. <http://dx.doi.org/10.1016/j.amjcard.2008.01.021>.

KEY WORDS ACC/AHA Scientific Statements, athletes, blood pressure measurement, cardiovascular abnormalities, hypertension

Marfan syndrome, an autosomal dominant disorder of connective tissue with an estimated prevalence of 1 in 5,000 to 10,000, is caused by abnormal fibrillin-1 attributable to mutations in the *FBN1* gene (2). Manifestations involve multiple organ systems, including the aorta, heart and valves, skeleton, eye, lungs, and dura. *FBN1* mutations can be identified in the vast majority of patients satisfying the revised Ghent criteria for Marfan syndrome (2). The diagnosis of Marfan syndrome is made by use of clinical criteria, imaging, family history, and genetic testing as outlined in the revised Ghent criteria (Tables 1 and 2) (2). Cardiovascular features of Marfan syndrome include mitral valve prolapse, mitral regurgitation, aortic root dilatation (most pronounced at the sinuses of Valsalva), and aortic dissection (2). The descending aorta, although less commonly involved in young patients, is also at risk for aneurysm formation and dissection.

Other genetically triggered aortic aneurysm syndromes and conditions associated with aortopathy may increase the risk of aortic dissection in competitive athletes. Loeys-Dietz syndrome is caused by mutations in *TGFBR1* and *TGFBR2* and is characterized by craniofacial features, arterial tortuosity, and aneurysms of the aorta and branch vessels, as well as increased risk of dissection at relatively small arterial dimensions (3). Vascular Ehlers-Danlos syndrome, caused by mutations in *COL3A1*, is associated with dissection and rupture of the aorta and branch vessels, even at relatively normal arterial dimensions.

TAA or dissection may be familial and is inherited as an autosomal dominant trait with decreased penetrance and variable expression. Mutations in several genes have been recognized as causing TAA disease, including *ACTA2*, *TGFBR1*, *TGFBR2*, *FBN1*, *MYH11*, *SMAD3*, *MLCK*, and *TGFB2*. Familial TAA syndromes may be associated with cerebral aneurysms or BAV; some patients have nonvascular manifestations (4,5).

BAV, which affects ~1% of the general population, may be associated with dilatation of the aortic root or

ascending aorta (6). BAV with or without TAA may be familial, and the specific gene loci responsible are yet to be determined. The prevalence of BAV in first-degree relatives of a person with BAV has been demonstrated to be ~9% (6). Cystic medial degeneration and abnormal aortic wall stress accompany BAV aortic disease independent of the valvular lesion (6). BAV with aortic aneurysm is a risk factor for aortic dissection (7). The risk of aortic dissection differs among genetically triggered aortopathies, being higher in those with Loeys-Dietz syndrome and Marfan syndrome than in BAV aortopathy.

MEASURING THE AORTIC ROOT AND ASCENDING AORTA

The ascending aorta may be divided into 2 segments, the aortic root and the upper ascending aorta. The aortic root begins at the aortic valve, includes the sinuses of Valsalva, and extends to the sinotubular junction. The upper portion of the ascending aorta begins at the sinotubular junction and rises to join the aortic arch. The normal aortic root diameter is dependent on multiple factors, including age, sex, body size, location of the aortic measurement, particular type of imaging modality used, and accuracy of measurement ascertainment (4,8). In adults, aortic diameters are larger in men than in women by 1 to 3 mm, whereas studies in children have not consistently

TABLE 1 Revised Ghent Criteria for the Diagnosis of Marfan Syndrome

In the absence of a family history of Marfan syndrome, any of the following:	
1. Dilated aorta (z score >2) and ectopia lentis = Marfan syndrome*	
2. Dilated aorta (z score >2) and <i>FBN1</i> mutation = Marfan syndrome	
3. Dilated aorta (z score >2) and systemic score >7 (see Table 2) = Marfan syndrome*	
4. Ectopia lentis and <i>FBN1</i> associated with known aortic dilatation = Marfan syndrome	
In the presence of a family history of Marfan syndrome, any of the following:	
5. Ectopia lentis and family history of Marfan syndrome = Marfan syndrome	
6. Systemic score >7 and family history of Marfan syndrome = Marfan syndrome*	
7. Dilated aorta (z score >2 at age ≥20 y; z score >3 at <20 y of age) and family history of Marfan syndrome = Marfan syndrome*	

*Caveat: Without discriminating features of another connective tissue disorder such as Loeys-Dietz syndrome, vascular Ehlers-Danlos syndrome, or Shprintzen-Goldberg syndrome, and after mutation analysis for *TGFBR1*, *TGFBR2*, *TGFB2*, *SMAD3*, *SKI*, *COL3A1*, or other genes as appropriate. Other genes/conditions will emerge with time. Modified with permission from Loeys et al. (2) Copyright © 2010, British Medical Journal Publishing Group.

TABLE 2 Scoring of Systemic Features in the Marfan Syndrome*

Feature	Points
Wrist and thumb sign	3
Wrist or thumb sign	1
Pectus carinatum deformity	2
Pectus excavatum or chest asymmetry	1
Hindfoot deformity	2
Plain pes planus	1
Pneumothorax	2
Lumbosacral dural ectasia	2
Protrusio acetabuli	2
Reduced upper-segment to lower-segment ratio (<0.85 in white adults; <0.78 in black adults) and increased arm span-to-height ratio (>1.05) and no severe scoliosis	1
Scoliosis or thoracolumbar kyphosis	1
Reduced elbow extension	1
Facial features (3 of 5): dolichocephaly, enophthalmos, down-slanting palpebral fissures, malar hypoplasia, retrognathia	1
Skin striae	1
Myopia (>3 diopters)	1
Mitral valve prolapse	1

Maximum total, 20 points; score >7 indicates systemic involvement.

*A detailed explanation of the systemic score and nosology may be found at <http://www.marfandx.org>. Modified with permission from Loeys et al. (2) Copyright © 2010, British Medical Journal Publishing Group.

demonstrated a sex difference in aortic diameter when corrected for body surface area (BSA) (8). Ascending aortic dimensions in adults are also related to age, sex, and BSA (9).

Variability in the measured aortic diameter may result from the type of imaging modality used, whether contrast is used, and whether internal or external aortic diameters are recorded. For example, transthoracic echocardiographic nomograms have reported aortic root diameters using sinus-to-sinus measurements from a leading-edge technique at end diastole (10), whereas *z*-score determinations validated in children have used maximal end-systolic diameter at the sinuses of Valsalva using inner-edge-to-inner-edge measurements (11). Measurements should be taken perpendicular to the axis of blood flow and should include the largest measured aortic diameter (whether at the sinuses of Valsalva or the ascending aorta) (4). Images taken from echocardiography, computed tomography (CT), or magnetic resonance imaging may overestimate the true aortic diameter if oblique slices are obtained. CT and magnetic resonance imaging measurements from sinus to commissure are generally smaller than echocardiographic measurements from sinus to sinus (8). CT or magnetic resonance imaging techniques are used when the extent of aortic enlargement is not adequately or completely visualized by the echocardiogram. Imaging techniques that avoid or minimize radiation are recommended whenever possible, particularly when serial assessment is anticipated. Regardless of which imaging technique is used, it is important that serial measurements be made at the same location by the same method for appropriate clinical correlation.

Older nomograms that predict normal and abnormal aortic dimensions are limited by such factors as failure to account for sex differences, limited age ranges of subjects studied (especially teenagers), marked jumps in “normal” aortic diameter based on age-range strata, and the use of small sample sizes (8,10).

***z* Scores**

Notably, *z* scores that incorporate height, weight, age, and sex are now preferred for determination of normal aortic diameter as opposed to a single aortic dimension (8). The *z* score describes how many standard deviations above or below a size or age-specific population mean a given measurement lies (12). They are especially useful for evaluation of cardiac dimensions in the young, whose normal values change during growth.

Aortic dilatation is recognized when the difference between the observed sinus of Valsalva diameter and the value predicted for age, sex, and BSA (*z* score) is >2.0, which corresponds to approximately the 98th percentile of the general population (8). A *z* score of 3 corresponds to the 99.9th percentile. Mild, moderate, and severe aortic

dilatation may be defined by *z*-score values of 2 to 3, 3.01 to 4.0, and >4.0, respectively (8,13). Reference values for ascending aortic diameter assessed by echocardiography are also available from large databases (14).

A formula for calculating aortic sinus of Valsalva diameter *z* scores was derived recently from a data set of 1207 healthy subjects >15 years old, in whom aortic root diameter ranged from 2.1 to 4.3 cm (8). Aortic dimensions were calculated by echocardiogram at end diastole from sinus to sinus using a leading-edge-to-leading-edge technique (Figure). *z* Scores are calculated from this database using the following equation (8) (online-only Data Supplement aortic *z*-score calculator):

$$\begin{aligned} \text{Expected aortic root size: } & 2.423 + (\text{age [years]} \times 0.009) \\ & + (\text{BSA [square meters]} \times 0.461) \\ & - (\text{sex [1 = man, 2 = woman]} \times 0.267); \\ \text{standard error of estimate} & = 0.261 \text{ cm} \end{aligned}$$

$$\begin{aligned} z \text{ Score} = & (\text{observed aortic root size} \\ & - \text{expected aortic root size}) / 0.261 \end{aligned}$$

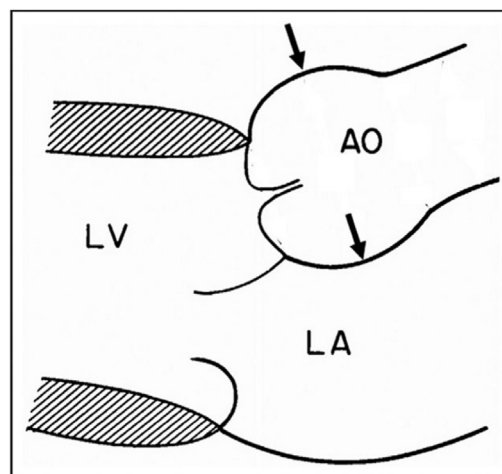
For example, a 22-year-old man with a BSA of 2.0 m² has an aortic root diameter of 4.1 cm at the sinuses of Valsalva. Thus, his expected aortic root size is calculated as follows:

$$2.423 + (22 \times 0.009) + (2.0 \times 0.461) - 0.267 = 3.276$$

$$\begin{aligned} & 4.1 (\text{observed aortic root size}) \\ & - 3.28 (\text{expected aortic root size}) = 0.824 \end{aligned}$$

$$0.824 / 0.261 (\text{standard error of estimate}) = 3.16$$

FIGURE Schematic of the Aortic Root Showing Measurement of the Aortic Root Diameter at Maximum Width Parallel to the Aortic Annular Plane by American Society of Echocardiography Leading-Edge Convention (Arrows)



AO indicates aorta; LA, left atrium; and LV, left ventricle. Reproduced with permission from Devereux et al. (8) Copyright © 2012, Elsevier Inc.

Thus, the *z* score is 3.16, which is significantly abnormal for this patient.

AORTIC DIMENSIONS IN ATHLETES

Intense physical exertion is associated with hemodynamic changes that increase aortic wall tension and may increase aortic dimension (15-17). Chronic intense weight training may influence aortic dimension (18). Furthermore, elite athletes have slightly larger aortas at the sinuses of Valsalva than nonathletic control subjects (19). Although mild aortic enlargement may be a normal adaptation to intense training, large increases in aortic size are unusual in athletes and when present are more consistent with an underlying pathological aortopathy, which may be exacerbated by exercise training (19).

TALL ATHLETES

Although increasing BSA is associated with larger aortic diameters, there is a nonlinear relationship, with a plateau, between aortic root dimensions and height (>189 cm or 74.5 inches in men; >175 cm or 69 inches in women) and BSA (>2.3 m²) in very tall people (20). A small proportion of athletes will have an aortic dimension slightly greater than the diameter considered to be at the upper limits of normal (i.e., >2 standard deviations above the mean, or *z* score >2) (15,16). Therefore, it is important to avoid attributing the enlarged aorta in tall (or large) athletes solely to height, BSA, or a physiological response to exercise (19). Mild aortic dilation in an athlete should trigger evaluation to determine whether an underlying aortopathy is present and whether the aortic size conveys an increased risk to the athlete.

We underscore that for athletes with aortic *z* scores above the normal range for age, sex, and BSA (i.e., *z* score >2 to 2.5), evaluation by a knowledgeable specialist, and often by a multidisciplinary team that includes a medical geneticist and cardiologist, is recommended to exclude an underlying disorder associated with aortic dilatation (such as Marfan syndrome, familial TAA syndrome, or BAV disease). Indeed, systemic features of some disorders may be subtle and often overlap with those in the general population. Referral to a specialized center with expertise in the clinical and genetic evaluation of genetic aortic disease may be necessary in some instances. In selected cases, we recognize that it may not be possible to distinguish pathological aortic dilatation from a nonpathological aortic size when the aortic measurement mildly exceeds the normal range in very tall people or in those with large BSA, especially when there is only a single evaluation at only 1 point in time.

OUTCOME AND RISK OF AORTIC DISSECTION AND RUPTURE

There is a paucity of data examining the long-term outcome of athletes with unexplained aortic dilatation (16,17). Of 2317 Italian athletes, 17 males (ages 25 ± 7 years; height 188 ± 10 cm; BSA 2.17 ± 0.25 m²) had aortic diameters >40 mm and were allowed to continue participation (16,17). Over an 8 ± 5 year follow-up, the aortic root increased mildly in diameter from 40.9 ± 1.3 to 42.9 ± 3.6 mm in these 17 athletes, and none experienced acute aortic dissection. Two athletes had progressive aortic dilation to 50 mm by ages 38 and 50 years, respectively (17).

The risk of aortic dissection in the general population is related to many factors, foremost of which is the severity of aortic dilation, and is sometimes triggered acutely by heavy weight lifting or strenuous exercise, including competitive sports (21-22a). However, some patients with acute aortic dissection do not have a markedly dilated aorta at the time of dissection (23,24). In a series of 177 patients without the Marfan syndrome phenotype or BAV who incurred an acute type A dissection, aortic diameter was <50 mm in 42% and <45 mm in 21% at the time of dissection. Furthermore, 12% of women had a dissection at an aortic diameter <40 mm (23). Similarly, in the International Registry of Acute Aortic Dissection, 40% of acute type A dissections occurred with aortic diameters <50 mm (24). There is no evidence that β-blockers, angiotensin receptor blockers, or angiotensin-converting enzyme inhibitors protect athletes from aortic dissection or rupture during intense competitive sports.

There are no prospective data available regarding the risks of competitive athletics in patients who have undergone surgical correction for aortic aneurysm or dissection; however, after aortic root replacement, patients with Marfan syndrome, Loeys-Dietz syndrome, and familial TAA disease remain at risk for distal aortic complications (3,5,25,26). Additionally, BAV aortopathy may involve aortic segments distal to the root (27).

PRIOR RECOMMENDATIONS FOR ATHLETES

The 36th Bethesda Conference Report (2005) recommended that athletes with “unequivocal aortic root enlargement” (therein defined as >40 mm in adults, >2 standard deviations beyond the mean for BSA in children and adolescents, or a *z* score of >2) only participate in low-intensity competitive sports (class IA sports) (28). Characterizing an aortic diameter of >40 mm as enlarged in males is an arbitrary but also useful definition, because very few apparently healthy young male athletes have been reported with aortic root diameters >40 mm (17,19). For example, in a study of >2,000 Italian athletes, the 99th

percentile value of aortic diameter by echocardiogram was 40 mm in males and 34 mm in females (16). In a Japanese study, only 6 of 1,562 male athletes (0.38%) had an aortic root dimension >40 mm, and 2 of these also had phenotypic features of the Marfan syndrome (29). In an evaluation of >1000 female Italian athletes, the 99th percentile for aortic size at the sinuses of Valsalva was 34 mm, and no woman had an aortic diameter >36 mm (16,17).

Guidelines for participation in competitive sports have been lacking for those patients with mildly dilated aortic dimensions (i.e., z scores of 2 to 2.5, or 1 to 2 mm above the normal ranges described above) and no diagnosis of an underlying connective tissue disorder, family history of aortic disease, or pathogenic gene mutation associated with aneurysm disease (17). In this situation, there are difficult individual choices regarding sports participation to be made on a case-by-case basis. Discussion with the athlete, parents (when appropriate), and coaches/trainers should include full disclosure and transparency regarding the potential risks of further training and competition. For instance, the mildly dilated aorta may represent a pathological aortic condition, and the aorta may dilate further with continued exercise and athletic participation, or it may dilate years later. In such a person, absence of a pathogenic genetic mutation does not exclude risk. Furthermore, although the absolute risk of aortic dissection or rupture in this clinical situation is unknown, it is not zero. If participation in competitive sports is continued, close aortic surveillance (i.e., every 6 to 12 months) with echocardiography or magnetic resonance angiography (MRA) should be performed to assess aortic dimension (17). The frequency of imaging is dependent on the absolute size of the aorta, the z score, stability of the aortic size, and the intensity of the sport. In the athlete with a mildly dilated aorta, continued aortic enlargement should not be regarded as physiological but rather consistent with an underlying aortopathy; disqualification from competition should result if the aorta continues to enlarge. Because some athletes identified with only a mildly dilated aortic root have required aortic aneurysm surgery several years later, long-term aortic surveillance is recommended even after engagement in the competitive athletic lifestyle has terminated (17,29).

Recommendations

1. Athletes with Marfan syndrome should undergo echocardiographic (and in some instances MRA or CT) measurement of the aortic root dimension every 6 to 12 months, depending on aortic size (*Class I; Level of Evidence C*).
2. Athletes with unexplained TAA, familial TAA syndrome, or known pathogenic mutation leading to a familial TAA syndrome (*ACTA2*, *MYH11*, *FBN1*, *TGFBR1*, *TGFBR2*, *MLCK*, *SMAD3*, *TGFB2*, and others) should undergo echocardiographic and (depending on the diagnosis) MRA or CT surveillance every 6 to 12 months to evaluate for progression of aortic or branch vessel disease (*Class I; Level of Evidence C*).
3. Athletes with aortic dimensions mildly above the normal range (z scores 2 to 2.5 or aortic root diameters measuring 40 to 41 mm in tall men or 36 to 38 mm in tall women) and no features of Marfan syndrome, Loeys-Dietz syndrome, or familial TAA syndrome should undergo echocardiographic or MRA surveillance every 6 to 12 months, with imaging frequency dependent on aortic size and stability of measurements (*Class I; Level of Evidence C*).
4. Athletes with BAV can participate in all competitive athletics if the aortic root and ascending aorta are not dilated (i.e., z score <2, or <2 standard deviations from the mean, or <40 mm in adults). The function of the BAV (whether stenotic or regurgitant) is also important in determining participation recommendations (see Task Force 5 on valvular heart disease [30]) (*Class I; Level of Evidence C*).
5. Athletes with BAV and aortic dimensions above the normal range (scores 2 to 3 or aortic diameters measuring 40 to 42 mm in men or 36 to 39 mm in women) should undergo echocardiographic or MRA surveillance of the aorta every 12 months, with more frequent imaging recommended for increasing aortic z score (*Class I; Level of Evidence C*).
6. It is reasonable for athletes with Marfan syndrome to participate in low and moderate static/low dynamic competitive sports (classes IA and IIA; see definition of sports classification in Task Force 1 report [31]) if they do not have ≥ 1 of the following (*Class IIa; Level of Evidence C*):
 - a. Aortic root dilatation (i.e., z score >2, or aortic diameter >40 mm, or >2 standard deviations from the mean relative to BSA in children or adolescents <15 years old)
 - b. Moderate to severe mitral regurgitation
 - c. Left ventricular systolic dysfunction (ejection fraction <40%)
 - d. Family history of aortic dissection at an aortic diameter <50 mm
7. It is reasonable for athletes with an unexplained TAA, familial TAA syndrome, or known pathogenic mutation leading to familial TAA syndrome (*ACTA2*, *MYH11*, *FBN1*, *TGFBR1*, *TGFBR2*, *MLCK*, *SMAD3*, *TGFB2*, and others) to participate in low static, low dynamic competitive sports (class IA) if they do not have ≥ 1 of the following (*Class IIa; Level of Evidence C*):
 - a. Aortic root dilatation (i.e., score >2, or aortic diameter >40 mm, or >2 standard deviations from the mean relative to BSA for children and adolescents <15 years old)

- b. Moderate to severe mitral regurgitation
 - c. Family history of aortic dissection
 - d. Cerebrovascular disease
 - e. Branch vessel aneurysm or dissection
8. It is reasonable for athletes with Loeys-Dietz syndrome or vascular Ehlers-Danlos syndrome to participate in low static, low dynamic sports (class IA) if they do not have any of the following (*Class IIa; Level of Evidence C*):
 - a. Aortic enlargement (score >2) or dissection, or branch vessel enlargement
 - b. Moderate to severe mitral regurgitation
 - c. Extracardiac organ system involvement that makes participation hazardous
 9. It is reasonable for athletes with surgical correction of the aortic root or ascending aorta for aneurysm disease or dissection and no evidence of residual aortic enlargement or dissection to participate in low static, low dynamic sports (class IA) that do not include the potential for bodily collision (*Class IIa; Level of evidence C*).
 10. For athletes with a BAV and a mild to moderately dilated aorta (score 2 to 3.5 or aortic root or ascending aortic diameters measuring 40 to 42 mm in men or 36 to 39 mm in women) and no features of associated connective tissue disorder or familial TAA syndrome, participation in low and moderate static and dynamic competitive sports with a low likelihood of significant bodily contact (classes IA, IB, IC, IIA, IIB, and IIC) may be considered. For these athletes, avoidance of intense weight training should be considered (*Class IIb; Level of Evidence C*).
 11. For athletes with aortic dimensions mildly above the normal range (scores 2 to 2.5 or aortic root diameters measuring 40 to 41 mm in tall men or 35 to 37 mm in tall women) and no features of Marfan syndrome, Loeys-Dietz syndrome, familial TAA syndrome, or BAV, participation in all competitive athletics may be considered after a comprehensive evaluation for an underlying genetic condition associated with aortopathy is performed. This may include analysis for mutations in *FBN1* and other genes associated with aortopathies in certain circumstances (*Class IIb; Level of Evidence C*).
 12. For athletes with aortic dimensions mildly above the normal range (scores 2 to 2.5 or aortic root diameters measuring 40 to 41 mm in tall men or 35 to 37 mm in tall women) and no features of Marfan syndrome, Loeys-Dietz syndrome, familial TAA syndrome, or BAV, avoidance of intense weight training may be considered (*Class IIb; Level of Evidence C*).
 13. For athletes with a BAV and a dilated aorta measuring 43 to 45 mm, participation in low-intensity competitive sports (class IA) with a low likelihood of bodily contact may be considered (*Class IIb; Level of Evidence C*).
 14. Athletes with Marfan syndrome, familial TAA syndrome, Loeys-Dietz syndrome, unexplained aortic aneurysm, vascular Ehlers-Danlos syndrome, or a related aortic aneurysm disorder should not participate in any competitive sports that involve intense physical exertion or the potential for bodily collision (*Class III; Level of Evidence C*).
 15. Athletes with BAV and a severely dilated aorta (score >3.5 to 4 or >43 mm in men or >40 mm in women) should not participate in any competitive sports that involve the potential for bodily collision (*Class III; Level of Evidence C*).
 16. Athletes with BAV and a markedly dilated aorta (>45 mm) should not participate in any competitive sports (*Class III; Level of Evidence C*).
 17. Athletes with chronic aortic dissection or branch vessel arterial aneurysm or dissection should not participate in any competitive sports (*Class III; Level of Evidence C*).

DISCLOSURES

Writing Group Disclosures

Writing Group Member	Employment	Research Grant	Other Research Support	Speakers Bureau/Honoraria	Expert Witness	Ownership Interest	Consultant/Advisory Board	Other
Alan C. Braverman	Washington University	None	None	None	None	None	None	None
Kevin M. Harris	Minneapolis Heart Institute Foundation at Abbott Northwestern Hospital	None	None	None	None	None	None	None
Richard J. Kovacs	Indiana University	None	None	None	None	None	None	None
Barry J. Maron	Minneapolis Heart Institute Foundation	None	None	None	None	None	None	None

This table represents the relationships of writing group members that may be perceived as actual or reasonably perceived conflicts of interest as reported on the Disclosure Questionnaire, which all members of the writing group are required to complete and submit. A relationship is considered to be "significant" if (a) the person receives \$10,000 or more during any 12-month period, or 5% or more of the person's gross income; or (b) the person owns 5% or more of the voting stock or share of the entity, or owns \$10,000 or more of the fair market value of the entity. A relationship is considered to be "modest" if it is less than "significant" under the preceding definition.

Reviewer Disclosures

Reviewer	Employment	Research Grant	Other Research Support	Speakers Bureau/Honoraria	Expert Witness	Ownership Interest	Consultant/Advisory Board	Other
Gregory Piazza	Brigham and Women's Hospital	None	None	None	None	None	None	None
David I. Silverman	Hartford Hospital	None	None	None	None	None	None	None
Karen K. Stout	University of Washington	None	None	None	None	None	None	None
Lars G. Svensson	Cleveland Clinic	None	None	None	None	None	None	None
Luciana T. Young	Ann & Robert H. Lurie Children's Hospital of Chicago/Northwestern University	None	None	None	None	None	None	None

This table represents the relationships of reviewers that may be perceived as actual or reasonably perceived conflicts of interest as reported on the Disclosure Questionnaire, which all reviewers are required to complete and submit. A relationship is considered to be "significant" if (a) the person receives \$10,000 or more during any 12-month period, or 5% or more of the person's gross income; or (b) the person owns 5% or more of the voting stock or share of the entity, or owns \$10,000 or more of the fair market value of the entity. A relationship is considered to be "modest" if it is less than "significant" under the preceding definition.

REFERENCES

- Maron BJ, Doerer JJ, Haas TS, Tierney DM, Mueller FO. Sudden deaths in young competitive athletes: analysis of 1866 deaths in the United States, 1980-2006. *Circulation*. 2009;119:1085-92. <http://dx.doi.org/10.1161/CIRCULATIONAHA.108.804617>.
- Loeys BL, Dietz HC, Braverman AC, Callewaert BL, De Backer J, Devereux RB, Hilhorst-Hofstee Y, Jondeau G, Faivre L, Milewicz DM, Pyeritz RE, Sponseller PD, Wordsworth P, De Paepe AM. The revised Ghent nosology for the Marfan syndrome. *J Med Genet*. 2010;47:476-85. <http://dx.doi.org/10.1136/jmg.2009.072785>.
- Loeys BL, Schwarze U, Holm T, Callewaert BL, Thomas GH, Pannu H, De Backer JF, Oswald GL, Symoens S, Manouvrier S, Roberts AE, Faravelli F, Greco MA, Pyeritz RE, Milewicz DM, Coucke PJ, Cameron DE, Braverman AC, Byers PH, De Paepe AM, Dietz HC. Aneurysm syndromes caused by mutations in the TGF-beta receptor. *N Engl J Med*. 2006;355:788-98. <http://dx.doi.org/10.1056/NEJMoa055695>.
- Hiratzka LF, Bakris GL, Beckman JA, Bersin RM, Carr VF, Casey DE Jr., Eagle KA, Hermann LK, Isselbacher EM, Kazerooni EA, Kouchoukos NT, Lytle BW, Milewicz DM, Reich DL, Sen S, Shinn JA, Svensson LG, Williams DM. 2010 ACCF/AHA/AATS/ACR/ASA/SCA/SCAI/SIR/STS/SVM guidelines for the diagnosis and management of patients with thoracic aortic disease: a report of the American College of Cardiology Foundation/American Heart Association Task Force on Practice Guidelines, American Association for Thoracic Surgery, American College of Radiology, American Stroke Association, Society of Cardiovascular Anesthesiologists, Society for Cardiovascular Angiography and Interventions, Society of Interventional Radiology, Society of Thoracic Surgeons, and Society for Vascular Medicine [published correction appears in *J Am Coll Cardiol*. 2010;62:1039]. *J Am Coll Cardiol*. 2010;55:e27-130. <http://dx.doi.org/10.1016/j.jacc.2010.02.015>.
- Milewicz DM, Regalado ES, Guo DC. Treatment guidelines for thoracic aortic aneurysms and dissections based on the underlying causative gene. *J Thorac Cardiovasc Surg*. 2010;140 suppl:S2-4. <http://dx.doi.org/10.1016/j.jtcvs.2010.07.027>.
- Braverman AC. Aortic involvement in patients with a bicuspid aortic valve. *Heart*. 2011;97:506-13. <http://dx.doi.org/10.1136/hrt.2009.183871>.
- Michelen HI, Khanna AD, Mahoney D, Margaryan E, Topilsky Y, Suri RM, Eidem B, Edwards WD, Sundt TM 3rd, Enriquez-Sarano M. Incidence of aortic complications in patients with bicuspid aortic valves. *JAMA*. 2011;306:1104-12. <http://dx.doi.org/10.1001/jama.2011.1286>.
- Devereux RB, de Simone G, Arnett DK, Best LG, Boerwinkle E, Howard BV, Kitzman D, Lee ET, Mosley TH Jr., Weder A, Roman MJ. Normal limits in relation to age, body size and gender of two-dimensional echocardiographic aortic root dimensions in persons ≥15 years of age. *Am J Cardiol*. 2012;110:1189-94. <http://dx.doi.org/10.1016/j.amjcard.2012.05.063>.
- Biaggi P, Matthews F, Braun J, Rousson V, Kaufmann PA, Jenni R. Gender, age, and body surface area are the major determinants of ascending aorta dimensions in subjects with apparently normal echocardiograms. *J Am Soc Echocardiogr*. 2009;22:720-5. <http://dx.doi.org/10.1016/j.echo.2009.03.012>.
- 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.

11. Colan SD, McElhinney DB, Crawford EC, Keane JF, Lock JE. Validation and re-evaluation of a discriminant model predicting anatomic suitability for biventricular repair in neonates with aortic stenosis. *J Am Coll Cardiol*. 2006;47:1858-65. <http://dx.doi.org/10.1016/j.jacc.2006.02.020>.
12. Chubb H, Simpson JM. The use of Z-scores in paediatric cardiology. *Ann Pediatr Cardiol*. 2012;5:179-84. <http://dx.doi.org/10.4103/0974-2069.99622>.
13. Lang RM, Bierig M, Devereux RB, Flachskampf FA, Foster E, Pellikka PA, Picard MH, Roman MJ, Seward J, Shanewise JS, Solomon SD, Spencer KT, Sutton MS, Stewart WJ, Chamber Quantification Writing Group, American Society of Echocardiography's Guidelines and Standards Committee, European Association of Echocardiography. Recommendations for chamber quantification: a report from the American Society of Echocardiography's Guidelines and Standards Committee and the Chamber Quantification Writing Group, developed in conjunction with the European Association of Echocardiography, a branch of the European Society of Cardiology. *J Am Soc Echocardiogr*. 2005; 18:1440-63. <http://dx.doi.org/10.1016/j.echo.2005.10.005>.
14. Campens L, Demulier L, De Groote K, Vandekerckhove K, De Wolf D, Roman MJ, Devereux RB, De Paepe A, De Backer J. Reference values for echocardiographic assessment of the diameter of the aortic root and ascending aorta spanning all age categories. *Am J Cardiol*. 2014;114:914-20. <http://dx.doi.org/10.1016/j.amjcard.2014.06.024>.
15. D'Andrea A, Cocchia R, Riegler L, Scarafie R, Salerno G, Gravino R, Vriz O, Citro R, Limongelli G, Di Salvo G, Cuomo S, Caso P, Russo MG, Calabrò R, Bossone E. Aortic root dimensions in elite athletes. *Am J Cardiol*. 2010;105:1629-34. <http://dx.doi.org/10.1016/j.amjcard.2010.01.028>.
16. Pelliccia A, Di Paolo FM, De Blasiis E, Quattrini FM, Pisicchio C, Guerra E, Culasso F, Maron BJ. Prevalence and clinical significance of aortic root dilatation in highly trained competitive athletes. *Circulation*. 2010;122:698-706. <http://dx.doi.org/10.1161/CIRCULATIONAHA.109.901074>.
17. Pelliccia A, Di Paolo FM, Quattrini FM. Aortic root dilatation in athletic population. *Prog Cardiovasc Dis*. 2012;54:432-7. <http://dx.doi.org/10.1016/j.pcad.2012.01.004>.
18. Babaei Bigi MA, Aslani A. Aortic root size and prevalence of aortic regurgitation in elite strength trained athletes. *Am J Cardiol*. 2007;100:528-30. <http://dx.doi.org/10.1016/j.amjcard.2007.02.108>.
19. Iskandar A, Thompson PD. A meta-analysis of aortic root size in elite athletes. *Circulation*. 2013;127: 791-8. <http://dx.doi.org/10.1161/CIRCULATIONAHA.112.000974>.
20. Reed CM, Richey PA, Pulliam DA, Somes GW, Alpert BS. Aortic dimensions in tall men and women. *Am J Cardiol*. 1993;71:608-10.
21. Braverman AC. Medical management of thoracic aortic aneurysm disease. *J Thorac Cardiovasc Surg*. 2013;145 suppl:S2-6. <http://dx.doi.org/10.1016/j.jtcvs.2012.11.062>.
22. Hatzaras I, Tranquilli M, Coady M, Barrett PM, Bible J, Elefteriades JA. Weight lifting and aortic dissection: more evidence for a connection. *Cardiology*. 2007;107: 103-6. <http://dx.doi.org/10.1159/000094530>.
- 22a. Harris KM, Tung M, Haas TS, Maron BJ. Under-recognition of aortic and aortic valve disease and the risk for sudden death in competitive athletes. *J Am Coll Cardiol*. 2015;65:860-2. <http://dx.doi.org/10.1016/j.jacc.2014.09.094>.
23. Parish LM, Gorman JH 3rd, Kahn S, Plappert T, St John-Sutton MG, Bavaria JE, Gorman RC. Aortic size in acute type A dissection: implications for preventive ascending aortic replacement. *Eur J Cardiothorac Surg*. 2009;35:941-5. <http://dx.doi.org/10.1016/j.ejcts.2008.12.047>.
24. Pape LA, Tsai TT, Isselbacher EM, Oh JK, O'Gara PT, Evangelista A, Fattori R, Meinhardt G, Trimarchi S, Bossone E, Suzuki T, Cooper JV, Froehlich JB, Nienaber CA, Eagle KA, International Registry of Acute Aortic Dissection (IRAD) Investigators. Aortic diameter ≥ 5.5 cm is not a good predictor of type A aortic dissection: observations from the International Registry of Acute Aortic Dissection (IRAD). *Circulation*. 2007;116:1120-7. <http://dx.doi.org/10.1161/CIRCULATIONAHA.107.702720>.
25. Engelfriet PM, Boersma E, Tijssen JG, Bouma BJ, Mulder BJ. Beyond the root: dilatation of the distal aorta in Marfan's syndrome. *Heart*. 2006;92:1238-43. <http://dx.doi.org/10.1136/hrt.2005.081638>.
26. Mimoun L, Detaint D, Hamroun D, Arnoult F, Delorme G, Gautier M, Milleron O, Meuleman C, Raoux F, Boileau C, Vahanian A, Jondeau G. Dissection in Marfan syndrome: the importance of the descending aorta. *Eur Heart J*. 2011;32:443-9. <http://dx.doi.org/10.1093/eurheartj/ehq434>.
27. Kari FA, Fazel SS, Mitchell RS, Fischbein MP, Miller DC. Bicuspid aortic valve configuration and aortopathy pattern might represent different pathophysiologic substrates. *J Thorac Cardiovasc Surg*. 2012; 144:516-7. <http://dx.doi.org/10.1016/j.jtcvs.2012.05.035>.
28. Maron BJ, Ackerman MJ, Nishimura RA, Pyeritz RE, Towbin JA, Udelson JE. Task Force 4: HCM and other cardiomyopathies, mitral valve prolapse, myocarditis, and Marfan syndrome. *J Am Coll Cardiol*. 2005;45: 1340-5. <http://dx.doi.org/10.1016/j.jacc.2005.02.011>.
29. Kinoshita N, Mimura J, Obayashi C, Katsukawa F, Onishi S, Yamazaki H. Aortic root dilatation among young competitive athletes: echocardiographic screening of 1929 athletes between 15 and 34 years of age. *Am Heart J*. 2000;139:723-8.
30. Bonow RO, Nishimura RA, Thompson PD, Udelson JE, on behalf of the American Heart Association Electrocardiography and Arrhythmias Committee of the Council on Clinical Cardiology, Council on Cardiovascular Disease in the Young, Council on Cardiovascular and Stroke Nursing, Council on Functional Genomics and Translational Biology, and the American College of Cardiology. Eligibility and disqualification recommendations for competitive athletes with cardiovascular abnormalities: Task Force 5: valvular heart disease: a scientific statement from the American Heart Association and American College of Cardiology. *J Am Coll Cardiol*. 2015 In Press. <http://dx.doi.org/10.1016/j.jacc.2015.09.037>.
31. Levine BD, Baggish AL, Kovacs RJ, Link MS, Maron MS, Mitchell JH, on behalf of the American Heart Association Electrocardiography and Arrhythmias Committee of the Council on Clinical Cardiology, Council on Cardiovascular Disease in the Young, Council on Cardiovascular and Stroke Nursing, Council on Functional Genomics and Translational Biology, and the American College of Cardiology. Eligibility and disqualification recommendations for competitive athletes with cardiovascular abnormalities: Task Force 1: classification of sports: dynamic, static, and impact: a scientific statement from the American Heart Association and American College of Cardiology. *J Am Coll Cardiol*. 2015 In Press. <http://dx.doi.org/10.1016/j.jacc.2015.09.033>.

KEY WORDS ACC/AHA Scientific Statements, aortic diseases, athletes, bicuspid aortic valve, familial thoracic aortic aneurysm, Marfan syndrome

Eligibility and Disqualification Recommendations for Competitive Athletes With Cardiovascular Abnormalities: Task Force 8: Coronary Artery Disease

James E. Udelson, MD, FAHA, FACC*
Richard J. Kovacs, MD, FAHA, FACC*

exercise (1). There is universal agreement that vigorous exercise, such as athletic competition, acutely, albeit transiently, increases the risk of SCD and AMI in previously healthy people (1). Vigorous exercise also transiently increases the risk for SCD and AMI in people with diagnosed ASCAD. These events may be caused by plaque disruption, but SCD in these patients may also be produced by malignant arrhythmias caused by demand ischemia or originating in areas of myocardial scar (1). In addition to ASCAD, other coronary conditions such as coronary

Permissions: Multiple copies, modification, alteration, enhancement, and/or distribution of this document are not permitted without the express permission of the American College of Cardiology. Requests may be completed online via the Elsevier site (<http://www.elsevier.com/about/policies/author-agreement/obtaining-permission>).

vasospasm, myocardial bridging, and coronary dissection, as well as infection such as Kawasaki disease, vasculitis, and cardiac transplant vasculopathy, may also cause acute cardiac events during exercise. The present section makes recommendations on how to evaluate patients with disease of the coronary arteries and make appropriate recommendations for athletic competition. Anomalous coronary arteries are considered in the Task Force 4 report (2).

We searched PubMed for English language articles reporting exercise-related issues related to coronary diseases. This search produced no clinical trials examining how competitive athletes with coronary artery diseases should be advised regarding vigorous exercise in general or athletic competition in particular. Consequently, the following recommendations are based on case series, case reports, and consensus among the committee members.

ATHEROSCLEROTIC CORONARY ARTERY DISEASE

Patients with ASCAD can be divided into clinically manifest or symptomatic and clinically concealed or asymptomatic subgroups. The former have either experienced an acute cardiac event or have symptoms consistent with inducible myocardial ischemia, or they have findings of ischemia identified by a diagnostic testing modality such as exercise testing with or without adjunctive nuclear or echocardiographic imaging. This group includes those with “silent ischemia” who have no symptoms but have ischemia documented by provocative testing. Patients with clinically concealed ASCAD are presently and previously asymptomatic and are diagnosed as having ASCAD by the presence of coronary artery calcification on computerized tomography or by the presence of non-calcified plaque by coronary computed tomography angiography but do not have evidence of ischemia on provocative testing.

Evaluation and recommendations for patients with ASCAD are based on the following assumptions: 1) The risk of an acute exertion-related cardiac event is greater in those who have had a previous acute cardiac syndrome and lower in those whose ASCAD is clinically silent and was diagnosed by such techniques as coronary artery calcification scanning or computed tomography angiography. 2) The risk of an acute exertion-related cardiac event increases with increasing extent of coronary artery disease, reduced left ventricular systolic function, the presence and extent of ischemia, and increased electrical instability. Unstable or “vulnerable” plaques are often lipid rich (3), so it is also likely that the risk of an exertion-related plaque disruption can be reduced by aggressive lipid-lowering treatment, which has been shown to

reduce the lipid content of atherosclerotic plaques (4). 3) Patients with clinically manifest ASCAD should strongly consider deferring their possible return to athletic competition to permit lesion regression and regression of lipid from the plaque. The length of this delay is not defined, but some have suggested 2 years, because substantial lesion regression has been documented to occur within 2 years of aggressive lipid management (5).

Recommendations

1. **Athletes with ASCAD should undergo maximal exercise testing to evaluate exercise tolerance, the presence of inducible ischemia, and the presence of exercise-induced electrical instability. Testing should be performed on the subject’s standard medical regimen, including β -adrenergic blocking medications (Class I; Level of Evidence C).**
2. **Athletes with ASCAD should undergo an evaluation of left ventricular function (Class I; Level of Evidence C).**
3. **Once informed of the results of the evaluations contained in recommendations 1 and 2, adult patients with ASCAD should participate in the decision as to whether the health and psychological benefits of exercise for them outweigh the risk (Class I; Level of Evidence C).**
4. **Athletes with ASCAD should undergo aggressive risk factor reduction with high-intensity statin therapy to reduce the chance of plaque disruption (6) (Class I; Level of Evidence A).**
5. **It is reasonable for athletes with clinically concealed ASCAD to participate in all competitive activities if their resting left ventricular ejection fraction is >50% and they have no inducible ischemia or electrical instability (Class IIb; Level of Evidence C).**
6. **It is reasonable for patients with clinically manifest ASCAD to participate in all competitive activities if their resting left ventricular ejection fraction is >50%, they are asymptomatic, and they have no inducible ischemia or electrical instability (Class IIb; Level of Evidence C).**
7. **It is reasonable to restrict patients with clinically manifest ASCAD that does not fulfill the criteria in recommendation 6 to sports with low dynamic and low to moderate static demands (Class IIb; Level of Evidence C).**
8. **It is reasonable to prohibit patients with clinically manifest ASCAD from competitive sport participation:**
 - a. **For at least 3 months after an AMI or coronary revascularization procedure (Class IIb; Level of Evidence C);**
 - b. **If they have increasing frequency or worsening symptoms of myocardial ischemia (Class IIb; Level of Evidence C).**

CORONARY ARTERY SPASM

Focal coronary artery spasm, usually in the presence of various degrees of coronary atherosclerosis, is a defined but uncommon cause of life-threatening arrhythmias and SCD (7,8). It can also be identified in the absence of identifiable atherosclerotic lesions by provocation studies (9). Coronary artery spasm in its classic form usually occurs with little or minimally obstructive coronary artery lesions. Although exercise-induced spasm during stress testing has been documented, it is uncommon. In most instances, it was induced during pharmacological stress tests with either dobutamine or adenosine. Reports of cardiac arrest survivors in whom coronary artery spasm has been identified as the mechanism of cardiac arrest are limited (10), although the presence of coronary vasomotor spasm identifies people with a higher risk of sudden death than the general population (11). However, susceptibility to spasm is not constant over time, being dependent on the state of the endothelium. Finally, there are also few data to suggest a specific propensity to coronary artery spasm and consequent arrhythmias in competitive athletes. When coronary vasospasm is identified or strongly suspected during exercise, treatment should be initiated with calcium blockers and nitrates to reduce the possibility of spasm and to control symptoms.

Recommendations

1. It is reasonable to restrict the small subset with silent ischemia caused by coronary artery spasm who have had documented life-threatening arrhythmias and in whom the absence of clinical pain impedes identification of an adequate response to therapy (12) to sports with low dynamic and low to moderate static demands (Class IIa; Level of Evidence C).
2. It is reasonable that athletes whose symptoms and objective evidence of spasm can be controlled with medications be allowed to participate in all levels of competition (Class IIb; Level of Evidence C).

SPONTANEOUS CORONARY ARTERY DISSECTION

Spontaneous coronary artery dissection refers to dissection of the coronary arteries without underlying atherosclerosis (13). Spontaneous coronary artery dissection is associated with late pregnancy and the peripartum state, female hormonal therapy, Marfan syndrome, exercise, chest trauma (13), and fibromuscular dysplasia (14). It is a rare cause of exercise-related cardiac events but should be considered in any young person who develops an acute cardiac syndrome during vigorous exercise or after sports-related chest trauma.

Recommendation

1. There are insufficient data to provide definitive recommendations for sports participation, but because spontaneous dissection can occur with exertion, it is reasonable that patients with prior spontaneous coronary artery dissection be restricted to participation in sports with low to moderate dynamic and low to moderate static demands (Class IIa; Level of Evidence C).

MYOCARDIAL BRIDGING

Myocardial bridging is diagnosed when a portion of a major epicardial coronary artery is completely covered by myocardium. Myocardial bridging is commonly observed by angiography as coronary artery compression during systole. It is usually asymptomatic and of no clinical consequence but has been rarely associated with exercise-induced ischemia and exercise-related acute cardiac events (15). Pathological studies suggest that vessels whose tunneled length is long and deeper than 3 mm beneath the epicardium create the greatest vulnerability for cardiac events.

Recommendations

1. It is reasonable for athletes with myocardial bridging and no evidence of myocardial ischemia during adequate stress testing to participate in all competitive sports (Class IIa; Level of Evidence C).
2. It is reasonable to restrict athletes with myocardial bridging of an epicardial coronary artery and objective evidence of myocardial ischemia or prior myocardial infarction to sports with low to moderate dynamic and low to moderate static demands (Class IIa; Level of Evidence C).
3. It is reasonable to restrict athletes who have undergone surgical resection of the myocardial bridge or stenting of the bridge to low-intensity sports for 6 months after the procedure. If such athletes have no subsequent evidence of ischemia, they may participate in all competitive sports (Class IIa; Level of Evidence C).

KAWASAKI DISEASE

Kawasaki disease is an acute febrile illness of unknown pathogenesis that is among the leading causes of acquired heart disease in children. Kawasaki disease can produce coronary artery aneurysms that predispose to myocardial ischemia, myocardial infarction, and SCD. Aneurysms can be divided by their internal diameter into small (<1.5 times normal, or <5 mm), moderate (1.5 to 4 times normal, or 5 to 8 mm), and large (>4 times normal, or >8 mm) aneurysms (16). Prompt recognition and treatment of the acute phase of Kawasaki disease can reduce the cardiac

complications, but 20% of untreated people and 4% of those treated with aspirin and intravenous immunoglobulin still develop coronary artery aneurysms (17). Risk scores for prediction of coronary artery aneurysm development are imperfect, and the broad use of intravenous immunoglobulin is recommended. Ongoing surveillance of patients after the acute phase is recommended, including serial stress tests in those patients with manifest coronary artery disease (18). Treatment of coronary artery aneurysms with antiplatelet agents, anticoagulant agents, or myocardial revascularization must be considered in evaluating decisions about a patient's return to competition.

Recommendations

1. Patients with ≥ 1 large coronary aneurysms should continue antiplatelet therapy and possibly anticoagulant therapy. It is also reasonable for annual stress tests to be performed and activity to be guided by results, similar to adults with ASCAD (*Class I; Level of Evidence C*).
2. Patients with myocardial infarction or revascularization should follow the guidance for adults with ASCAD (*Class I; Level of Evidence A*).
3. Collision sports should be avoided in patients undergoing antiplatelet therapy (*Class I; Level of Evidence C*).
4. In the absence of exercise-induced ischemia or arrhythmias, it is reasonable for patients to participate in low- to moderate-intensity static and dynamic competitive sports. Patients with persistent small to medium-sized aneurysms in ≥ 1 coronary arteries should continue antiplatelet therapy and undergo ongoing surveillance (*Class IIa; Level of Evidence C*).
5. Patients with no coronary aneurysms during the convalescent phase and with no exercise-induced ischemia or arrhythmias may be considered for participation in all sports starting 8 weeks after the illness has resolved (*Class IIb; Level of Evidence C*).
6. Patients with transient coronary aneurysms and with no exercise-induced ischemia or arrhythmias may be considered for participation in all sports 8 weeks after illness resolution. Risk reassessment is recommended at 3- to 5-year intervals or according to current guidelines (*Class IIb; Level of Evidence C*).

CORONARY VASCULITIS

Coronary vasculitis attributable to causes other than Kawasaki disease may affect competitive athletes of any age but is rare. These diseases include polyarteritis nodosa, Takayasu arteritis, Buerger disease, and other specific and nonspecific forms of coronary arteritis (16). SCD has been reported in previously healthy young people with

unsuspected coronary vasculitis at autopsy (17). In a series of 50 cases of SCD associated with nonatherosclerotic coronary pathology (12 of whom died during or immediately after physical exertion), 6 of the 50 (12%) had autopsy evidence of coronary vasculitis (18).

There is no evidence that athletes are predisposed to coronary vasculitis at rates higher than the general age-corrected population or that the course of these athletes' disease is any different from that of the general population. There is no information in the medical literature to suggest care of the athlete should differ.

Recommendations

1. Athletes who have recovered from coronary vasculitis can participate in all sports without restriction (*Class I; Level of Evidence C*).
2. Athletes with coronary vasculitis are likely at increased risk for acute cardiac events during training or competition. It is reasonable to restrict participation in sports until the vasculitis has resolved (*Class IIa; Level of Evidence C*).

CARDIAC TRANSPLANT CORONARY VASCULOPATHY

The coronary arteries of orthotopic transplanted hearts develop a diffuse vasculopathy that is the leading cause of death in transplant recipients. Because the transplanted heart is initially denervated, recipients require surveillance, because they may not experience classic symptoms of cardiac ischemia. Our literature search did not detect any reports of exercise-related cardiac events in cardiac transplant recipients either because transplant vasculopathy is not associated with the same increase in exercise events as classic atherosclerosis or because too few transplant patients have participated in competitive events to highlight this issue.

Recommendations

1. The transplant cardiologist should make the final recommendations for athletic participation for cardiac transplant recipients (*Class I; Level of Evidence C*).
2. It is reasonable for cardiac transplant recipients participating in competitive athletics to 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 regimen (*Class IIa; Level of Evidence C*).
3. It is reasonable for cardiac transplant recipients with an ejection fraction $>50\%$, no evidence of cardiac ischemia, and no electrical instability to participate in all competitive activities commensurate with their exercise tolerance (*Class IIa; Level of Evidence C*).

DISCLOSURES

Writing Group Disclosures

Writing Group Member	Employment	Research Grant	Other Research Support	Speakers Bureau/Honoraria	Expert Witness	Ownership Interest	Consultant/Advisory Board	Other
Paul D. Thompson	Hartford Hospital	None	None	None	None	None	None	None
Richard J. Kovacs	Indiana University	None	None	None	None	None	None	None
Benjamin D. Levine	University of Texas Southwestern Medical Center	NSBRI*	None	None	None	None	None	None
Robert J. Myerburg	University of Miami	None	None	None	None	None	None	None
James E. Udelson	Tufts Medical Center	None	None	None	None	None	None	None

This table represents the relationships of writing group members that may be perceived as actual or reasonably perceived conflicts of interest as reported on the Disclosure Questionnaire, which all members of the writing group are required to complete and submit. A relationship is considered to be "significant" if (a) the person receives \$10,000 or more during any 12-month period, or 5% or more of the person's gross income; or (b) the person owns 5% or more of the voting stock or share of the entity, or owns \$10,000 or more of the fair market value of the entity. A relationship is considered to be "modest" if it is less than "significant" under the preceding definition.

*Modest.

Reviewer Disclosures

Reviewer	Employment	Research Grant	Other Research Support	Speakers Bureau/Honoraria	Expert Witness	Ownership Interest	Consultant/Advisory Board	Other
Ami B. Bhatt	Massachusetts General Hospital	None	None	None	None	None	None	None
Stephan D. Fihn	VA Puget Sound Health Care System	None	None	None	None	None	None	None
Robert A. Vogel	Pritikin, University of Maryland	NFL†	None	None	None	None	None	None

This table represents the relationships of reviewers that may be perceived as actual or reasonably perceived conflicts of interest as reported on the Disclosure Questionnaire, which all reviewers are required to complete and submit. A relationship is considered to be "significant" if (a) the person receives \$10,000 or more during any 12-month period, or 5% or more of the person's gross income; or (b) the person owns 5% or more of the voting stock or share of the entity, or owns \$10,000 or more of the fair market value of the entity. A relationship is considered to be "modest" if it is less than "significant" under the preceding definition.

†Significant.

REFERENCES

- Parker MW, Thompson PD. Assessment and management of atherosclerosis in the athletic patient. *Prog Cardiovasc Dis*. 2012;54:416-22. <http://dx.doi.org/10.1016/j.pcad.2012.02.001>.
- Van Hare GF, Ackerman MJ, Evangelista JK, Kovacs RJ, Myerburg RJ, Shafer KM, Warnes CA, Washington RL, on behalf of the American Heart Association Electrocardiography and Arrhythmias Committee of the Council on Clinical Cardiology, Council on Cardiovascular Disease in the Young, Council on Cardiovascular and Stroke Nursing, Council on Functional Genomics and Translational Biology, and the American College of Cardiology. Eligibility and disqualification recommendations for competitive athletes with cardiovascular abnormalities: Task Force 4: congenital heart disease: a scientific statement from the American Heart Association and American College of Cardiology. *J Am Coll Cardiol*. 2015 In Press. <http://dx.doi.org/10.1016/j.jacc.2015.09.036>.
- Libby P. Molecular and cellular mechanisms of the thrombotic complications of atherosclerosis. *J Lipid Res*. 2009;50 suppl:S352-7. <http://dx.doi.org/10.1194/jlr.R800099-JLR200>.
- Ballantyne CM, Raichlen JS, Nicholls SJ, Erbel R, Tardif JC, Brener SJ, Cain VA, Nissen SE, ASTEROID Investigators. Effect of rosuvastatin therapy on coronary artery stenoses assessed by quantitative coronary angiography: a study to evaluate the effect of rosuvastatin on intravascular ultrasound-derived coronary atheroma burden. *Circulation*. 2008;117:2458-66. <http://dx.doi.org/10.1161/CIRCULATIONAHA.108.773747>.
- Zhao XQ, Dong L, Hatsukami T, Phan BA, Chu B, Moore A, Lane T, Neradilek MB, Polissar N, Monick D, Lee C, Underhill H, Yuan C. MR imaging of carotid plaque composition during lipid-lowering therapy: a prospective assessment of effect and time course. *J Am Coll Cardiol Img*. 2011;4:977-86. <http://dx.doi.org/10.1016/j.jcmg.2011.06.013>.
- Stone NJ, Robinson JG, Lichtenstein AH, Bairey Merz CN, Blum CB, Eckel RH, Goldberg AC, Gordon D, Levy D, Lloyd-Jones DM, McBride P, Schwartz JS, Shero ST, Smith SC Jr., Watson K, Wilson PW. 2013 ACC/AHA guideline on the treatment of blood cholesterol to reduce atherosclerotic cardiovascular risk in adults: a report of the American College of Cardiology/American Heart Association Task Force on Practice Guidelines [published correction appears in *J Am Coll Cardiol*. 2014;63:3024-5]. *J Am Coll Cardiol*. 2014;63:2869-934. <http://dx.doi.org/10.1016/j.jacc.2013.11.002>.
- Yasue H, Takizawa A, Nagao M, Nishida S, Horie M, Kubota J, Omote S, Takaoka K, Okumura K. Long-term prognosis for patients with variant angina and influential factors. *Circulation*. 1988;78:1-9.
- Lanza GA, Sestito A, Sgueglia GA, Infusino F, Manolfi M, Crea F, Maseri A. Current clinical features, diagnostic assessment and prognostic determinants of patients with variant angina. *Int J Cardiol*. 2007;118:41-7. <http://dx.doi.org/10.1016/j.ijcard.2006.06.016>.
- Ong P, Athanasiadis A, Borgulya G, Mahrholdt H, Kaski JC, Sechtem U. High prevalence of a pathological response to acetylcholine testing in patients with stable angina pectoris and unobstructed coronary arteries: the ACOVA Study (Abnormal COronary VAsomotion in patients with stable angina and unobstructed coronary arteries). *J Am Coll Cardiol*. 2012;59:655-62. <http://dx.doi.org/10.1016/j.jacc.2011.11.015>.
- Krahn AD, Healey JS, Chauhan V, Birnie DH, Simpson CS, Champagne J, Gardner M, Sanatani S, Exner DV, Klein GJ, Yee R, Skanes AC, Gula LJ, Goltz MH. Systematic assessment of patients with unexplained cardiac arrest: Cardiac Arrest Survivors With Preserved Ejection Fraction Registry (CASPER) [published correction appears in *Circulation*. 2010;121:e460]. *Circulation*. 2009;120:278-85. <http://dx.doi.org/10.1161/CIRCULATIONAHA.109.853143>.

11. Takagi Y, Yasuda S, Tsunoda R, Ogata Y, Seki A, Sumiyoshi T, Matsui M, Goto T, Tanabe Y, Sueda S, Sato T, Ogawa S, Kubo N, Momomura S, Ogawa H, Shimokawa H, Japanese Coronary Spasm Association. Clinical characteristics and long-term prognosis of vasospastic angina patients who survived out-of-hospital cardiac arrest: multicenter registry study of the Japanese Coronary Spasm Association. *Circ Arrhythm Electrophysiol.* 2011;4:295–302. <http://dx.doi.org/10.1161/CIRCEP.110.959809>.
12. Myerburg RJ, Kessler KM, Mallon SM, Cox MM, deMarchena E, Interian A Jr., Castellanos A. Life-threatening ventricular arrhythmias in patients with silent myocardial ischemia due to coronary-artery spasm. *N Engl J Med.* 1992;326:1451–5. <http://dx.doi.org/10.1056/NEJM199205283262202>.
13. Kalaga RV, Malik A, Thompson PD. Exercise-related spontaneous coronary artery dissection: case report and literature review. *Med Sci Sports Exerc.* 2007;39:1218–20. <http://dx.doi.org/10.1249/mss.0b013e318060114f>.
14. Michelis KC, Olin JW, Kadian-Dodov D, d'Escamard V, Kovacic JC. Coronary artery manifestations of fibromuscular dysplasia. *J Am Coll Cardiol.* 2014;64:1033–46. <http://dx.doi.org/10.1016/j.jacc.2014.07.014>.
15. Baggish AL, Thompson PD. The Athlete's Heart 2007: diseases of the coronary circulation. *Cardiol Clin.* 2007;25:431–40, vi. <http://dx.doi.org/10.1016/j.ccl.2007.07.003>.
16. Kato H, Sugimura T, Akagi T, Sato N, Hashino K, Maeno Y, Kazue T, Eto G, Yamakawa R. Long-term consequences of Kawasaki disease: a 10- to 21-year follow-up study of 594 patients. *Circulation.* 1996;94:1379–85.
17. 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.
18. Newburger JW, Takahashi M, Gerber MA, Gewitz MH, Tani LY, Burns JC, Shulman ST, Bolger AF, Ferrieri P, Baltimore RS, Wilson WR, Baddour LM, Levison ME, Pallasch TJ, Falace DA, Taubert KA, Committee on Rheumatic Fever, Endocarditis, and Kawasaki Disease, Council on Cardiovascular Disease in the Young, American Heart Association. 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, American Heart Association [published correction appears in *Pediatrics.* 2005;115:1118]. *Pediatrics.* 2004;114:1708–33. <http://dx.doi.org/10.1542/peds.2004-2182>.

KEY WORDS ACC/AHA Scientific Statements, athletes, coronary artery disease, sudden cardiac death

AHA/ACC SCIENTIFIC STATEMENT

Eligibility and Disqualification Recommendations for Competitive Athletes With Cardiovascular Abnormalities: Task Force 9: Arrhythmias and Conduction Defects

A Scientific Statement From the American Heart Association and American College of Cardiology

Douglas P. Zipes, MD, FAHA, MACC,
*Chair**

Mark S. Link, MD, FACC*
 Michael J. Ackerman, MD, PhD,
 FACC*
 Richard J. Kovacs, MD, FAHA, FACC*

Robert J. Myerburg, MD, FACC*
 N.A. Mark Estes III, MD, FACC*

A broad range of variations in heart rates and rhythms, specific cardiac arrhythmias, and atrioventricular (AV) and intraventricular conduction disturbances are observed in athletes. Although most are common among nonathletes as well, the special circumstances and pressures related to athletic performance demand a high level of attention. The distinction between normal variants, often exaggerated by the specific physiology of the conditioned athlete, and arrhythmias

that may be symptomatic or life-threatening may be significant challenges.

BRADYCARDIA

Sinus Bradycardia

Sinus bradycardia, defined as a sinus rate <60 beats per minute (bpm), is common in the athlete (1). Generally, it is attributed to enhanced vagal tone

*On behalf of the American Heart Association Electrocardiography and Arrhythmias Committee of the Council on Clinical Cardiology, Council on Cardiovascular Disease in the Young, Council on Cardiovascular and Stroke Nursing, Council on Functional Genomics and Translational Biology, and the American College of Cardiology.

The American Heart Association and the American College of Cardiology make every effort to avoid any actual or potential conflicts of interest that may arise as a result of an outside relationship or a personal, professional, or business interest of a member of the writing panel. Specifically, all members of the writing group are required to complete and submit a Disclosure Questionnaire showing all such relationships that might be perceived as real or potential conflicts of interest. The Preamble and other Task Force reports for these proceedings are available online at www.onlinejacc.org (J Am Coll Cardiol 2015;XX:000-000; 000-000; 000-000; 000-000; 000-000; 000-000; 000-000; 000-000; 000-000; 000-000; 000-000; 000-000; and 000-000).

This statement was approved by the American Heart Association Science Advisory and Coordinating Committee on June 24, 2015, and the American Heart Association Executive Committee on July 22, 2015, and by the American College of Cardiology Board of Trustees and Executive Committee on June 3, 2015.

The American College of Cardiology requests that this document be cited as follows: Zipes DP, Link MS, Ackerman MJ, Kovacs RJ, Myerburg RJ, Estes NAM 3rd; on behalf of the American Heart Association Electrocardiography and Arrhythmias Committee of the Council on Clinical Cardiology, Council on Cardiovascular Disease in the Young, Council on Cardiovascular and Stroke Nursing, Council on Functional Genomics and Translational Biology, and the American College of Cardiology. Eligibility and disqualification recommendations for competitive athletes with cardiovascular abnormalities: Task Force 9: arrhythmias and conduction defects: a scientific statement from the American Heart Association and American College of Cardiology. J Am Coll Cardiol 2015;xx:•••-•••.

This article has been copublished in *Circulation*.

Copies: This document is available on the World Wide Web sites of the American Heart Association (<http://my.americanheart.org>) and the American College of Cardiology (www.acc.org). For copies of this document, please contact Elsevier Inc. Reprint Department via fax (212-633-3820) or e-mail (reprints@elsevier.com).

Permissions: Multiple copies, modification, alteration, enhancement, and/or distribution of this document are not permitted without the express permission of the American College of Cardiology. Requests may be completed online via the Elsevier site (<http://www.elsevier.com/about/policies/author-agreement/obtaining-permission>).

caused by conditioning and is thus physiological. Occasionally, heart rates can be as slow as 30 to 40 bpm at rest in the highly conditioned athlete and decrease to <30 bpm during sleep. Some athletes with marked sinus bradycardia will exhibit periods of low atrial or junctional escape rhythms with rates of 40 to 60 bpm. This is a normal phenomenon, and these will become suppressed with exercise-induced increases in the sinus rate.

Evaluation of the athlete with sinus bradycardia includes a careful history to determine whether the athlete has symptoms related to the bradycardia. In addition, physical examination and an ECG are warranted, with selective use of additional tests such as an echocardiogram and exercise stress test if underlying structural heart disease is suggested. Stress testing can also be used to verify a normal rate response to exercise, if judged to be necessary. The same approach applies to the sinus arrhythmia commonly observed in the athlete. Generally, asymptomatic sinus pauses or sinus arrest (<3 seconds) are not considered clinically significant unless accompanied by symptoms. Pauses of longer duration may fall within the spectrum of physiological responses to athletic conditioning; however, when accompanied by symptoms, sinus bradycardia, sinoatrial exit block, and sick sinus syndrome with pauses at the termination of a supraventricular tachycardia (SVT) are considered abnormal. Athletes with symptoms potentially associated with these arrhythmias should have an ECG, 24-hour ambulatory monitoring, and an exercise test. Clinical assessment for structural heart disease and noninvasive assessment of sinus node function with ambulatory monitoring and stress testing are also appropriate in symptomatic patients or those with resting heart rates <30 bpm or pauses >3 seconds.

Invasive electrophysiology studies (EPS) play a very limited role in the assessment of sinus node function. An athlete with symptoms related to sinus bradycardia caused by high vagal tone related to training should restrict athletic training and have clinical reassessment of symptoms and sinus node function (1). Patients with symptomatic bradycardia not responsive to other measures such as deconditioning or the withholding of nonessential medications that are contributing to the bradycardia may need to be treated with a permanent pacemaker, although this is very rarely needed in the athlete (2,3).

Recommendations

- 1. Athletes with sinus bradycardia, sinus exit block, sinus pauses, and sinus arrhythmia without symptoms can participate in all competitive athletic activities unless otherwise excluded by underlying structural heart disease or other arrhythmias (Class I; Level of Evidence C).**

- 2. Athletes with symptomatic bradycardia should be evaluated for structural heart disease and be treated for the bradycardia, generally by an implanted pacemaker. They should be restricted from training and athletic competition while being evaluated. If treatment of the bradycardia eliminates symptoms, they can participate in athletic training and competition unless otherwise excluded by structural heart disease or other arrhythmias (Class I; Level of Evidence C).**

AV BLOCK

Athletes with AV block should be assessed for symptoms attributable to the block with a history and for any underlying structural heart disease with a cardiovascular examination and ECG. Other tests, including an echocardiogram, ambulatory monitoring, exercise stress test, and invasive EPS, should be used in a selective fashion.

First-Degree AV Block

In asymptomatic athletes with structurally normal hearts who have first-degree AV block identified on a pre-participation or other incidental ECG, the PR interval will shorten during a stress test in most cases. However, stress testing is rarely necessary for the evaluation of an athlete with a PR interval <0.3 second and a normal QRS duration. An echocardiogram is not necessary unless the cardiovascular examination or ECG suggests structural heart disease. If the QRS complex is abnormal, or the PR interval is excessively prolonged (≥ 0.3 second), an exercise stress test, 24-hour ambulatory monitor, and echocardiogram are warranted. EPS is rarely necessary but might be performed in selected cases, such as those with exercise-induced AV block suspected of having type II AV block, to determine the site and duration of conduction delay (AV node or intra-His/infra-His) and ensure that the patient is not at risk for progression to higher-degree block that would cause symptoms. Patients with congenitally corrected transposition of the great arteries can exhibit first-degree AV block with very little else on physical examination.

Recommendations

- 1. Asymptomatic athletes with no structural heart disease and first-degree AV block (PR interval <0.3 ms) can participate in all competitive sports unless there are findings that indicate that the person is at risk for progression to higher-degree block that would symptoms (Class I; Level of Evidence C).**
- 2. Asymptomatic athletes with first-degree AV block, in whom type I second-degree AV block appears with exercise, should be evaluated further for possible intra-His or infra-His block with EPS (Class I; Level of Evidence C).**

- 3. If structural heart disease is present, athletic restrictions should be recommended as appropriate for the type of structural heart disease (Class I; Level of Evidence C).**

Type I Second-Degree (Wenckebach) AV Block

Wenckebach type I AV nodal block can be present in otherwise normal, well-trained endurance athletes. Type I second-degree AV block (i.e., Wenckebach) is observed more commonly during sleep in athletes than in the daytime when they are awake. Athletes should be assessed for symptoms attributable to the block and for any underlying structural heart disease with an echocardiogram. In asymptomatic or symptomatic athletes with Wenckebach block, a history, physical examination, ECG, echocardiogram, and exercise stress test may be considered. If the QRS complex is abnormal, or the shortest PR interval is excessively prolonged (≥ 0.3 second), 24-hour ECG recording or other ambulatory monitor is warranted. EPS is rarely necessary but might be performed in highly selected cases to determine the site and duration of conduction delay and ensure that the patient is not at risk for progression to higher-degree block that would cause symptoms.

Recommendations

- 1. Asymptomatic athletes with structurally normal hearts and Wenckebach AV block (type I second-degree AV block) with improvement in conduction with exercise or recovery can participate in all competitive sports (Class I; Level of Evidence C).**
- 2. Asymptomatic athletes with structurally abnormal hearts with improvement in Wenckebach AV block with exercise can participate in all competitive sports, unless there are restrictions based on heart disease (Class I; Level of Evidence C).**
- 3. Athletes with Wenckebach AV block that does not improve with exercise should be evaluated with an EPS for intra-His or infra-His block that may require pacemaker therapy (Class I; Level of Evidence C).**
- 4. In athletes with Wenckebach AV block and coexisting bundle-branch block or with any indication that they are at risk for progression to higher-degree AV block, EPS should be performed to identify the presence of intra-His-Purkinje or infra-His-Purkinje block that may require pacemaker therapy (Class I; Level of Evidence C).**

Type II Second-Degree (Mobitz) AV Block

Type II second-degree (Mobitz) AV block is abnormal in athletes. Athletes with type II second-degree AV block should be assessed with a history, physical examination and echocardiogram regardless of symptoms. In addition,

it is important to distinguish 2:1 Wenckebach physiology at the level of the AV node from true Mobitz type II AV block. This can usually be achieved by a stress test, but EPS may be required in rare cases. Generally, Mobitz type II second-degree AV block is considered an indication for a permanent pacemaker (2,3). The recommendations for evaluation and treatment of Mobitz type II second-degree AV block are the same as those for acquired complete heart block below.

Recommendations

- 1. Athletes with Mobitz type II second-degree AV block with a wide QRS, including isolated right bundle-branch block (RBBB) should receive a permanent pacemaker (Class I; Level of Evidence C). Restrictions for athletic participation for those with pacemakers are in the section on “Athletes With Permanent Pacemakers.”**
- 2. Permanent pacemaker implantation is reasonable for athletes with asymptomatic Mobitz type II second-degree AV block with a narrow QRS (Class IIa; Level of Evidence C).**

Complete RBBB

Athletes with a complete RBBB should have a cardiac evaluation with a history, physical examination, ECG, echocardiogram, and stress test. Ambulatory monitoring and EPS can be used in a very selective fashion in patients with documentation of symptoms possibly attributable to progression to type II second-degree AV block or complete heart block (4). Progression is more likely if left anterior fascicular block accompanies the RBBB.

Recommendation

- 1. Athletes with RBBB, who do not develop periods of type II second-degree AV block or complete heart block spontaneously or during exercise and who have no symptoms or heart disease identified by appropriate testing that otherwise precludes participation, can participate in all competitive athletics (Class I; Level of Evidence C).**

Complete Left Bundle-Branch Block

Athletes with a complete left bundle-branch block (LBBB) should have a cardiac evaluation with a history, physical examination, ECG, echocardiogram, and stress test. Ambulatory monitoring and EPS can be useful in patients with documentation of, or symptoms possibly attributable to, progression to type II second-degree AV block or complete heart block. Acquired LBBB may be associated with syncope from paroxysmal AV block. In patients with syncope or presyncope, an invasive EPS should be strongly considered to exclude intra-Hisian or infra-Hisian block.

In contrast, rate-dependent LBBB in the absence of symptoms or structural heart disease may be benign, but long-term data are lacking. However, because rate-dependent LBBB, particularly if at slow rates, often occurs in the presence of structural heart disease, a more complete evaluation is necessary to exclude the latter (5).

Recommendations

1. Athletes with permanent or rate-dependent LBBB who do not develop spontaneous type II second-degree AV block (Mobitz) or complete heart block and who have no symptoms or heart disease identified by appropriate testing that otherwise precludes participation, can participate in all competitive athletics (*Class I; Level of Evidence C*).
2. In athletes with concerning symptoms, an EPS is recommended. An athlete with a normal HV interval and a normal AV conduction response to pacing can participate in all competitive sports unless otherwise restricted by their structural heart disease (*Class I; Level of Evidence C*).
3. Athletes with abnormal AV conduction characterized by an HV interval >90 ms or a His-Purkinje block should have pacemaker implantation (*Class I; Level of Evidence C*).

Congenital High-Grade or Complete Heart Block

Athletes with congenital complete heart block, and the rare cases of congenital advanced type II second-degree heart block, should be evaluated with a history, physical examination, ECG, echocardiogram, 24-hour ambulatory monitor, and exercise stress test. The exercise stress test protocol should be to maximum level of performance to assess ability to exercise to a level comparable to the relevant athletic activity. In recent years, there has been a trend to implant pacemakers in all patients with congenital complete heart block because of concern of evolution of left ventricular dysfunction and heart failure over time (6,7).

Recommendations

1. Asymptomatic athletes without heart disease who have a junctional escape rhythm that has a QRS duration <120 ms, resting ventricular rates >40 bpm that increase appropriately with exertion, and exercise capacity that approximates that of the relevant sport can participate in athletic activity without restriction (*Class I; Level of Evidence C*).
2. Athletes with symptomatic heart block, resting ventricular rates <40 bpm, or ventricular escape rhythm with a QRS width >120 ms should have a pacemaker implanted before they participate in competitive sports. Before athletes are allowed to resume sports,

an exercise test should be conducted to ensure patient safety and that the exercise capacity of the athlete is similar to that required for the relevant sport (*Class I; Level of Evidence C*).

3. Athletes with structural heart disease and congenital complete heart block should be restricted from, or allowed to participate in, competitive athletics based on the recommendations for the type of structural heart disease with or without a permanent pacemaker (*Class I; Level of Evidence C*).

Acquired Complete Heart Block

Athletes with acquired complete heart block should be evaluated with a history, physical examination, ECG, echocardiogram, and additional diagnostic testing as is clinically appropriate. Acquired complete heart block, unless caused by completely reversible factors, is an indication for placement of a permanent pacemaker (2,3).

Recommendations

1. Athletes with acquired complete heart block should have a permanent pacemaker placed regardless of symptoms, type of structural heart disease, and exercise capacity unless the heart block is attributable to completely reversible causes and resolves completely (*Class I; Level of Evidence C*).
2. Athletes with structural heart disease and acquired complete heart block should be restricted from, or allowed to participate in, athletic activities based on the recommendations for the type of structural heart disease (*Class I; Level of Evidence C*).
3. Before athletes with a permanent pacemaker are allowed to engage in athletic activities, an exercise test should be conducted to ensure that the exercise capacity of the athlete is similar to that required by the relevant sport (*Class I; Level of Evidence C*).

ATHLETES WITH PERMANENT PACEMAKERS

Many of the patterns of bradycardia and AV conduction variants observed in athletes do not require consideration of pacemaker therapy, but a few of the conditions described have clear indications. The presence of a pacemaker is not an automatic impediment to clearance for athletic participation. The presence or absence of underlying structural heart disease, level of pacemaker dependence, risk of damage to device, and symptoms are relevant modifiers.

Recommendations

1. Generally, athletes with permanent pacemakers should be cleared for athletic participation if there are

no limiting structural heart conditions or symptoms (Class I; Level of Evidence C).

- 2. Athletes who are completely pacemaker dependent should not engage in sports in which there is a risk of collision that could result in damage to the pacemaker system (Class I; Level of Evidence C).**
- 3. Athletes treated with a pacemaker who are not pacemaker dependent may participate in sports with a risk of collision or trauma if they understand and accept the risk of damage to the pacemaker system and they have no structural heart disease that precludes participation (Class I; Level of Evidence C).**
- 4. For athletes with permanent pacemakers, protective equipment should be considered for participation in contact sports that have the potential to damage the implanted device (Class I; Level of Evidence C).**

SUPRAVENTRICULAR TACHYCARDIA

SVTs are not more common in athletes than in the general population of a similar age distribution, with the possible exception of atrial fibrillation (AF) (8,9). Treatment of these SVTs with catheter ablation is likely to achieve a permanent cure and in general is preferable to lifelong therapy with pharmacological agents. SVT-associated symptoms include palpitations, weakness, lightheadedness, and occasionally syncope, all of which may impair athletic performance, although the vast majority of SVTs are not life threatening. Symptoms do not distinguish between the different SVTs, and thus, a symptom-rhythm correlation is required. Rarely, a person with a sustained form of SVT, such as atrial flutter (AFL) or AF, or more commonly in young people, atrial or junctional tachycardias or the permanent form of junctional tachycardia, can present with a tachycardia-induced cardiomyopathy. The differential diagnosis of SVTs in the athlete includes sinus tachycardia, although this tachycardia should be relatively easily diagnosed by use of resting ECGs (10).

Atrial Fibrillation

There are some data suggesting that athletes are at increased risk of AF, and in particular vagally mediated AF (8,9,11). Athletes may be particularly prone to AF because of the high vagal tone associated with extreme fitness, as well as cardiac remodeling, which includes changes in chamber size and pressure. Other causes, including fibrosis, inflammation, and sympathetic discharge, can also play a role. All athletes with AF should undergo a workup that includes thyroid function tests, ECGs, echocardiograms, and queries for drug use, including performance-enhancing agents and illicit drugs. Athletes with AF should be evaluated for hypertension and coronary artery disease. Further testing is warranted in some cases, including cardiac magnetic resonance imaging and stress

testing. Patients with underlying cardiac disease such as dilated cardiomyopathies, hypertrophic cardiomyopathy, Brugada syndrome, and catecholaminergic ventricular tachycardia (VT) have an increased risk of AF. AF in a child or adolescent athlete is uncommon and should suggest a familial inheritance or the presence of an accessory pathway.

The management options for AF in athletes include rate control or rhythm control. Rate control, although an option, may not be ideal for competitive athletes because of the focus on performance and difficulty ensuring adequate rate control during an athletic performance. A rhythm control strategy is thus the preferred method of treatment in athletes. Rhythm control can be achieved with antiarrhythmic agents or ablation procedures. Increasingly, ablation has shown a sustained benefit, particularly in those with paroxysmal AF in the presence of a normal heart, which would likely be most athletes with AF (12); however, longer-term observations are necessary to determine benefits over many years. Antiarrhythmic drug therapy has efficacy and side effect concerns, including proarrhythmic risk. In some cases, withdrawal from competitive sports or attempts at deconditioning might be chosen. Conversely, some athletes may choose to avoid any therapy and still participate because they tolerate short episodes of AF during competition.

The other component of management is anticoagulation. Most athletes will have a low risk of systemic thromboemboli as manifested by a low CHADS₂ score or a CHA₂DS₂-VASC score of zero, and anticoagulation will rarely be necessary. If anticoagulation is used, athletes should be restricted from participation in high-impact contact sports because of the bleeding risk.

Recommendations

- 1. Athletes with AF should undergo a workup that includes thyroid function tests, queries for drug use, ECG, and echocardiogram (Class I; Level of Evidence B).**
- 2. Athletes with low-risk AF that is well tolerated and self-terminating may participate in all competitive sports without therapy (Class I; Level of Evidence C).**
- 3. In athletes with AF, when antithrombotic therapy, other than aspirin, is indicated, it is reasonable to consider the bleeding risk in the context of the specific sport before clearance (Class IIa; Level of Evidence C).**
- 4. Catheter ablation for AF could obviate the need for rate control or antiarrhythmic drugs and should be considered (Class IIa; Level of Evidence B).**

Atrial Flutter

AFL may also be more common in the athlete. The workup for AFL is identical to that of AF: thyroid function tests,

queries for drug use, ECGs, and an echocardiogram. Anticoagulation and rate control are also similar to that of AF. However, given the high cure rates of ablation and the low complication risk, AFL ablation should be the rhythm control strategy of choice for those with typical cavotricuspid isthmus-dependent flutter.

Recommendations

- 1. Athletes with AFL should undergo an evaluation that includes thyroid function tests, queries for drug use, ECG, and echocardiogram (Class I; Level of Evidence B).**
- 2. Catheter ablation for typical AFL has a high likelihood of success and should be considered (Class I; Level of Evidence B).**
- 3. When anticoagulation, other than with aspirin, is indicated in an athlete, it is reasonable to consider the bleeding risk in the context of the specific sport before clearance (Class IIa; Level of Evidence C).**

AV Nodal Reentry Tachycardia, AV Reciprocating Tachycardia, Atrial Tachycardia

These 3 tachycardias, AV nodal reentry tachycardia (AVNRT), AV reciprocating tachycardia (AVRT), and atrial tachycardia (AT), are considered together because of the many similarities they share (10), such as acute onset and termination, rates between 150 and 250 bpm, a regular ventricular rhythm, largely narrow QRS complex, and termination with adenosine. The latter is more likely to be effective in AVNRT and AVRT than AT. In addition, AT can exhibit a progressive rate increase at the onset and a gradual slowing before termination. The surface ECG may not reliably distinguish between these 3, and both acute and long-term treatments for these 3 SVTs are similar. AVNRT occurs because of dual AV nodal physiology; AVRT because of a bypass tract that allows conduction between the atria and ventricle other than via the AV node; and AT because of microreentrant circuits, automatic foci, and possibly triggered activity. The ECG in AT might be confused with the permanent form of junctional tachycardia or atypical AVNRT because of a long RP interval, but it is unlikely to be confused with AVRT and typical AVNRT, which have a short RP interval. If pre-excitation is present on a surface ECG, then AVRT is likely; however, a definite diagnosis often requires an invasive EPS. Occasionally, these SVTs can present as a wide-complex tachycardia if a bypass tract is present or if there is aberrant ventricular conduction of RBBB or LBBB. Treatment options include β -adrenergic blocking agents, nondihydropyridine calcium channel antagonists, multiple antiarrhythmic agents, and catheter ablation. Given the high success rates of catheter ablation and the low complication rate, catheter ablation is the treatment of choice in this young healthy population.

There is no clear consensus regarding the asymptomatic athlete with an ECG that demonstrates preexcitation. There is concern regarding the increased but unquantifiable risk of sudden cardiac death (SCD), most notably among athletes with accessory pathways having short refractory periods that allow very rapid ventricular rates during AF. A few studies and opinions have advocated risk stratification for asymptomatic people with an ECG that shows preexcitation (13). A recent consensus statement, endorsed by the Heart Rhythm Society and the Pediatric and Congenital Electrophysiology Society, recommends that people aged <21 years undergo initial stress testing to determine whether there is sudden and complete loss of preexcitation during exercise, which would denote low risk because of an accessory pathway with a long refractory period (14). If a person cannot be ascertained as being at low risk by stress testing, then an invasive EPS is advocated, with ablation if the bypass tract has a high risk for SCD because of an effective refractory period ≤ 250 ms.

Recommendations

- 1. Athletes with regular, acute-onset SVTs should undergo cardiac assessment with ECG and echocardiogram (Class I; Level of Evidence B).**
- 2. The treatment of choice for athletes with regular, acute-onset, symptomatic SVTs should be catheter ablation (Class I; Level of Evidence C).**
- 3. Athletes with short refractory period bypass tracts capable of anterograde conduction and a history of paroxysmal AF should have an ablation of the accessory pathway before clearance for competitive sports because of risk for life-threatening arrhythmias (Class I; Level of Evidence B).**
- 4. In athletes with asymptomatic preexcitation, it is reasonable to attempt risk stratification with stress testing to determine whether the preexcitation abruptly terminates at low heart rates. If low risk is unclear, it is reasonable to recommend invasive electrophysiological evaluation, with ablation of the bypass tract if it is deemed high risk for SCD because of a refractory period ≤ 250 ms (Class IIa; Level of Evidence B).**

VENTRICULAR ARRHYTHMIAS

A variety of ventricular arrhythmias can occur in competitive athletes across the age spectrum relevant to this document. Generally, the appearance of any ventricular arrhythmia requires evaluation before clearance for participation in athletic activities, but the level of workup depends on the specific pattern of the arrhythmias, whether they are symptom-provoking or not, and whether they occur in the presence of structural, molecular, or inflammatory heart diseases.

Premature Ventricular Complexes

Premature ventricular complexes (PVCs) are most commonly benign, but their appearance requires at least a minimal level of evaluation before clearance. The major distinctions to be made are whether they are isolated or occur in the presence of a transient or chronic cardiac abnormality, as well as how they respond to exercise (15). The minimal level of testing to acquire prognostic information is a 12-lead ECG and exercise stress test (16). In most instances, an echocardiogram will also be performed to rule out a structural abnormality that cannot be identified by either the ECG or stress test. Other imaging studies can be considered, based on the circumstances of the specific arrhythmias noted. These include computed tomography and magnetic resonance imaging for disorders such as cardiomyopathies, anomalous coronary artery origins, and subclinical myocarditis. In addition, a 24-hour ambulatory monitor may be helpful in determining the frequency and pattern of the arrhythmias. PVCs recorded at a frequency of >2,000 per 24 hours have a higher likelihood of association with underlying cardiac disease (15), estimated at 30% in this subgroup. It is reasonable to conclude that palpitations caused by PVCs in the absence of heart disease that occur at rest, are suppressed with exercise, and are not accompanied by periods of nonsustained VT (NSVT; at most, PVC couplets) are benign and should not limit full participation in competitive physical activities (17). For the purpose of this recommendation, multiform/multifocal single PVCs may be equivalent to uniform/unifocal PVCs in terms of risk assessment, as in the case of other clinical settings (18). PVCs that become more frequent or convert to runs of NSVT during exercise should lead to further evaluation, depending on findings on the initial noninvasive testing (19).

PVCs observed in the conditioned athlete without heart disease may decrease on deconditioning and reappear with reconditioning. This pattern does not indicate independently heightened risk in the absence of other risk markers, and with continued training, the frequency of ectopy decreases (20). There may be as yet unrecognized implications for higher risk of SCD associated with intense exercise in subjects in the general population who do not exercise regularly (21). This observation should be considered in deconditioned athletes who immediately begin a very intense conditioning program.

Disorders that should be considered are structural abnormalities such as occult coronary artery disease and coronary artery anomalies, including myocardial bridging, early evolution of hypertrophic cardiomyopathy, and arrhythmogenic right ventricular cardiomyopathy. Athletes with persisting frequent PVCs should remain under surveillance over time for early evidence of development of PVC-induced cardiomyopathy. Annual

cardiological evaluation is required in athletes with PVCs >2,000 per 24 hours (15). Contrast-enhanced cardiac magnetic resonance may detect subtle changes seen in hypertrophic cardiomyopathy and myocarditis (22). One study suggests that electroanatomic mapping in athletes with ventricular arrhythmias may identify evidence of subtle cardiomyopathies (23). The small number of subjects studied predominantly had sustained or NSVT or very frequent PVCs. Molecular disorders possibly associated with increased PVCs that should be considered are the various channelopathies, including long-QT syndrome and catecholaminergic polymorphic VT, and transient disorders such as a viral myocarditis should be considered. If there is evidence for the latter, the athlete should be retested after resolution of myocarditis.

Recommendations

1. Athletes with single PVCs and complex forms no greater than couplets at rest and during exercise testing without structural heart disease can participate in all competitive sports. The exercise testing protocol should be based on maximal performance rather than achieving 80% to 100% of the target heart rate to come as close as possible to the level of exertion achieved during their competitive sport (*Class I; Level of Evidence C*).
2. Athletes with PVCs at rest that increase in frequency during exercise or exercise testing and convert to repetitive forms should have further evaluation by appropriate imaging or monitoring strategies before clearance for participation in high-intensity sports. If uncontrollable exercise-induced arrhythmias produce symptoms of lightheadedness or near-syncope, fatigue, or dyspnea, the athlete should be limited to competitive sports below the level at which marked frequency increase or symptoms evolved during testing (*Class I; Level of Evidence C*).
3. Athletes with defined structural heart disease who are considered high risk based on the specific heart disease and who have PVCs with or without treatment should be limited to low-intensity class IA competitive sports. This statement applies whether or not PVCs in this setting are suppressed by drug therapy (*Class I; Level of Evidence C*). Some degree of risk can still be present during class IA sports, however, depending on the nature of the heart disease.
4. Ablation of PVCs may be considered in symptomatic patients with frequent PVCs resistant to medical therapy (*Class IIb; Level of Evidence C*).

Nonsustained VT

NSVT, defined as ≥ 3 consecutive PVCs up to a maximum duration of 30 seconds of repetitive activity that does not provoke cardiovascular collapse, has a higher probability

of reflecting an underlying disorder than single PVCs (19). Nonetheless, short runs of NSVT may be normal, but the potential for significant abnormalities must determine the workup and decision making. NSVT may occur as monomorphic or polymorphic forms. In general, patterns that are monomorphic and tend to be slower (e.g., <150 bpm) are more likely to be benign than those that are polymorphic and faster. In all cases, the minimum workup should include a 12-lead ECG and stress test, including echocardiography, either as part of the stress test or separately. A 24-hour ambulatory monitor should also be conducted, with the patient instructed to perform his or her usual levels of exercise with the monitor in place. The same limitations in regard to symptomatic worsening of the arrhythmias that are described for PVCs apply to NSVT as well. Athletes with NSVT at rest that is suppressed with exercise and who have no evidence of structural heart disease, molecular/genetic disorders, or transient abnormalities at the time of evaluation can be cleared for competitive athletics without limitations. If structural heart disease is identified, the athlete should be limited to class IA competitive sports.

Recommendations

- 1. Athletes with a structurally normal heart and no evidence of molecular/genetic or inflammatory disorders with suppression of the arrhythmia during exercise can participate in competitive athletics at any level. The exercise testing protocol should be based on maximum performance rather than achieving 80 to 100% of the target heart rate to come as close as possible to the level of exertion achieved during the athlete's competitive sport. Consideration of advanced therapy such as catheter ablation in an attempt to cure the runs of NSVT is optional (Class I; Level of Evidence C).**
- 2. For athletes without structural heart disease who have NSVT that is suppressed by drug therapy, especially β -blockers, documentation of both ambient and exercise-induced NSVT should be required before general clearance for participation in higher-level competitive athletics. Specifically, the athlete should not compete in sports with a classification greater than IA unless it is documented by exercise testing or electrophysiological testing that the arrhythmia is no longer inducible under the circumstances in which it was induced before therapy (Class I; Level of Evidence C). β -Blockers might exacerbate exercise-induced asthma.**
- 3. Athletes with structural disorders or active myocarditis and documented NSVT should only participate in low-intensity class IA sports. In the case of myocarditis, reevaluation is recommended after there is clinical and laboratory evidence of healing of the myocarditis, with return to athletics a minimum of 3 months after clinical resolution (Class I; Level of Evidence C).**

Sustained Monomorphic VT

Sustained monomorphic VT may be a benign arrhythmia, but it has a higher probability of reflecting an underlying structural disorder. Generally, the benign forms of sustained monomorphic VT appear at low levels of exercise and are suppressed during higher levels, although catecholamine-dependent forms of right ventricular outflow tract tachycardia may occur with increasing physical stress. The forms that are present at rest or at low levels of activity and are suppressed with greater levels of activity do not require therapy if the patient is asymptomatic, whereas those that appear with exercise or appear to be catecholamine dependent often respond to β -blocker therapy. In the absence of structural heart disease, athletes with this pattern, particularly if relatively slow (<150 bpm during peak activity) and asymptomatic, can be cleared to participate in athletics without restrictions, but the workup to reach this level of recommendation must be thorough, including stress testing and appropriate imaging, particularly to exclude occult heart disease. The prognosis for these patterns occurring at faster rates (e.g., >170 bpm) is less clear. Ablation is a reasonable therapy for idiopathic sustained monomorphic VT. If successful and there is no recurrence after a reasonable time interval (3 months), then return to play is allowed. For patients with structural, molecular, or inflammatory disorders who have sustained monomorphic VT at rest or exercise, athletic activity is prohibited. For acute forms of myocarditis, return to athletic activities is permissible if and when the disorder resolves.

Recommendations

- 1. Athletes with structurally normal hearts and monomorphic sustained VT amenable to catheter ablation who undergo ablation and remain free of spontaneous or induced VT at least 3 months after the procedure can resume full competitive activities (Class I; Level of Evidence C).**
- 2. Athletes with structurally normal hearts and monomorphic sustained VT who elect to undergo drug suppression with pharmacological therapy should not compete in any sports for at least 3 months after the last VT episode. In the absence of clinical recurrences or inducibility of the arrhythmia by exercise/exercise testing or EPS, all competitive sports may then be permitted (Class I; Level of Evidence C).**
- 3. For the athlete with structural heart disease and sustained monomorphic VT, moderate- and high-intensity competition is contraindicated regardless of apparent therapeutic response, although participation in low-intensity class IA competitive sports is permitted (Class III; Level of Evidence C).**

Sustained Polymorphic VT, Ventricular Flutter, and Ventricular Fibrillation

Athletes who manifest these arrhythmias in the presence or absence of structural heart disease or defined molecular/genetic disorders generally receive implantable cardioverter-defibrillators (ICDs). Athletes who have these arrhythmias in the setting of transient inflammatory or electrolyte disorders may be an exception in that they may not receive ICDs, and if they remain free of episodes of these arrhythmias for 3 months after resolution of the inflammatory process, they may be considered for reevaluation of clearance to participate.

Recommendations

- 1. Athletes who have survived a cardiac arrest caused by ventricular fibrillation or VT or who have had documented symptomatic rapid VT associated with a defined nonreversible cardiac abnormality (structural or molecular) or unidentified cause should have an ICD placed. See “Athletes With ICDs” for recommendations regarding competitive sports participation after ICD implantation (Class I; Level of Evidence A).**
- 2. Class IIb athletes who have survived a cardiac arrest caused by ventricular fibrillation or VT or who have had documented symptomatic rapid VT associated with a defined reversible abnormality (e.g., resolved acute myocarditis or a controllable electrolyte abnormality) may be considered for reinstitution of participation after reevaluation at 3 months (Class I; Level of Evidence C).**

SYNCOPE

Syncope is a transient loss of consciousness caused by transient global cerebral hypoperfusion characterized by rapid onset, short duration, and spontaneous complete recovery (24–26). Syncope in the athlete can result from relatively benign causes such as cerebral hypoperfusion because of physiology similar to that found with the common faint or neurally mediated syncope (27–30). Less frequently, syncope results from serious cardiovascular conditions that result in transient loss of cerebral blood flow because of an obstruction or arrhythmias associated with underlying structural heart disease (31). Primary electrical disorders can result in syncope in the absence of any structural heart disease (32).

Syncope or presyncope in an athlete mandates a thorough evaluation by a qualified clinician (33). The purpose of the evaluation is to determine the cause of syncope, with particular emphasis on detecting structural or electrical heart disease that may lead to sudden death. The evaluation should include a detailed history that includes specific details of the event and observations of witnesses when available. The distinction between syncope during exercise and postexertional syncope is

clinically important. Most syncopal episodes that occur immediately after exercise are benign. This pattern is believed to be a result of transient postural hypotension caused by lower-extremity pooling of blood once the athlete stops the activity (from exercise-induced vasodilation) and the resultant impairment of cardiac baroreflexes (34). It may be potentiated by relative or absolute bradycardia attributable to a parasympathetic surge at the cessation of exercise. By contrast, syncope during exercise has a higher probability of being caused by serious underlying cardiovascular disease; however, neurally mediated syncope also can be induced by prolonged intense exercise.

The history should include asking about a family history of syncope, cardiovascular disease, and sudden death. A careful physical examination with particular attention to the cardiovascular examination should be performed in all athletes. Subsequent diagnostic testing in all patients should include an ECG and an echocardiogram, with selective use of additional cardiovascular tests. These tests may include a tilt table test, exercise stress test, ambulatory monitoring, and an implantable loop monitor. The sensitivity and specificity of tilt table testing for the diagnosis of syncope in the competitive athlete are lower than for the general population, and some experts believe there is not a role for tilt testing in the workup (35). For those patients in whom the cause of syncope remains uncertain, especially if the syncope raises concern for arrhythmic causes, contrast-enhanced magnetic resonance imaging, cardiac computed tomography, coronary angiography, and invasive electrophysiological testing may be indicated. Provocative testing with stress testing, epinephrine, procainamide, or isoproterenol should be considered to identify otherwise concealed cases of long-QT syndrome, catecholaminergic polymorphic VT, and Brugada syndrome. Genetic testing may be clinically useful in selected cases (36).

Neurally mediated syncope is generally compatible with continued athletic participation once measures are taken to mitigate the syncope. The primary responsibility of the clinician is to definitively exclude structural heart disease or primary electrical disorders that may predispose to sudden death or recurrent syncope. In a significant minority of athletes, the cause of syncope cannot be established despite a thorough evaluation. Athletes with syncope of unknown cause should not participate in athletics in which the transient loss of consciousness can be hazardous.

Recommendations

- 1. Athletes with exercise-induced syncope should be restricted from all competitive athletics until evaluated by a qualified medical professional (Class I; Level of Evidence B).**
- 2. Athletes with syncope should be evaluated with a history, physical examination, ECG, and selective use**

of other diagnostic tests when there is suspicion of structural heart disease or primary electrical abnormalities that may predispose to recurrent syncope or sudden death (Class I; Level of Evidence C).

- 3. Athletes with syncope caused by structural heart disease or primary electrical disorders should be restricted from athletic activities according to the recommendations for their specific underlying cardiovascular condition (Class I; Level of Evidence C).**
- 4. Athletes with neurally mediated syncope can resume all athletic activities once measures are demonstrated to prevent recurrent syncope (Class I; Level of Evidence C).**
- 5. Athletes with syncope of unknown cause, based on a ruling out of structural or molecular pathogenesis, should not participate in athletics in which transient loss of consciousness can be hazardous (Class III; Level of Evidence C).**

ATHLETES WITH ICDs

As ICDs achieved recognition of efficacy for primary and secondary prevention of SCD, based on clinical trial and observational data, the specific question of participation of ICD recipients in competitive athletics arose. Although the various guideline documents have not addressed this issue directly, the 36th Bethesda Conference offered both general opinion (37) and several disease-specific recommendations that athletes with ICDs should limit competitive sports to class IA-level activities. This was based largely on reasoned notions, in the absence of observational data, concerning the effect of the physiology and biochemistry of high-intensity activities and underlying structural disease states on reliability of device therapy, the possibility of device malfunction, and the risk

of injury to the athlete or damage to the device by trauma. Appropriate or inappropriate discharges were also cited as potential concerns. The recommendation against competition sports participation by athletes with ICDs is being reevaluated on the basis of reported practice patterns and recently generated observational data (38,39).

Recommendations

- 1. ICD indications for competitive athletes should not differ from those applicable to the general population with appropriate diagnoses and clinical profiles (Class I; Level of Evidence C).**
- 2. Recommendations should be based on existing evidence for benefit and risk and should include discussions of potential impact on sport-specific participation and performance (Class I; Level of Evidence C).**
- 3. Participation in sports classified as IA for athletes with an ICD is reasonable if they are free of episodes of ventricular flutter or ventricular fibrillation requiring device therapy for 3 months (Class IIa; Level of Evidence C).**
- 4. Participation in sports with higher peak static and dynamic components than class IA may be considered if the athlete is free of episodes of ventricular flutter or ventricular fibrillation requiring device therapy for 3 months. The decision regarding athletic participation should be made with consideration of, and counseling of, the athlete regarding the higher likelihood of appropriate and inappropriate shocks and the potential for device-related trauma in high-impact sports (Class IIb; Level of Evidence C).**
- 5. The desire of the athlete to continue athletic competition should not represent the primary indication for use of an ICD (Class III; Level of Evidence C).**

DISCLOSURES

Writing Group Disclosures

Writing Group Member	Employment	Research Grant	Other Research Support	Speakers Bureau/Honoraria	Expert Witness	Ownership Interest	Consultant/Advisory Board	Other
Douglas P. Zipes	Indiana University	None	None	None	None	None	None	None
Michael J. Ackerman	Mayo Clinic	NIH (R01 grants)†	None	None	None	None	Boston Scientific*; Gilead Sciences*; Medtronic*; St. Jude Medical*	Transgenomic†
N.A. Mark Estes III	Tufts Medical Center	None	None	None	None	None	Medtronic*; St. Jude Medical†; Boston Scientific†	None
Richard J. Kovacs	Indiana University	None	None	None	None	None	None	None
Mark S. Link	Tufts Medical Center	None	None	None	None	None	None	None
Robert J. Myerburg	University of Miami	None	None	None	None	None	None	None

This table represents the relationships of writing group members that may be perceived as actual or reasonably perceived conflicts of interest as reported on the Disclosure Questionnaire, which all members of the writing group are required to complete and submit. A relationship is considered to be "significant" if (a) the person receives \$10,000 or more during any 12-month period, or 5% or more of the person's gross income; or (b) the person owns 5% or more of the voting stock or share of the entity, or owns \$10,000 or more of the fair market value of the entity. A relationship is considered to be "modest" if it is less than "significant" under the preceding definition.

*Modest.

†Significant.

Reviewer Disclosures

Reviewer	Employment	Research Grant	Other Research Support	Speakers Bureau/Honoraria	Expert Witness	Ownership Interest	Consultant/Advisory Board	Other
Cristina Basso	University of Padua Medical School (Italy)	None	None	None	None	None	None	None
Susan P. Etheridge	University of Utah	None	None	None	None	None	SADS board*	None
Timothy F. Feltes	Nationwide Children's Hospital/Ohio State University	None	None	None	None	None	None	None
Samuel O. Jones IV	US Air Force, USUHS	None	None	None	None	None	None	None
Brian Olshansky	Executive Health Resources†	None	None	None	None	None	Biocontrol*; Boston Scientific*; Boehringer Ingelheim*; Daiichi Sankyo*	
Satish Raj	University of Calgary (Canada)	None	None	None	None	None	None	None

This table represents the relationships of reviewers that may be perceived as actual or reasonably perceived conflicts of interest as reported on the Disclosure Questionnaire, which all reviewers are required to complete and submit. A relationship is considered to be "significant" if (a) the person receives \$10,000 or more during any 12-month period, or 5% or more of the person's gross income; or (b) the person owns 5% or more of the voting stock or share of the entity, or owns \$10,000 or more of the fair market value of the entity. A relationship is considered to be "modest" if it is less than "significant" under the preceding definition.

*Modest.

†Significant.

REFERENCES

- Zipes DP, Ackerman MJ, Estes NAM 3rd, Grant AO, Myerburg RJ, Van Hare G. Task Force 7: arrhythmias: 36th Bethesda Conference: eligibility recommendations for competitive athletes with cardiovascular abnormalities. *J Am Coll Cardiol*. 2005;45:1354-63. <http://dx.doi.org/10.1016/j.jacc.2005.02.014>.
- Epstein AE, DiMarco JP, Ellenbogen KA, Estes NA 3rd, Freedman RA, Gettes LS, Gillinov AM, Gregoratos G, Hammill SC, Hayes DL, Hlatky MA, Newby LK, Page RL, Schoenfeld MH, Silka MJ, Stevenson LW, Sweeney MO. ACC/AHA/HRS 2008 guidelines for device-based therapy of cardiac rhythm abnormalities: a report of the American College of Cardiology/American Heart Association Task Force on Practice Guidelines (Writing Committee to Revise the ACC/AHA/NASPE 2002 Guideline Update for Implantation of Cardiac Pacemakers and Antiarrhythmia Devices) [published correction appears in *J Am Coll Cardiol*. 2009;53:1473]. *J Am Coll Cardiol*. 2008;51:e1-62. <http://dx.doi.org/10.1016/j.jacc.2008.02.032>.
- Tracy CM, Epstein AE, Darbar D, DiMarco JP, Dunbar SB, Estes NA 3rd, Ferguson TB Jr., Hammill SC, Karasik PE, Link MS, Marine JE, Schoenfeld MH, Shanker AJ, Silka MJ, Stevenson LW, Stevenson WG, Varosy PD, Ellenbogen KA, Freedman RA, Gettes LS, Gillinov AM, Gregoratos G, Hayes DL, Page RL, Stevenson LW, Sweeney MO. 2012 ACCF/AHA/HRS focused update of the 2008 guidelines for device-based therapy of cardiac rhythm abnormalities: a report of the American College of Cardiology Foundation/American Heart Association Task Force on Practice Guidelines and the Heart Rhythm Society [corrected] [published correction appears in *J Am Coll Cardiol*. 2013;60:2604-5]. *J Am Coll Cardiol*. 2013;60:1297-313. <http://dx.doi.org/10.1016/j.jacc.2012.07.009>.
- Kim JH, Noseworthy PA, McCarty D, Yared K, Weiner R, Wang F, Wood MJ, Hutter AM, Picard MH, Baggish AL. Significance of electrocardiographic right bundle branch block in trained athletes. *Am J Cardiol*. 2011;107:1083-9. <http://dx.doi.org/10.1016/j.amjcard.2010.11.037>.
- Fisch C, Zipes DP, McHenry PL. Rate dependent aberrancy. *Circulation*. 1973;48:714-24.
- Moak JP, Barron KS, Houglen TJ, Wiles HB, Balaji S, Sreeram N, Cohen MH, Nordenberg A, Van Hare GF, Friedman RA, Perez M, Cecchin F, Schneider DS, Nehgme RA, Buyon JP. Congenital heart block: development of late-onset cardiomyopathy, a previously underappreciated sequela. *J Am Coll Cardiol*. 2001;37:238-42.
- Bordachar P, Zachary W, Ploux S, Labrousse L, Haissaguerre M, Thambhoji JB. Pathophysiology, clinical course, and management of congenital complete atrioventricular block. *Heart Rhythm*. 2013;10:760-6. <http://dx.doi.org/10.1016/j.hrthm.2012.12.030>.
- Furlanello F, Bertoldi A, Dallago M, Galassi A, Fernando F, Biffi A, Mazzone P, Pappone C, Chierchia S. Atrial fibrillation in elite athletes. *J Cardiovasc Electrophysiol*. 1998;9 suppl:S63-8.
- Grimsmo J, Grundvold I, Maehlum S, Arnesen H. High prevalence of atrial fibrillation in long-term endurance cross-country skiers: echocardiographic findings and possible predictors: a 28-30 years follow-up study. *Eur J Cardiovasc Prev Rehabil*. 2010;17:100-5. <http://dx.doi.org/10.1097/HJR.0b013e32833226be>.
- Page RL, Joglar JA, Al-Khatib SM, et al. 2015 ACC/AHA/HRS guideline for the management of adult patients with supraventricular tachycardia: a report of the American College of Cardiology/American Heart Association Task Force on Clinical Practice Guidelines and the Heart Rhythm Society. [published online ahead of print September 23, 2015]. *J Am Coll Cardiol*. <http://dx.doi.org/10.1016/j.jacc.2015.09.019>.
- Koopman P, Nuyens D, Garweg C, La Gerche A, De Buck S, Van Casteren L, Alzand B, Willems R, Heidbuchel H. Efficacy of radiofrequency catheter ablation in athletes with atrial fibrillation. *Europace*. 2011;13:1386-93. <http://dx.doi.org/10.1093/europace/eur142>.
- Calkins H, Kuck KH, Cappato R, Brugada J, Camm AJ, Chen SA, Crijns HJ, Damiano RJ Jr., Davies DW, DiMarco J, Edgerton J, Ellenbogen K, Ezekowitz MD, Haines DE, Haissaguerre M, Hindricks G, Ilescu Y, Jackman W, Jalife J, Jais P, Kalman J, Keane D, Kim YH, Kirchhof P, Klein G, Kottkamp H, Kumagai K, Lindsay BD, Mansour M, Marchlinski FE, McCarthy PM, Mont JL, Morady F, Nademanee K, Nakagawa H, Natale A, Nattel S, Packer DL, Pappone C, Prystowsky E, Raviele A, Reddy V, Ruskin JN, Shemin RJ, Tsao HM, Wilber D, Heart Rhythm Society Task Force on Catheter and Surgical Ablation of Atrial Fibrillation. 2012 HRS/EHRA/ECAS expert consensus statement on catheter and surgical ablation of atrial fibrillation: recommendations for patient selection, procedural techniques, patient management and follow-up, definitions, endpoints, and research trial design: a report of the Heart Rhythm Society (HRS) Task Force on Catheter and Surgical Ablation of Atrial Fibrillation. Developed in partnership with the European Heart Rhythm Association (EHRA), a registered branch of the European Society of Cardiology (ESC) and the European Cardiac Arrhythmia Society (ECAS); and in collaboration with the American College of Cardiology (ACC), American Heart Association (AHA), the Asia Pacific Heart Rhythm Society (APHRS), and the Society of Thoracic Surgeons (STS): endorsed by the governing bodies of the American College of Cardiology Foundation, the American Heart Association, the European Cardiac Arrhythmia Society, the European Heart Rhythm Association, the Society of Thoracic Surgeons, the Asia Pacific Heart Rhythm Society, and the Heart Rhythm Society. *Heart Rhythm*. 2012;9:632-96.e21. <http://dx.doi.org/10.1016/j.hrthm.2011.12.016>.
- Pappone C, Manguso F, Santinelli R, Vicedomini G, Sala S, Paglino G, Mazzone P, Lang CC, Gulletta S, Augello G, Santinelli O, Santinelli V. Radiofrequency ablation in children with asymptomatic Wolff-Parkinson-White syndrome. *N Engl J Med*. 2004;351:1197-205. <http://dx.doi.org/10.1056/NEJMoa040625>.
- Cohen MI, Triedman JK, Cannon BC, Davis AM, Drago F, Janousek J, Klein GJ, Law IH, Morady FJ, Paul T, Perry JC, Sanatani S, Tanel RE. PACES/HRS

expert consensus statement on the management of the asymptomatic young patient with a Wolff-Parkinson-White (WPW, ventricular preexcitation) electrocardiographic pattern: developed in partnership between the Pediatric and Congenital Electrophysiology Society (PACES) and the Heart Rhythm Society (HRS). Endorsed by the governing bodies of PACES, HRS, the American College of Cardiology Foundation (ACCF), the American Heart Association (AHA), the American Academy of Pediatrics (AAP), and the Canadian Heart Rhythm Society (CHRS). *Heart Rhythm*. 2012;9:1006-24.

15. Biffi A, Pelliccia A, Verdile L, Fernando F, Spataro A, Caselli S, Santini M, Maron BJ. Long-term clinical significance of frequent and complex ventricular tachyarrhythmias in trained athletes. *J Am Coll Cardiol*. 2002;40:446-52.

16. Steriotis AK, Nava A, Rigato I, Mazzotti E, Daliento L, Thiene G, Basso C, Corrado D, Bauce B. Noninvasive cardiac screening in young athletes with ventricular arrhythmias. *Am J Cardiol*. 2013;111:557-62. <http://dx.doi.org/10.1016/j.amjcard.2012.10.044>.

17. Lampert R. Evaluation and management of arrhythmia in the athletic patient. *Prog Cardiovasc Dis*. 2012;54:423-31. <http://dx.doi.org/10.1016/j.pcad.2012.01.002>.

18. Myerburg RJ, Castellanos A. Cardiac arrest and sudden cardiac death. In: Bonow RO, Mann DL, Zipes DP, Libby P, editors. *Braunwald's Heart Disease: A Textbook of Cardiovascular Medicine*. Oxford, United Kingdom: Elsevier; 2010.

19. Heidbüchel H, Corrado D, Biffi A, Hoffmann E, Panhuyzen-Goedkoop N, Hoogsteen J, Delise P, Hoff PI, Pelliccia A, Study Group on Sports Cardiology of the European Association for Cardiovascular Prevention and Rehabilitation. Recommendations for participation in leisure-time physical activity and competitive sports of patients with arrhythmias and potentially arrhythmogenic conditions, part II: ventricular arrhythmias, channelopathies and implantable defibrillators. *Eur J Cardiovasc Prev Rehabil*. 2006;13:676-86. <http://dx.doi.org/10.1097/01.hjr.0000239465.26132.29>.

20. Biffi A, Maron BJ, Culasso F, Verdile L, Fernando F, Di Giacinto B, Di Paolo FM, Spataro A, Delise P, Pelliccia A. Patterns of ventricular tachyarrhythmias associated with training, deconditioning and retraining in elite athletes without cardiovascular abnormalities. *Am J Cardiol*. 2011;107:697-703. <http://dx.doi.org/10.1016/j.amjcard.2010.10.049>.

21. 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. <http://dx.doi.org/10.1056/NEJM200011093431902>.

22. Friedrich MG, Sechtem U, Schulz-Menger J, Holmvang G, Alakaj P, Cooper LT, White JA, Abdel-Aty H, Gutberlet M, Prasad S, Aletras A, Laissy JP, Paterson I, Filipchuk NG, Kumar A, Pauschinger M, Liu P, International Consensus Group on Cardiovascular

Magnetic Resonance in Myocarditis. Cardiovascular magnetic resonance in myocarditis: a JACC White Paper. *J Am Coll Cardiol*. 2009;53:1475-87. <http://dx.doi.org/10.1016/j.jacc.2009.02.007>.

23. Dello Russo A, Pieroni M, Santangeli P, Bartoletti S, Casella M, Pelargonio G, Smaldone C, Bianco M, Di Biase L, Bellocci F, Zeppilli P, Fiorentini C, Natale A, Tondo C. Concealed cardiomyopathies in competitive athletes with ventricular arrhythmias and an apparently normal heart: role of cardiac electroanatomical mapping and biopsy. *Heart Rhythm*. 2011;8:1915-22. <http://dx.doi.org/10.1016/j.hrthm.2011.07.021>.

24. Moya A, Sutton R, Ammirati F, Blanc JJ, Brignole M, Dahm JB, Deharo JC, Gajek J, Gjesdal K, Krahn A, Massin M, Pepi M, Pezawas T, Ruiz Granell R, Sarasin F, Ungar A, van Dijk JG, Walma EP, Wieling W. Guidelines for the diagnosis and management of syncope (version 2009). *Eur Heart J*. 2009;30:2631-71.

25. Freeman R, Wieling W, Axelrod FB, Benditt DG, Benarroch E, Biaggioni I, Cheshire WP, Chelmsky T, Cortelli P, Gibbons CH, Goldstein DS, Hainsworth R, Hiltz MJ, Jacob G, Kaufmann H, Jordan J, Lipsitz LA, Levine BD, Low PA, Mathias C, Raj SR, Robertson D, Sandroni P, Schatz I, Schondorff R, Stewart JM, van Dijk JG. Consensus statement on the definition of orthostatic hypotension, neurally mediated syncope and the postural tachycardia syndrome. *Clin Auton Res*. 2011;21:69-72. <http://dx.doi.org/10.1007/s10286-011-0119-5>.

26. Strickberger SA, Benson DW, Biaggioni I, Callans DJ, Cohen MI, Ellenbogen KA, Epstein AE, Friedman P, Goldberger J, Heidenreich PA, Klein GJ, Knight BP, Morillo CA, Myerburg RJ, Sila CA. AHA/ACCF scientific statement on the evaluation of syncope: from the American Heart Association Councils on Clinical Cardiology, Cardiovascular Nursing, Cardiovascular Disease in the Young, and Stroke, and the Quality of Care and Outcomes Research Interdisciplinary Working Group; and the American College of Cardiology Foundation. *J Am Coll Cardiol*. 2006;47:473-84. <http://dx.doi.org/10.1016/j.jacc.2005.12.019>.

27. Link MS, Estes NA 3rd. How to manage athletes with syncope. *Cardiol Clin*. 2007;25:457-466, vii. <http://dx.doi.org/10.1016/j.ccl.2007.07.005>.

28. Grubb BP. Clinical practice. Neurocardiogenic syncope. *N Engl J Med*. 2005;352:1004-10. <http://dx.doi.org/10.1056/NEJMcip042601>.

29. Asplund CA, O'Connor FG, Noakes TD. Exercise-associated collapse: an evidence-based review and primer for clinicians. *Br J Sports Med*. 2011;45:1157-62. <http://dx.doi.org/10.1136/bjsports-2011-090378>.

30. Colivicchi F, Ammirati F, Santini M. Epidemiology and prognostic implications of syncope in young competing athletes. *Eur Heart J*. 2004;25:1749-53. <http://dx.doi.org/10.1016/j.ehj.2004.07.011>.

31. Khoo C, Chakrabarti S, Arbour L, Krahn AD. Recognizing life-threatening causes of syncope. *Cardiol Clin*. 2013;31:51-66. <http://dx.doi.org/10.1016/j.ccl.2012.10.005>.

32. Krahn AD, Healey JS, Simpson CS, Chauhan VS, Birnie DH, Champagne J, Gardner M, Sanatani S, Chakrabarti S, Yee R, Skanes AC, Leong-Sit P, Ahmad K, Gollob MH, Klein GJ, Gula LJ, Sheldon RS. Sentinel symptoms in patients with unexplained cardiac arrest: from the Cardiac Arrest Survivors With Preserved Ejection Fraction Registry (CASPER). *J Cardiovasc Electrophysiol*. 2012;23:60-6. <http://dx.doi.org/10.1111/j.1540-8167.2011.02185.x>.

33. O'Connor FG, Levine BD, Childress MA, Asplund CA, Oriscello RG. Practical management: a systematic approach to the evaluation of exercise-related syncope in athletes. *Clin J Sport Med*. 2009;19:429-34. <http://dx.doi.org/10.1097/JSM.0b013e3181b732c3>.

34. Levine BD, Lane LD, Buckley JC, Friedman DB, Blomqvist CG. Left ventricular pressure-volume and Frank-Starling relations in endurance athletes: implications for orthostatic tolerance and exercise performance. *Circulation*. 1991;84:1016-23.

35. Hastings JL, Levine BD. Syncope in the athletic patient. *Prog Cardiovasc Dis*. 2012;54:438-44. <http://dx.doi.org/10.1016/j.pcad.2012.02.003>.

36. Ackerman MJ, Priori SG, Willems S, Berul C, Brugada R, Calkins H, Camm AJ, Ellorin PT, Gollob M, Hamilton R, Hersberger RE, Judge DP, Le Marec H, McKenna WJ, Schulze-Bahr E, Semsarian C, Towbin JA, Watkins H, Wilde A, Wolpert C, Zipes DP. HRS/EHRA expert consensus statement on the state of genetic testing for the channelopathies and cardiomyopathies: this document was developed as a partnership between the Heart Rhythm Society (HRS) and the European Heart Rhythm Association (EHRA). *Heart Rhythm*. 2011;8:1308-39. <http://dx.doi.org/10.1016/j.hrthm.2011.05.020>.

37. Maron BJ, Zipes DP. Introduction: eligibility recommendations for competitive athletes with cardiovascular abnormalities-general considerations. *J Am Coll Cardiol*. 2005;45:1318-21. <http://dx.doi.org/10.1016/j.jacc.2005.02.006>.

38. Lampert R, Cannom D, Olshansky B. Safety of sports participation in patients with implantable cardioverter defibrillators: a survey of Heart Rhythm Society members. *J Cardiovasc Electrophysiol*. 2006;17:11-5. <http://dx.doi.org/10.1111/j.1540-8167.2005.00331.x>.

39. Lampert R, Olshansky B, Heidbüchel H, Lawless C, Saarel E, Ackerman M, Calkins H, Estes NA, Link MS, Maron BJ, Marcus F, Scheinman M, Wilkoff BL, Zipes DP, Berul CI, Cheng A, Law I, Loomis M, Barth C, Brandt C, Dziura J, Li F, Cannom D. Safety of sports for athletes with implantable cardioverter-defibrillators: results of a prospective, multinational registry. *Circulation*. 2013;127:2021-30. <http://dx.doi.org/10.1161/CIRCULATIONAHA.112.000447>.

KEY WORDS ACC/AHA Scientific Statements, arrhythmias, athletes, cardiovascular abnormalities, conduction defects

AHA/ACC SCIENTIFIC STATEMENT

Eligibility and Disqualification Recommendations for Competitive Athletes With Cardiovascular Abnormalities: Task Force 10: The Cardiac Channelopathies

A Scientific Statement From the American Heart Association and American College of Cardiology

Michael J. Ackerman, MD, PhD, FACC, *Chair**

Douglas P. Zipes, MD, FAHA, MACC*

Richard J. Kovacs, MD, FAHA, FACC*

Barry J. Maron, MD, FACC*

The cardiac channelopathies are a collection of primary, genetically mediated heart rhythm disorders (also referred to as the primary electrical disorders) that are generally associated with a structurally normal heart and a propensity for syncope, seizures, or sudden cardiac arrest precipitated by a channelopathy-mediated episode of nonsustained or sustained polymorphic ventricular tachycardia (torsade de pointes) or ventricular fibrillation. These cardiac channelopathies include long-QT syndrome (LQTS), catecholaminergic polymorphic ventricular tachycardia (CPVT), Brugada syndrome (BrS), early repolarization syndrome, short-QT syndrome, and potentially idiopathic ventricular

fibrillation. Approximately 1 in 1,000 people are affected by a cardiac channelopathy, with LQTS being most common, involving an estimated 1 in 2,000 people (1).

Presently, these channelopathies should be viewed as potentially lethal but highly treatable conditions. However, unlike the various bradyarrhythmias and tachyarrhythmias detailed in the Task Force 9 report (2), there remains significant variability and heterogeneity among pediatric and adult heart rhythm specialists in terms of their ability to diagnose, risk stratify, and treat patients with these conditions. For example, in 1 study, 40% of the patients who received

*On behalf of the American Heart Association Electrocardiography and Arrhythmias Committee of the Council on Clinical Cardiology, Council on Cardiovascular Disease in the Young, Council on Cardiovascular and Stroke Nursing, Council on Functional Genomics and Translational Biology, and the American College of Cardiology.

[illegible]

This statement was approved by the American Heart Association Science Advisory and Coordinating Committee on June 24, 2015, and the American Heart Association Executive Committee on July 22, 2015, and by the American College of Cardiology Board of Trustees and Executive Committee on June 3, 2015.

The American College of Cardiology requests that this document be cited as follows: Ackerman MJ, Zipes DP, Kovacs RJ, Maron BJ; on behalf of the American Heart Association Electrocardiography and Arrhythmias Committee of the Council on Clinical Cardiology, Council on Cardiovascular Disease in the Young, Council on Cardiovascular and Stroke Nursing, Council on Functional Genomics and Translational Biology, and the American College of Cardiology. Eligibility and disqualification recommendations for competitive athletes with cardiovascular abnormalities: Task Force 10: the cardiac channelopathies: a scientific statement from the American Heart Association and American College of Cardiology. *J Am Coll Cardiol* 2015;xx:●●●●●●.

This article has been copublished in *Circulation*.

Copies: This document is available on the World Wide Web sites of the American Heart Association (<http://my.americanheart.org>) and the American College of Cardiology (www.acc.org). For copies of this document, please contact Elsevier Inc. Reprint Department via fax (212-633-3820) or e-mail (reprints@elsevier.com).

Permissions: Multiple copies, modification, alteration, enhancement, and/or distribution of this document are not permitted without the express permission of the American College of Cardiology. Requests may be completed online via the Elsevier site (<http://www.elsevier.com/about/policies/author-agreement/obtaining-permission>).

a second opinion evaluation at a LQTS specialty center for a previously rendered diagnosis of LQTS by a heart rhythm specialist were reclassified as otherwise normal, having insufficient evidence to merit that diagnostic consideration (3). This is explained in part by the advanced knowledge and training required to evaluate and treat these less common channelopathies. Accordingly, any return-to-play decision for an athlete suspected of having a cardiac channelopathy necessitates that the athlete be evaluated, risk stratified, treated, and counseled by a heart rhythm specialist or genetic cardiologist with sufficient experience and expertise in these syndromes (4).

For the most part, restriction from virtually all competitive sports has been the guideline-based recommendation since 2005 for athletes with a cardiac channelopathy, regardless of the underlying channelopathy (5,6). This universal recommendation was given despite the observation that exercise or competitive athletics has only been established as a potentially proarrhythmic trigger for CPVT and LQTS (particularly LQT1) (7,8).

Since 2005, there have been 4 fundamental developments that inform these current recommendations. First, genetic testing is now a widely available clinical test used routinely in the evaluation of a patient with a suspected channelopathy. The first Heart Rhythm Society/European Heart Rhythm Association-sponsored guideline as to the clinical use of genetic testing for the cardiac channelopathies was published in 2011 (9).

Second, despite increased discovery of more family members (athletes and nonathletes alike) with genotype positive/phenotype-negative (i.e., concealed disease) status secondary to the availability and use of genetic testing, there has been no report of athletes with concealed channelopathic substrates in the United States experiencing their sentinel event during sport. Thus, consistent with our expert opinion-based recommendations from a decade ago, there has been no observational evidence to support the European position to disqualify an athlete based solely on a positive genetic test (5,6).

Nevertheless, it remains prudent for an athlete with a channelopathy, whether concealed or manifest, to exercise simple precautionary measures, including 1) avoidance of QT-prolonging drugs for athletes with LQTS (<http://www.crediblemeds.org>), 2) avoidance of drugs that exacerbate the BrS in affected athletes (<http://www.brugadadrugs.org>), 3) electrolyte/hydration replenishment and avoidance of dehydration for all, 4) avoiding/treating hyperthermia from febrile illnesses or training-related heat exhaustion/heat stroke for athletes with either LQTS or BrS, 5) acquisition of a personal automatic external defibrillator as part of the

athlete's personal sports safety gear, and 6) establishing an emergency action plan with the appropriate school/team officials.

Third, observational evidence, derived from a large series of athletes with either concealed, electrocardiographically manifest, or symptomatic LQTS who chose to remain competitive despite the 2005 guideline-based recommendations for their disqualification, now exists (10,11). In this single-center study of LQTS athletes, only 1 of the 130 athletes with LQTS (LQT1 specifically) experienced 2 LQT1-triggered events that resulted in appropriate ventricular fibrillation-terminating implantable cardioverter-defibrillator (ICD) therapies while playing baseball on 1 occasion and soccer on another occasion in >650 athlete-years of observation. An important caveat is that every athlete underwent an extensive 2- to 3-day evaluation that included being diagnosed, risk stratified, treated, and counseled by a single LQTS specialist. This program's experience has been reproduced independently in a study involving sports participation in genotype-positive children at another center (12).

At this point in time, no similar data exist for athletes with CPVT. Given that CPVT is likely the channelopathy most vulnerable to exercise as a proarrhythmic trigger, the likelihood of a CPVT-triggered breakthrough event despite β -blocker use is much higher than in LQTS (7), and the potential for an arrhythmia/ICD storm is greatest in patients with CPVT (13), competitive sports (beyond class IA sports) are not recommended for the athlete with CPVT and documented exercise-induced frequent premature ventricular contractions/nonsustained ventricular tachycardia. Whether or not such an athlete could be cleared in the setting of combination drug therapy (for example, β -blockers and flecainide) or after left cardiac sympathetic denervation would require consultation with a CPVT disease specialist.

Fourth, the observational experience from the North American ICD Sports Registry currently comprising >340 athletes with an ICD suggests that these athletes with an ICD can continue to participate with negligible mortality (0 deaths with 31 months' average follow-up to date) and no discernible excess in damage to the implanted device or inappropriate shocks to the patient (13). The most common heart disease represented among these athletes with an ICD was LQTS, followed by hypertrophic cardiomyopathy and arrhythmogenic right ventricular cardiomyopathy.

Despite these 4 new developments over the past decade, there remains an overall lack of data or evidence regarding the true risk that an athlete with a channelopathy faces by remaining in competitive sports. As such, these recommendations are buttressed by only Level of Evidence C.

For the purposes of this document, a previously symptomatic athlete describes one who has experienced at least 1 channelopathy-triggered/suspected syncope, seizure, or aborted/resuscitated cardiac arrest. On the other hand, an athlete with a concealed channelopathy describes an asymptomatic athlete with a positive genetic test who lacks electrocardiographic evidence on a 12-lead ECG at rest (i.e., corrected QT interval <460 ms for LQTS, no spontaneous type 1 Brugada electrocardiographic pattern in the right precordial leads for BrS, no horizontal or downsloping early repolarization pattern in the inferolateral leads for early repolarization syndrome, or corrected QT interval >380 ms for short-QT syndrome) or during exercise stress testing for CPVT (i.e., no exercise-induced premature ventricular contractions in bigeminy, couplets, or worse). An athlete with a concealed channelopathy is also referred to as genotype positive/phenotype negative.

In addition, for the purposes of this document, disease-specific treatments may include either drug therapy, denervation therapy (i.e., left cardiac sympathetic denervation for LQTS and CPVT), device therapy (generally an ICD rather than a pacemaker if device therapy is indicated), or a combination thereof. The athlete's treatment program should be based primarily on the severity of the disease phenotype and should not be unduly influenced by the patient's athlete status. In other words, an ICD should not be implanted just because the patient happens to be an athlete in order for the patient to remain an athlete. This individualized treatment program should be sought from a center or program dedicated to patients with cardiac channelopathies and implemented by a heart rhythm specialist or genetic cardiologist with sufficient experience and expertise with these disorders (4). Finally, it may be prudent to temporarily restrict an athlete who experiences a cardiac event and is suspected of having a channelopathy or the athlete with a known channelopathy who experiences a breakthrough cardiac event for 3 months to ensure adequate time for evaluation, counseling, and initiation or modification of the athlete's treatment program.

Recommendations

1. For athletes with a suspected/diagnosed cardiac channelopathy, a comprehensive evaluation by a heart rhythm specialist or genetic cardiologist with sufficient experience and expertise with these disorders is recommended (*Class I; Level of Evidence C*).
2. It is recommended that symptomatic athletes with any suspected or diagnosed cardiac channelopathy be restricted from all competitive sports until a comprehensive evaluation has been completed, the athlete and his or her family are well informed, a treatment program has been implemented, and the athlete has been asymptomatic on therapy for 3 months (*Class I; Level of Evidence C*).
3. It is reasonable for an asymptomatic athlete with genotype-positive/phenotype-negative (i.e., concealed channelopathy) LQTS, CPVT, BrS, early repolarization syndrome, idiopathic ventricular fibrillation, or short-QT syndrome to participate in all competitive sports with appropriate precautionary measures, including 1) avoidance of QT-prolonging drugs for athletes with LQTS (<http://www.crediblemeds.org>), 2) avoidance of drugs that exacerbate the BrS in affected athletes (<http://www.brugadadrugs.org>), 3) electrolyte/hydration replenishment and avoidance of dehydration for all, 4) avoidance or treatment of hyperthermia from febrile illnesses or training-related heat exhaustion or heat stroke for athletes with either LQTS or BrS, 5) acquisition of a personal automatic external defibrillator as part of the athlete's personal sports safety gear, and 6) establishment of an emergency action plan with the appropriate school or team officials (*Class IIa; Level of Evidence C*).
4. Competitive sports participation may be considered for an athlete with either previously symptomatic or electrocardiographically evident BrS, early repolarization syndrome, or short-QT syndrome assuming appropriate precautionary measures and disease-specific treatments are in place and that the athlete has been asymptomatic on treatment for at least 3 months (*Class IIb; Level of Evidence C*). If therapy includes an ICD, refer to the Task Force 9 report (2).
5. For an athlete with either symptomatic LQTS or electrocardiographically manifest LQTS (i.e., corrected QT interval >470 ms in males or >480 ms in females), competitive sports participation (except competitive swimming in a previously symptomatic LQT1 host) may be considered after institution of treatment and appropriate precautionary measures assuming the athlete has been asymptomatic on treatment for at least 3 months (*Class IIb; Level of Evidence C*). If treatment includes an ICD, refer to the Task Force 9 report (2) for recommendations regarding restrictions after the procedure, lead replacements, and so forth.
6. For an athlete with previously symptomatic CPVT or an asymptomatic CPVT athlete with exercise-induced premature ventricular contractions in bigeminy, couplets, or nonsustained ventricular tachycardia, participation in competitive sports is not recommended except for class IA sports (*Class III; Level of Evidence C*). Exceptions to this limitation should be made only after consultation with a CPVT specialist.

DISCLOSURES

Writing Group Disclosures

Writing Group Member	Employment	Research Grant	Other Research Support	Speakers Bureau/Honoraria	Expert Witness	Ownership Interest	Consultant/Advisory Board	Other
Michael J. Ackerman	Mayo Clinic	NIH (R01 grants) [†]	None	None	None	None	Boston Scientific*; Gilead Sciences*; Medtronic*; St. Jude Medical*	Transgenomic [†]
Richard J. Kovacs	Indiana University	None	None	None	None	None	None	None
Barry J. Maron	Minneapolis Heart Institute Foundation	None	None	None	None	None	None	None
Douglas P. Zipes	Indiana University	None	None	None	None	None	None	None

This table represents the relationships of writing group members that may be perceived as actual or reasonably perceived conflicts of interest as reported on the Disclosure Questionnaire, which all members of the writing group are required to complete and submit. A relationship is considered to be "significant" if (a) the person receives \$10,000 or more during any 12-month period, or 5% or more of the person's gross income; or (b) the person owns 5% or more of the voting stock or share of the entity, or owns \$10,000 or more of the fair market value of the entity. A relationship is considered to be "modest" if it is less than "significant" under the preceding definition.

*Modest.

[†]Significant.

Reviewer Disclosures

Reviewer	Employment	Research Grant	Other Research Support	Speakers Bureau/Honoraria	Expert Witness	Ownership Interest	Consultant/Advisory Board	Other
Linnea M. Baudhuin	Mayo Clinic	None	None	None	None	None	None	None
Robert M. Campbell	Children's Healthcare of Atlanta	None	None	None	None	None	None	None
Michael S. Emery	Greenville Health System	None	None	None	None	None	None	None
Bulent Gorenek	Eskisehir Osmangazi University, Eskisehir (Turkey)	None	None	None	None	None	None	None

This table represents the relationships of reviewers that may be perceived as actual or reasonably perceived conflicts of interest as reported on the Disclosure Questionnaire, which all reviewers are required to complete and submit. A relationship is considered to be "significant" if (a) the person receives \$10,000 or more during any 12-month period, or 5% or more of the person's gross income; or (b) the person owns 5% or more of the voting stock or share of the entity, or owns \$10,000 or more of the fair market value of the entity. A relationship is considered to be "modest" if it is less than "significant" under the preceding definition.

REFERENCES

- Schwartz PJ, Stramba-Badiale M, Crotti L, Pedrazzini M, Besana A, Bosi G, Gabbarini F, Goulene K, Insolia R, Mannarino S, Mosca F, Nespola L, Rimini A, Rosati E, Salice P, Spazzolini C. Prevalence of the congenital long-QT syndrome. *Circulation*. 2009;120:1761-7. <http://dx.doi.org/10.1161/CIRCULATIONAHA.109.863209>.
- Zipes DP, Link MS, Ackerman MJ, Kovacs RJ, Myerburg RJ, Estes NAM 3rd, on behalf of the American Heart Association Electrocardiography and Arrhythmias Committee of the Council on Clinical Cardiology, Council on Cardiovascular Disease in the Young, Council on Cardiovascular and Stroke Nursing, Council on Functional Genomics and Translational Biology, and the American College of Cardiology. Eligibility and disqualification recommendations for competitive athletes with cardiovascular abnormalities: Task Force 9: arrhythmias and conduction defects: a scientific statement from the American Heart Association and American College of Cardiology. *J Am Coll Cardiol*. 2015 In Press. <http://dx.doi.org/10.1016/j.jacc.2015.09.041>.
- Taggart NW, Haglund CM, Tester DJ, Ackerman MJ. Diagnostic miscues in congenital long-QT syndrome. *Circulation*. 2007;115:2613-20. <http://dx.doi.org/10.1161/CIRCULATIONAHA.106.661082>.
- Priori SG, Wilde AA, Horie M, Cho Y, Behr ER, Berul C, Blom N, Brugada J, Chiang CE, Huikuri H, Kannankeril P, Krahm A, Leenhardt A, Moss A, Schwartz PJ, Shimizu W, Tomaselli G, Tracy C. Executive summary: HRS/EHRA/APHRS expert consensus statement on the diagnosis and management of patients with inherited primary arrhythmia syndromes. *Heart Rhythm*. 2013;10:e85-108.
- Zipes DP, Ackerman MJ, Estes NAM 3rd, Grant AO, Myerburg RJ, Van Hare G. Task Force 7: arrhythmias. *J Am Coll Cardiol*. 2005;45:1354-63. <http://dx.doi.org/10.1016/j.jacc.2005.02.014>.
- Pelliccia A, Fagard R, Bjørnstad HH, Anastassakis A, Arbustini E, Assanelli D, Biffi A, Borjesson M, Carré F, Corrado D, Delise P, Dorwarth U, Hirth A, Heidbuchel H, Hoffmann E, Mellwig KP, Panhuyzen-Goedkoop N, Pisani A, Solberg EE, van-Buuren F, Vanhees L, Blomstrom-Lundqvist C, Deligiannis A, Dugmore D, Glikson M, Hoff PI, Hoffmann A, Hoffmann E, Horstkotte D, Nordrehaug JE, Oudhof J, McKenna WJ, Penco M, Priori S, Reybrouck T, Senden J, Spataro A, Thiene G, Study Group of Sports Cardiology of the Working Group of Cardiac Rehabilitation and Exercise Physiology, Working Group of Myocardial and Pericardial Diseases of the European Society of Cardiology. Recommendations for competitive sports participation in athletes with cardiovascular disease: a consensus document from the Study Group of Sports Cardiology of the Working Group of Cardiac Rehabilitation and Exercise Physiology and the Working Group of Myocardial and Pericardial Diseases of the European Society of Cardiology. *Eur Heart J*. 2005;26:1422-45. <http://dx.doi.org/10.1093/eurheartj/ehi325>.
- Priori SG, Napolitano C, Memmi M, Colombi B, Drago F, Gasparini M, DeSimone L, Coltorti F, Bloise R, Keegan R, Cruz Filho FE, Vignati G, Benatar A, DeLogu A. Clinical and molecular characterization of patients with catecholaminergic polymorphic ventricular tachycardia. *Circulation*. 2002;106:69-74.
- Schwartz PJ, Priori SG, Spazzolini C, Moss AJ, Vincent GM, Napolitano C, Denjoy I, Guicheney P, Breithardt G, Keating MT, Towbin JA, Beggs AH, Brink P, Wilde AA, Toivonen L, Zareba W, Robinson JL,

Timothy KW, Corfield V, Wattanasirichaigoon D, Corbett C, Haverkamp W, Schulze-Bahr E, Lehmann MH, Schwartz K, Coumel P, Bloise R. Genotype-phenotype correlation in the long-QT syndrome: gene-specific triggers for life-threatening arrhythmias. *Circulation*. 2001;103:89-95.

9. Ackerman MJ, Priori SG, Willems S, Berul C, Brugada R, Calkins H, Camm AJ, Ellinor PT, Gollob M, Hamilton R, Hershberger RE, Judge DP, Le Marec H, McKenna WJ, Schulze-Bahr E, Semsarian C, Towbin JA, Watkins H, Wilde A, Wolpert C, Zipes DP. HRS/EHRA expert consensus statement on the state of genetic testing for the channelopathies and cardiomyopathies this document was developed as a partnership between the Heart Rhythm Society (HRS) and the European Heart Rhythm

Association (EHRA). *Heart Rhythm*. 2011;8:1308-39. <http://dx.doi.org/10.1016/j.hrthm.2011.05.020>.

10. Johnson JN, Ackerman MJ. Competitive sports participation in athletes with congenital long QT syndrome. *JAMA*. 2012;308:764-5. <http://dx.doi.org/10.1001/jama.2012.9334>.

11. Johnson JN, Ackerman MJ. Return to play? Athletes with congenital long QT syndrome. *Br J Sports Med*. 2013;47:28-33. <http://dx.doi.org/10.1136/bjsports-2012-091751>.

12. Aziz PF, Sweeten T, Vogel RL, et al. Sports participation in genotype positive children with long QT syndrome. *JACEP*. 2015;1:62-70. <http://dx.doi.org/10.1016/j.jacep.2015.03.006>.

13. Lampert R, Olshansky B, Heidebuchel H, Lawless C, Saarel E, Ackerman MJ, Calkins H, Estes NAM, Link MS, Maron BJ, Marcus F, Scheinman M, Wilkoff BL, Zipes DP, Berul CI, Cheng A, Law I, Loomis M, Barth C, Brandt C, Dziura J, Li F, Cannom D. Safety of sports for athletes with implantable cardioverter-defibrillators: results of a prospective, multinational registry. *Circulation*. 2013;127:2021-30. <http://dx.doi.org/10.1161/CIRCULATIONAHA.112.000447>.

KEY WORDS ACC/AHA Scientific Statements, athletes, Brugada syndrome, cardiovascular abnormalities, channelopathies, heart rhythm disorders, long QT syndrome

A Scientific Statement From the American Heart Association and American College of Cardiology

Richard J. Kovacs, MD, FAHA, FACC*
Aaron L. Baggish, MD, FACC*
Robert J. Myerburg, MD, FACC*

Athletic governing organizations maintain updated lists of prohibited substances (2). The prohibition of these agents is based on preventing an unfair athletic advantage and eliminating the health risks of doping. Generally, these drugs fall into categories that include anabolic agents, hormones and related substances, β -adrenergic agonists, stimulants, and

Permissions: Multiple copies, modification, alteration, enhancement, and/or distribution of this document are not permitted without the express permission of the American College of Cardiology. Requests may be completed online via the Elsevier site (<http://www.elsevier.com/about/policies/author-agreement/obtaining-permission>).

diuretic agents (1,2). Multiple masking agents are also prohibited because they are used to hide or prevent detection of a banned substance (1,2). Drugs used for enhancement of oxygen transfer, such as erythropoietin, or techniques of autotransfusion are also prohibited (1,2). Many drugs and substances considered “recreational” rather than performance enhancing, including narcotics, cannabinoids, and alcohol, are also prohibited (1,2).

Of the many adverse effects of performance-enhancing substances, those that affect the cardiovascular system are among the most serious and will be the focus of this document (3). This section also summarizes the best available, albeit limited, data on the adverse cardiovascular effects of prohibited substances in athletes. In addition, strategies for effective implementation of anti-doping programs will be discussed, and specific recommendations for healthcare professionals will be made. To ensure harmonized, coordinated, and effective antidoping programs at the international and national level with regard to detection, deterrence, and prevention of doping, a World Anti-Doping Code has been accepted by almost all international athletic organizations (4). Ultimately, all stakeholders, including athletic governing organizations, athletes, trainers, and physicians, have a shared responsibility to discourage the use of doping in sports.

The evidence base for performance-enhancing drugs and substances is subject to limitations not usually encountered in the assessment of risk and benefit for cardiovascular drugs approved by the U.S. Food and Drug Administration (FDA). Scientifically designed studies of efficacy are lacking, and many reports or opinions are subjective and often specific to an individual sport. The application of randomized clinical trials has not been feasible and in many cases may be considered unethical because of the listing of the drug or substance on lists of banned substances (5). Searches of the medical literature for randomized trials demonstrate very few clinical trials that evaluated the efficacy and safety of performance-enhancing drugs or substances. One prospective randomized trial of supraphysiological doses of testosterone combined with strength training demonstrated an increased fat-free mass and muscle size and strength in normal men with this steroid (6). The ClinicalTrials.gov Web site does not list any currently enrolling trials when searched under the terms of *sports* or *performance* (7). Because many of the substances in question are regulated by the FDA as food supplements, claims of efficacy are not substantiated by randomized clinical trials.

The evidence base for safety is somewhat more extensive but is also limited by its observational nature and the absence of randomized trials with placebo controls in most cases. Excellent summaries of the

detrimental cardiovascular effects of performance-enhancing substances have been published (5,8). FDA efforts are largely directed at individual product recalls and warning letters for unwarranted claims rather than published trial data. However, in its ban of ephedra-containing dietary supplements in the United States in 2004, the FDA based its decision on the principle of “unreasonable risk,” a risk-benefit analytical method based on even a small potential for harm in the absence of any scientifically reliable support for benefit (9). The FDA avoided the principle of “significant risk,” which would have required a higher level of scientific reliability of specific risk than was available (9). Gaps in the evidence base may continue to expand. The number of performance-enhancing substances available to athletes continues to increase, and the substances are readily available via the Internet. Large numbers of youth are being prescribed stimulant drugs to treat attention-deficit hyperactivity disorder, with a prevalence estimated to be as high as 10% of the relevant age group (10). Participation of these patients in competitive sports will require assessment of the risk and benefit. Finally, athletes will continue to explore new substances to enhance performance, without the benefit of adequate trials of efficacy or measures of safety published in the medical literature.

The term *antidoping program* refers to any organized system designed to prevent the use of banned substances in sport. Such programs have been designed and implemented with the dual objectives of ensuring fair sport competition and protecting the health of athletes. There are numerous key stakeholders in an effective antidoping program, including athletic governing bodies, athletic league directors and administrators, healthcare professionals, athletic trainers, coaches, and athletes themselves. Collectively, this group should work to promote awareness about the consequences of the use of performance-enhancing drugs and substances (education), design and implement transparent and evidence-based drug testing protocols (detection), impart and uphold fair sanctions for athletes who abuse performance-enhancing drugs and substances (enforcement), and provide resources for athletes who develop medical or psychiatric complications (treatment).

Athletic governing organizations play a crucial role in the effort to curb abuse of performance-enhancing drugs and substances among athletes. Historically, these organizations were created to generate and maintain lists of prohibited substances and to develop policies for the detection and punishment of users (1,2). These fundamental objectives remain their primary focus. The antidoping organization community now includes members at the international, regional, national, and local levels. Over the past decade, their role has expanded to include development of widespread

educational campaigns, support of scientific research focused on abuse, certification of clinical laboratories for testing, arbitration of complex cases with disputed athlete culpability, oversight of therapeutic use exemptions, and the creation of novel abuse detection strategies, including biological passports. Athletic governing bodies should continue to revise and update lists of banned substances as new agents become available. These lists should be published in easily accessible places, should be constructed in language that can be interpreted by stakeholders from all backgrounds, and should include known medical and psychological complications of use.

Athletes of all ages and across all competition levels should be educated with guidance from physicians and relevant athletic organizations regarding the risks of illicit drugs. This includes life-threatening consequences such as sudden death with cocaine use (11). The use of performance-enhancing drugs and substances such as anabolic-androgenic steroids, growth hormone, and red cell boosting agents, as well as medications such as diuretic agents, β_2 -adrenergic agonists, and glucocorticoids, may jeopardize athletic eligibility. Use of these drugs and substances, including many commercially available nutritional supplements, can be harmful and result in athletic disqualification. Athletes should disclose all prescription medication and supplement use to healthcare providers and governing organizations such that therapeutic use exemptions can be arranged when and if necessary.

A therapeutic use exemption is an official authorization from a governing agency that indicates that an athlete may take a prescription medication that is otherwise considered a banned substance without jeopardizing athletic eligibility. The international standard for the therapeutic exemption process was created in 2004 by the World Anti-Doping Agency and is updated on a regular basis (12). At the present time, national governing agencies are responsible for all aspects of the therapeutic drug exemption application and granting process. This is contingent on 3 key criteria: 1) The athlete would experience significant health problems without taking the prohibited substance or method; 2) the therapeutic use of the substance would not produce significant enhancement of performance; and 3) there is no reasonable therapeutic alternative to the use of the otherwise prohibited substance or method. The US Anti-Doping Agency provides an algorithm for determining an individual athlete's need for a therapeutic drug exemption based on the competition level and the medication in question (12). All U.S. athletes are required to submit applications through the U.S. Anti-Doping Agency. Medications in routine clinical practice that most frequently prompt the need for therapeutic drug exemption include

β_2 -adrenergic agonists, glucocorticoids, stimulants (including methylphenidate), and β_2 -adrenergic blockers. Appropriate therapeutic drug exemption use requires a collaborative approach between the athlete, clinician, and appropriate governing body. It is the athlete's responsibility to file an application for a therapeutic drug exemption if he or she is taking a banned medication. Clinicians play a crucial role in this process, because they must justify the necessity of the medication in question and the absence of comparable alternatives.

Athletes considering the use of banned or unregulated substances should be aware that the efficacy and safety of most agents have not been assessed in rigorous scientific fashion. Athletes should not ingest any substances in an attempt to improve performance or expedite recovery from injury or training unless prescribed by a healthcare professional who abides by governing organization recommendations.

Healthcare providers should recognize that performance-enhancing drug and substance abuse is a potential issue with each athlete encountered. This applies to asymptomatic athletes evaluated during health screening visits and those presenting with symptoms that suggest occult performance-enhancing drug and substance use. An essential element of the comprehensive clinical encounter with an athlete includes careful and direct questioning about performance-enhancing drug and substance use. Providers are encouraged to ask about access to and use of common agents by name and to counsel patients about the known and uncertain medical consequences of abuse. It is the responsibility of all clinicians who care for athletes to know which prescribed medications have been included on banned substance lists and to support an athlete's therapeutic exemption application when appropriate. Clinicians who discover performance-enhancing drug and substance abuse should counsel patients about the necessity of abstinence, treat all attendant medical complications, and refer the patient to specialists, including addiction counselors, sport psychologists, and medical subspecialists as deemed appropriate on a case-by-case basis.

Recommendations

- 1. Athletes should have their nutritional needs met through a healthy, balanced diet without dietary supplements (Class I; Level of Evidence C).**
- 2. As a matter of general policy, the use of performance-enhancing drugs and supplements should be prohibited by schools, universities, and other sponsoring/participating organizations as a condition for continued participation in athletic activities (Class I; Level of Evidence C).**
- 3. The principle of "unreasonable risk" (the potential for risk in the absence of defined benefit) should be the**

standard for banning or recommending avoidance of substances being evaluated for use by athletes (*Class I; Level of Evidence C*).

4. Prohibited stimulants and other medications should be subject to exceptions based on a specific medical benefit, such as a β_2 -adrenergic blocker or a bronchodilator. Medical need should be determined by a treating physician on a case-by-case basis and

authorized by the procedures defined by the US Anti-Doping Agency (*Class I; Level of Evidence B*).

5. Athletes should receive formal education and counseling by physicians and athletic department staff on the potential dangers of recreational drugs and performance-enhancing substances, including the risk of sudden death and myocardial infarction (*Class I; Level of Evidence C*).

DISCLOSURES

Writing Group Disclosures

Writing Group Member	Employment	Research Grant	Other Research Support	Speakers Bureau/Honoraria	Expert Witness	Ownership Interest	Consultant/Advisory Board	Other
N.A. Mark Estes III	Tufts University	None	None	None	None	None	Medtronic*; St. Jude Medical†; Boston Scientific†	None
Aaron Baggish	Massachusetts General Hospital, Harvard Medical School	None	None	None	None	None	None	None
Richard J. Kovacs	Indiana University	None	None	None	None	None	None	None
Robert J. Myerburg	University of Miami	None	None	None	None	None	None	None

This table represents the relationships of writing group members that may be perceived as actual or reasonably perceived conflicts of interest as reported on the Disclosure Questionnaire, which all members of the writing group are required to complete and submit. A relationship is considered to be "significant" if (a) the person receives \$10,000 or more during any 12-month period, or 5% or more of the person's gross income; or (b) the person owns 5% or more of the voting stock or share of the entity, or owns \$10,000 or more of the fair market value of the entity. A relationship is considered to be "modest" if it is less than "significant" under the preceding definition.

*Modest.

†Significant.

Reviewer Disclosures

Reviewer	Employment	Research Grant	Other Research Support	Speakers Bureau/Honoraria	Expert Witness	Ownership Interest	Consultant/Advisory Board	Other
Michael S. Emery	Greenville Health System	None	None	None	None	None	None	None
Michelle E. Grenier	University of Mississippi	None	None	None	None	None	None	None
Carolyn A. Hempel	University at Buffalo School of Pharmacy & Pharmaceutical Sciences	None	None	None	None	None	None	None
James E. Tisdale	Purdue University	American Heart Association Grant in Aid†; Indiana Clinical Translational Sciences Institute†	None	None	None	None	None	None

This table represents the relationships of reviewers that may be perceived as actual or reasonably perceived conflicts of interest as reported on the Disclosure Questionnaire, which all reviewers are required to complete and submit. A relationship is considered to be "significant" if (a) the person receives \$10,000 or more during any 12-month period, or 5% or more of the person's gross income; or (b) the person owns 5% or more of the voting stock or share of the entity, or owns \$10,000 or more of the fair market value of the entity. A relationship is considered to be "modest" if it is less than "significant" under the preceding definition.

†Significant.

REFERENCES

1. World Anti-Doping Agency (WADA) Code Web site. <https://www.wada-ama.org/en/>. Accessed August 24, 2015.
2. List of prohibited substances and methods. World Anti-Doping Agency (WADA) Web site. Available at: <https://www.wada-ama.org/en/what-we-do/prohibited-list>. Accessed December 24, 2014.
3. Estes NAM 3rd, Kloner R, Olshansky B, Virmani R. Task Force 9: drugs and performance-enhancing substances. *J Am Coll Cardiol*. 2005;45:1368-9.
4. World Anti-Doping Code. World Anti-Doping Agency (WADA) Web site. <https://www.wada-ama.org/en/what-we-do/the-code>. Accessed March 3, 2013.
5. Deligiannis A, Björnstad H, Carre F, Heidbüchel H, Kouidi E, Panhuyzen-Goedkoop NM, Pigozzi F, Schänzer W, Vanhees L, ESC Study Group of Sports Cardiology. ESC Study Group of Sports Cardiology position paper on adverse cardiovascular effects of doping in athletes. *Eur J Cardiovasc Prev Rehabil*. 2006;13:687-94. <http://dx.doi.org/10.1097/01.hjr.0000224482.95597.7a>.
6. Bhasin S, Storer TW, Berman N, Callegari C, Clevenger B, Phillips J, Bunnell TJ, Tricker R, Shirazi A, Casaburi R. The effects of supraphysiologic doses of testosterone on muscle size and strength in normal men. *N Engl J Med*. 1996;335:1-7. <http://dx.doi.org/10.1056/NEJM199607043350101>.
7. [ClinicalTrials.gov](http://clinicaltrials.gov) registry and results database. <http://clinicaltrials.gov/>. Accessed March 3, 2013.
8. Dhar R, Stout CW, Link MS, Homoud MK, Weinstock J, Estes NA 3rd. Cardiovascular toxicities of performance-enhancing substances in sports [published correction appears in *Mayo Clin Proc*. 2006;81:133]. *Mayo Clin Proc*. 2005;80:1307-15. <http://dx.doi.org/10.4065/80.10.1307>.
9. FDA acts to remove ephedra-containing dietary supplements from market [news release]. Silver Spring, MD: US Food and Drug Administration, November 23, 2004. <http://www.fda.gov/NewsEvents/Newsroom/PressAnnouncements/2004/ucm108379.htm>. Accessed August 26, 2014.
10. Kratochvil CJ, Vaughan BS, Barker A, Corr L, Wheeler A, Madaan V. Review of pediatric attention deficit/hyperactivity disorder for the general psychiatrist. *Psychiatr Clin North Am*. 2009;32:39-56. <http://dx.doi.org/10.1016/j.psc.2008.10.001>.
11. Isner JM, Estes NA 3rd, Thompson PD, Costanzo-Nordin MR, Subramanian R, Miller G, Katsas G, Sweeney K, Stumer WQ. Acute cardiac events temporally related to cocaine abuse. *N Engl J Med*. 1986;315:1438-43. <http://dx.doi.org/10.1056/NEJM198612043152302>.
12. International Standard for Therapeutic Use Exemptions (ISTUE). World Anti-Doping Agency (WADA) Web site. <https://www.wada-ama.org/en/resources/therapeutic-use-exemption-tue/international-standard-for-therapeutic-use-exemptions-istue>. Accessed August 24, 2015.

KEY WORDS ACC/AHA Scientific Statements, athletes, cardiovascular abnormalities, drugs, performance-enhancing substances

AHA/ACC SCIENTIFIC STATEMENT

Eligibility and Disqualification

Recommendations for Competitive Athletes With Cardiovascular Abnormalities:

Task Force 12: Emergency Action Plans, Resuscitation, Cardiopulmonary Resuscitation, and Automated External Defibrillators

A Scientific Statement From the American Heart Association and American College of Cardiology

Mark S. Link, MD, FACC, *Chair**

Robert J. Myerburg, MD, FACC*
N.A. Mark Estes III, MD, FACC*

The ability to resuscitate cardiac arrest victims is a critical component of health-related topics in the athlete population. Even with screening, there will remain people who experience sudden cardiac arrest. An effective resuscitation strategy requires multiple elements, including planning for an event, appropriate team members who can provide cardiopulmonary resuscitation (CPR), rapid availability of automated external defibrillators (AEDs)

and other appropriate equipment, and calls for emergency medical services (EMS). The chain of survival as articulated by the American Heart Association (AHA) calls for immediate recognition of cardiac arrest and activation of EMS, early CPR, rapid defibrillation, effective advanced life support, and integrated post-cardiac arrest care (1,2). Inadequacy in any one of these facets will reduce the chances of survival.

*On behalf of the American Heart Association Electrocardiography and Arrhythmias Committee of the Council on Clinical Cardiology, Council on Cardiovascular Disease in the Young, Council on Cardiovascular and Stroke Nursing, Council on Functional Genomics and Translational Biology, and the American College of Cardiology.

[illegible]

This statement was approved by the American Heart Association Science Advisory and Coordinating Committee on June 24, 2015, and the American Heart Association Executive Committee on July 22, 2015, and by the American College of Cardiology Board of Trustees and Executive Committee on June 3, 2015.

The American College of Cardiology requests that this document be cited as follows: Link MS, Myerburg RJ, Estes NAM 3rd; on behalf of the American Heart Association Electrocardiography and Arrhythmias Committee of the Council on Clinical Cardiology, Council on Cardiovascular Disease in the Young, Council on Cardiovascular and Stroke Nursing, Council on Functional Genomics and Translational Biology, and the American College of Cardiology. Eligibility and disqualification recommendations for competitive athletes with cardiovascular abnormalities: Task Force 12: emergency action plans, resuscitation, cardiopulmonary resuscitation, and automated external defibrillators: a scientific statement from the American Heart Association and American College of Cardiology. *J Am Coll Cardiol* 2015;xx:xxxx-xxxx.

This article has been copublished in *Circulation*.

Copies: This document is available on the World Wide Web sites of the American Heart Association (<http://my.americanheart.org>) and the American College of Cardiology (www.acc.org). For copies of this document, please contact Elsevier Inc. Reprint Department via fax (212-633-3820) or e-mail (reprints@elsevier.com).

Permissions: Multiple copies, modification, alteration, enhancement, and/or distribution of this document are not permitted without the express permission of the American College of Cardiology. Requests may be completed online via the Elsevier site (<http://www.elsevier.com/about/policies/author-agreement/obtaining-permission>).

BASICS OF AEDS

AEDs are portable devices capable of detecting and terminating ventricular tachycardia and fibrillation. All require human input to place the pads and turn on the device. Some are fully automated in that they will analyze the rhythm and provide a shock if the arrhythmia is deemed shockable. However, most are semiautomated in that they require continued human input, including activation to analyze the rhythm, and then if the arrhythmia is deemed shockable, further activation to shock. Ease of use has been demonstrated for both automated and semiautomated AEDs. AEDs are manufactured by many companies, with subtle differences in sensing algorithms and shock energy.

The sensitivity and specificity of AEDs are excellent and likely better than human analysis of arrhythmias (3). In arrhythmia libraries, the sensitivity of most devices approaches 100%, as does the specificity (3). Whether one manufacturer's algorithms are more accurate than others is not clear. Some devices will correct for CPR artifact, analyze the quality of the CPR, or both. Nearly all current AEDs incorporate biphasic waveforms; however, the specifics of the waveform and the energy vary among manufacturers. In addition, some AEDs use escalating energies, whereas others have fixed energy output. Whether one type of waveform or energy level is better than another is not clear; however, the ability to terminate ventricular fibrillation is generally excellent.

AEDs may be used in children; however, the AHA recommends the use of pediatric dose attenuator systems and pediatric pads, if available, for people aged 1 to 8 years (3). AEDs require routine maintenance; battery life and system integrity require at least monthly checks, and pads have a limited shelf-life span of ≈ 2 years. Thus, AEDs should be part of an emergency action plan and should not be placed in isolation.

INITIAL RESPONSE TO SUSPECTED CARDIAC ARREST IN THE SPORTS ENVIRONMENT

The AHA guidelines regarding response to out-of-hospital cardiac arrest generally apply to the circumstances in which athlete-related cardiac arrests occur (4-6). These include immediate assessment of level of consciousness and cardiovascular status of the athlete who has collapsed unexpectedly, as well as institution of chain-of-survival actions when cardiac arrest is identified. Because sport-related cardiac arrest has a higher probability of being witnessed by appropriately trained bystander staff than does cardiac arrest that

occurs in the general population, a beneficial outcome is more likely if delays in recognition and responses are avoided. Although benign forms of syncope and near syncope may occur in these settings, it is important to recognize that the onset of cardiac arrest may be heralded by a brief period of drifting in and out of consciousness because of unstable rhythms before a full arrest. It should not be assumed that such patterns are benign.

An important additional factor in many sport-related incidents is the distinction between primary cardiac arrest and cardiac arrest caused by chest wall trauma (commotio cordis). It is critical to determine quickly whether impaired consciousness is associated with loss of pulse and respiration and to institute appropriate resuscitative therapy immediately. The responder must also distinguish loss of pulse caused by an extreme vagal response from a true cardiac arrest. Vagal responses are usually transient and may be associated with marked bradycardia and reduction of blood pressure; respirations typically continue. It is also important, but often difficult, to make the sometimes subtle distinctions between involuntary seizure-like movements associated with cardiac arrest and epilepsy-related seizures. Distinguishing between true spontaneous respirations and gasping respirations is also important, the latter being a part of cardiac arrest physiology (7), which supports the recognition of true cardiac arrest.

BASIC LIFE SUPPORT AND AED DEPLOYMENT FOR THE ATHLETE IN CARDIAC ARREST

If the pulse and spontaneous respirations are absent, it should be assumed that a cardiac arrest is present, and the initial steps in resuscitation should be effected immediately. If an AED is immediately available, it should be deployed simultaneous with the act of contacting emergency rescue personnel (4). In some sports settings, primarily during competitive events with large attendance, a rescue vehicle may be stationed at the scene, but that is less likely during practices or sports events in small facilities. Nonetheless, AEDs are being deployed increasingly in public venues, including schools, universities, and various sports and exercise facilities. Both campus security personnel and EMS should be contacted immediately. It is recommended that the devices be deployed in a manner that results in a maximum access time of 5 minutes to any site on a school campus or sporting venue (8-10).

While the AED is being brought to the victim's side and deployed, compression of the chest should be started by bystanders. According to the most recent

guidelines, compression alone (“hands-only CPR”) should be started at a rate of 100 to 120 compressions per minute without interruption for rescue breaths (1). Trained professional providers should include rescue breathing. As soon as the defibrillator is attached and powered up, the rhythm is analyzed. A single shock should be delivered if the device senses a shockable rhythm (11). If the initial rhythm is nonshockable (i.e., asystole or pulseless electrical activity), CPR should be continued. If an initial shock fails to restore a spontaneous rhythm with return of spontaneous circulation, compressions should be resumed for 2 minutes before another shock is attempted. The previous concept of delivering 2 or 3 consecutive shocks before resuming CPR is no longer advised, and no more than 1 shock at a time is given, with 2 minutes of chest compressions between each shock. Once emergency rescue personnel are on the scene, advanced life support activities will be implemented as needed. These may include intubation with respiratory management and pharmacological interventions.

The likelihood of survival with good neurological status is directly related to the time between onset of cardiac arrest, implementation of CPR, and return of spontaneous circulation. In the adequately prepared athletic environment, including both trained staff and appropriate equipment, with the onset of the event witnessed, it is a reasonable goal to begin CPR within 60 to 90 seconds and deliver an initial shock to an athlete with a shockable rhythm in <3 minutes.

EMERGENCY RESPONSE PLANS

Comprehensive emergency response plans are as important as the individual aspects of CPR and AEDs (6). The initial recognition of an arrest and immediate CPR must cascade into activation of the emergency response plan, which includes early access to a defibrillator and placement of calls to the local EMS (for example, 9-1-1 in most of the United States). An emergency response plan includes preparation for cardiac arrests, including anticipation of events, placement of AEDs and training of people to use them, access to emergency services, and simulations of real-life events. Included in emergency response plans are monthly AED checks for integrity and battery life.

Similar to treatment of other out-of-hospital cardiac arrest patients, therapeutic hypothermia (also referred to as targeted temperature management) should be started as soon as possible in the victim who is comatose after successful return of spontaneous circulation. Emergency response plans should consider transfer to facilities that are capable of therapeutic hypothermia (12,13).

EMERGENCY RESPONSE: LEGAL CONSIDERATIONS

There are multiple legal and regulatory considerations that minimize legal risks of AED ownership, use, or medical oversight (14). To address liability concerns, state and federal Good Samaritan legislation currently protects responders using AEDs (15,16). Good Samaritan legislation statutes provide immunity from claims of negligence for volunteers aiding others with CPR and AED use. The Federal Cardiac Arrest Survival Act (CASA) was enacted in 2000 with provisions to encourage AED use in federal buildings and to create immunity for AED users (15). CASA provides conditional immunity from legal liability for harm resulting from use or attempted use of an AED by lay responders. All 50 states have Good Samaritan laws that vary in scope and conditions but that supplement the basic protections from liability afforded by federal regulations (17). The state AED program requirements generally include the provisions of Good Samaritan immunity, medical oversight, agency notification, policies, quality assurance measures, training, AED maintenance, and postevent reporting. The AHA has developed a policy statement with the objective of guiding policymakers and other stakeholders in writing new legislation or revising existing legislation to remove potential barriers to implementation of emergency response programs that include AEDs (18). Those considering starting an AED program should consult and adhere to state regulations to minimize potential risks associated with AED ownership, oversight, or use. Healthcare professionals should be aware of the clinical benefits of AEDs and the limited liability associated with their use. They should also consider the potential liability that could arise from failure to use AEDs as a matter of prudent public protection.

Recommendations

- Schools and other organizations hosting athletic events or providing training facilities for organized competitive athletic programs should have an emergency action plan that incorporates basic life support and AED use within a broader plan to activate EMS (6,10) (Class I; Level of Evidence B).**
- Coaches and athletic trainers should be trained to recognize cardiac arrests and to implement timely and AHA guideline-directed CPR (100 to 120 beats per minute and compression depth of 2 inches) along with AED deployment (4,6) (Class I; Level of Evidence B).**
- AEDs should be available to all cardiac arrest victims within 5 minutes, in all settings, including competition, training, and practice (9,10) (Class I; Level of Evidence B).**
- Advanced post-cardiac arrest care, including targeted temperature management, should be available at sites to which patients are taken by EMS (19,20) (Class I; Level of Evidence A).**

DISCLOSURES

Writing Group Disclosures

Writing Group Member	Employment	Research Grant	Other Research Support	Speakers Bureau/Honoraria	Expert Witness	Ownership Interest	Consultant/Advisory Board	Other
Mark S. Link	Tufts Medical Center	None	None	None	None	None	None	None
N.A. Mark Estes III	Tufts Medical Center	None	None	None	None	None	Medtronic*; St. Jude Medical†	None
Robert J. Myerburg	University of Miami	None	None	None	None	None	None	None

This table represents the relationships of writing group members that may be perceived as actual or reasonably perceived conflicts of interest as reported on the Disclosure Questionnaire, which all members of the writing group are required to complete and submit. A relationship is considered to be "significant" if (a) the person receives \$10,000 or more during any 12-month period, or 5% or more of the person's gross income; or (b) the person owns 5% or more of the voting stock or share of the entity, or owns \$10,000 or more of the fair market value of the entity. A relationship is considered to be "modest" if it is less than "significant" under the preceding definition.

*Modest.

†Significant.

Reviewer Disclosures

Reviewer	Employment	Research Grant	Other Research Support	Speakers Bureau/Honoraria	Expert Witness	Ownership Interest	Consultant/Advisory Board	Other
Robert M. Campbell	Children's Healthcare of Atlanta	None	None	None	None	None	None	None
Frederick G. Kushner	West Jefferson Heart Clinic of Louisiana	None	None	None	None	None	None	None
Silvana M. Lawrence	Baylor College of Medicine	None	None	None	None	None	None	Vice President, Science and Research, Championship Hearts Foundation*
Wanchun Tang	University of Southern California	None	None	None	None	None	None	None
Roger White	Mayo Clinic	None	None	None	None	None	None	None
Dongmei Wu	Mount Sinai Medical Center	None	None	None	None	None	None	None

This table represents the relationships of reviewers that may be perceived as actual or reasonably perceived conflicts of interest as reported on the Disclosure Questionnaire, which all reviewers are required to complete and submit. A relationship is considered to be "significant" if (a) the person receives \$10,000 or more during any 12-month period, or 5% or more of the person's gross income; or (b) the person owns 5% or more of the voting stock or share of the entity, or owns \$10,000 or more of the fair market value of the entity. A relationship is considered to be "modest" if it is less than "significant" under the preceding definition.

*Modest.

REFERENCES

- Field JM, Hazinski MF, Sayre MR, Chameides L, Schexnayder SM, Hemphill R, Samson RA, Kattwinkel J, Berg RA, Bhanji F, Cave DM, Jauch EC, Kudenchuk PJ, Neumar RW, Peberdy MA, Perlman JM, Sinz E, Travers AH, Berg MD, Billi JE, Eigel B, Hickey RW, Kleinman ME, Link MS, Morrison LJ, O'Connor RE, Shuster M, Callaway CW, Cucchiara B, Ferguson JD, Rea TD, Vanden Hoek TL. Part 1: executive summary: 2010 American Heart Association guidelines for cardiopulmonary resuscitation and emergency cardiovascular care. *Circulation*. 2010;122 suppl 3:S640-56. <http://dx.doi.org/10.1161/CIRCULATIONAHA.110.970889>.
- Travers AH, Rea TD, Bobrow BJ, Edelson DP, Berg RA, Sayre MR, Berg MD, Chameides L, O'Connor RE, Swor RA. Part 4: CPR overview: 2010 American Heart Association guidelines for cardiopulmonary resuscitation and emergency cardiovascular care. *Circulation*. 2010;122 suppl 3:S676-84. <http://dx.doi.org/10.1161/CIRCULATIONAHA.110.970913>.
- Link MS, Atkins DL, Passman RS, Halperin HR, Samson RA, White RD, Cudnik MT, Berg MD, Kudenchuk PJ, Kerber RE. Part 6: electrical therapies: automated external defibrillators, defibrillation, cardioversion, and pacing: 2010 American Heart Association guidelines for cardiopulmonary resuscitation and emergency cardiovascular care [published correction appears in *Circulation*. 2011;123:e235]. *Circulation*. 2010;122 suppl 3:S706-19. <http://dx.doi.org/10.1161/CIRCULATIONAHA.110.970954>.
- Berg RA, Hemphill R, Abella BS, Aufderheide TP, Cave DM, Hazinski MF, Lerner EB, Rea TD, Sayre MR, Swor RA. Part 5: adult basic life support: 2010 American Heart Association guidelines for cardiopulmonary resuscitation and emergency cardiovascular care [published correction appears in *Circulation*. 2011;124:e402]. *Circulation*. 2010;122 suppl 3:S685-705. <http://dx.doi.org/10.1161/CIRCULATIONAHA.110.970939>.
- Kramer EB, Botha M, Drezner J, Abdelrahman Y, Dvorak J. Practical management of sudden cardiac arrest on the football field. *Br J Sports Med*. 2012;46:1094-6. <http://dx.doi.org/10.1136/bjsports-2012-091376>.
- Drezner JA, Courson RW, Roberts WO, Mosesso VN Jr., Link MS, Maron BJ, Inter-Association Task Force. Inter-association task force recommendations on emergency preparedness and management of sudden cardiac arrest in high school and college athletic programs: a consensus statement. *Heart Rhythm*. 2007;4:549-65. <http://dx.doi.org/10.1016/j.hrthm.2007.02.019>.
- Bobrow BJ, Zuercher M, Ewy GA, Clark L, Chikani V, Donahue D, Sanders AB, Hilwig RW, Berg RA, Kern KB.

Gasping during cardiac arrest in humans is frequent and associated with improved survival. *Circulation*. 2008;118:2550-4. <http://dx.doi.org/10.1161/CIRCULATIONAHA.108.799940>.

8. Hazinski MF, Markenson D, Neish S, Gerardi M, Hootman J, Nichol G, Taras H, Hickey R, O'Connor R, Potts J, van der Jagt E, Berger S, Schexnayder S, Garson A Jr., Doherty A, Smith S. 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. <http://dx.doi.org/10.1161/01.CIR.0000109486.45545.AD>.

9. Balady GJ, Chaitman B, Foster C, Froelicher E, Gordon N, Van Camp S. Automated external defibrillators in health/fitness facilities: supplement to the AHA/ACSM recommendations for cardiovascular screening, staffing, and emergency policies at health/fitness facilities. *Circulation*. 2002;105:1147-50.

10. Cave DM, Aufderheide TP, Beeson J, Ellison A, Gregory A, Hazinski MF, Hiratzka LF, Lurie KG, Morrison LJ, Mosesso VN Jr., Nadkarni V, Potts J, Samson RA, Sayre MR, Schexnayder SM, on behalf of the American Heart Association Emergency Cardiovascular Care Committee, Council on Cardiopulmonary, Critical Care, Perioperative and Resuscitation, Council on Cardiovascular Diseases in the Young; Council on Cardiovascular Nursing, Council on Clinical Cardiology, and Advocacy Coordinating Committee. Importance and implementation of training in cardiopulmonary resuscitation and automated external defibrillation in schools: a science advisory from the American Heart Association. *Circulation*. 2011;123:691-706. <http://dx.doi.org/10.1161/CIR.0b013e31820b5328>.

11. Jacobs I, Sunde K, Deakin CD, Hazinski MF, Kerber RE, Koster RW, Morrison LJ, Nolan JP, Sayre MR,

on behalf of Defibrillation Chapter Collaborators. Part 6: defibrillation: 2010 International Consensus on Cardiopulmonary Resuscitation and Emergency Cardiovascular Care Science With Treatment Recommendations. *Circulation*. 2010;122 suppl 2:S325-37. <http://dx.doi.org/10.1161/CIRCULATIONAHA.110.971010>.

12. Neumar RW, Otto CW, Link MS, Kronick SL, Shuster M, Callaway CW, Kudenchuk PJ, Ornato JP, McNally B, Silvers SM, Passman RS, White RD, Hess EP, Tang W, Davis D, Sinz E, Morrison LJ. Part 8: adult advanced cardiovascular life support: 2010 American Heart Association guidelines for cardiopulmonary resuscitation and emergency cardiovascular care [published corrections appear in *Circulation*. 2011;123:e236 and *Circulation*. 2013;128:e480]. *Circulation*. 2010;122 suppl 3:S729-67. <http://dx.doi.org/10.1161/CIRCULATIONAHA.110.970988>.

13. Peberdy MA, Callaway CW, Neumar RW, Geocadin RG, Zimmerman JL, Donnino M, Gabrielli A, Silvers SM, Zaritsky AL, Merchant R, Vanden Hoek TL, Kronick SL. Part 9: post-cardiac arrest care: 2010 American Heart Association guidelines for cardiopulmonary resuscitation and emergency cardiovascular care [published corrections appear in *Circulation*. 2011;123:e237 and *Circulation*. 2011;124:e403]. *Circulation*. 2010;122 suppl 3:S768-86. <http://dx.doi.org/10.1161/CIRCULATIONAHA.110.971002>.

14. England H, Weinberg PS, Estes NA 3rd. The automated external defibrillator: clinical benefits and legal liability. *JAMA*. 2006;295:687-90. <http://dx.doi.org/10.1001/jama.295.6.687>.

15. Cardiac Arrest Survival Act of 2000. Pub L No. 106-505, 114 Stat 2314.

16. US Department of Health and Human Services program support center Website. Guidelines for public access defibrillation programs in federal facilities.

<https://www.foh.hhs.gov/whatwedo/AED/HHSAED.ASP>. Accessed August 23, 2015.

17. National Conference of State Legislatures Website. State laws on cardiac arrest and defibrillators. <http://www.ncsl.org/research/health/laws-on-cardiac-arrest-and-defibrillators-aeds.aspx>. Accessed August 23, 2015.

18. Aufderheide T, Hazinski MF, Nichol G, Steffens SS, Buroker A, McCune R, Stapleton E, Nadkarni V, Potts J, Ramirez RR, Eigel B, Epstein A, Sayre M, Halperin H, Cummins RO. Community lay rescuer automated external defibrillation programs: key state legislative components and implementation strategies: a summary of a decade of experience for healthcare providers, policymakers, legislators, employers, and community leaders from the American Heart Association Emergency Cardiovascular Care Committee, Council on Clinical Cardiology, and Office of State Advocacy. *Circulation*. 2006;113:1260-70. <http://dx.doi.org/10.1161/CIRCULATIONAHA.106.172289>.

19. Bernard SA, Gray TW, Buist MD, Jones BM, Silvester W, Gutteridge G, Smith K. Treatment of comatose survivors of out-of-hospital cardiac arrest with induced hypothermia. *N Engl J Med*. 2002;346:557-63. <http://dx.doi.org/10.1056/NEJMoa003289>.

20. The Hypothermia After Cardiac Arrest Study Group. Mild therapeutic hypothermia to improve the neurologic outcome after cardiac arrest [published correction appears in *N Engl J Med*. 2002;346:1756]. *N Engl J Med*. 2002;346:549-56.

KEY WORDS ACC/AHA Scientific Statements, athletes, automated external defibrillator, cardiovascular abnormalities, CPR, emergency action plan, resuscitation

AHA/ACC SCIENTIFIC STATEMENT

Eligibility and Disqualification Recommendations for Competitive Athletes With Cardiovascular Abnormalities: Task Force 13: Commotio Cordis

A Scientific Statement From the American Heart Association and American College of Cardiology

Mark S. Link, MD, FACC, *Chair**

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

Commotio cordis is defined as sudden cardiac death triggered by a relatively innocent blow to the precordium (1). Although initially thought to be extremely rare, it is now increasingly reported in the United States and worldwide (2,3). Enhanced recognition of commotio cordis, rather than an increase in event frequency, likely accounts for the greater visibility of those events. Commotio cordis is one of the most common causes of sudden cardiac death in recreational and competitive sports, instantaneously resulting in a potentially fatal arrhythmia (4). Commotio cordis is distinct from cardiac contusion, in which structural damage to the heart with resultant

arrhythmias develops within 24 hours after severe chest impact (5).

RISK FACTORS FOR COMMOTIO CORDIS

Risk factors for a commotio cordis have been defined by a Commotio Cordis Registry of clinical events and an experimental swine model. Human cases occur largely in adolescent males (95% of cases), with a mean age of 14 years (2). Impacts occur over the left chest wall and are generally sustained with a hard spherical object such as a baseball, hockey puck, lacrosse ball, or softball. Collapse is instantaneous or

*On behalf of the American Heart Association Electrocardiography and Arrhythmias Committee of the Council on Clinical Cardiology, Council on Cardiovascular Disease in the Young, Council on Cardiovascular and Stroke Nursing, Council on Functional Genomics and Translational Biology, and the American College of Cardiology.

[illegible]

This statement was approved by the American Heart Association Science Advisory and Coordinating Committee on June 24, 2015, and the American Heart Association Executive Committee on July 22, 2015, and by the American College of Cardiology Board of Trustees on June 3, 2015.

The American College of Cardiology requests that this document be cited as follows: Link MS, Estes NAM 3rd, Maron BJ; on behalf of the American Heart Association Electrocardiography and Arrhythmias Committee of the Council on Clinical Cardiology, Council on Cardiovascular Disease in the Young, Council on Cardiovascular and Stroke Nursing, Council on Functional Genomics and Translational Biology, and the American College of Cardiology. Eligibility and disqualification recommendations for competitive athletes with cardiovascular abnormalities: Task Force 13: commotio cordis: a scientific statement from the American Heart Association and American College of Cardiology. J Am Coll Cardiol 2015;xx:●●●-●●●.

This article has been copublished in *Circulation*.

Copies: This document is available on the World Wide Web sites of the American Heart Association (<http://my.americanheart.org>) and the American College of Cardiology (www.acc.org). For copies of this document, please contact Elsevier Inc. Reprint Department via fax (212-633-3820) or e-mail (reprints@elsevier.com).

Permissions: Multiple copies, modification, alteration, enhancement, and/or distribution of this document are not permitted without the express permission of the American College of Cardiology. Requests may be completed online via the Elsevier site (<http://www.elsevier.com/about/policies/author-agreement/obtaining-permission>).

within a few seconds; when a defibrillator is used rapidly, the arrhythmia is typically ventricular fibrillation (VF).

An experimental model of commotio cordis has confirmed the arrhythmia induced by a chest blow is VF (6). Impact must occur over the cardiac silhouette, and harder balls are more likely to induce VF (6-8). In addition, this model has demonstrated the critical importance of timing in that only those blows that occur during a narrow time segment of the T-wave upstroke reliably produce VF (6). These laboratory experiments have also shown the importance of size and shape of the object (9). Blows must occur directly perpendicular to the chest wall to produce VF, and impact velocities optimal to produce commotio cordis are those just slightly less than velocities which produce cardiac damage (in the swine model 40 mph optimal; 50 mph creates cardiac damage) (10).

RESUSCITATION

Initially, it was thought that successful resuscitation was more difficult to achieve in commotio cordis victims than in sudden cardiac death in other conditions (1). This perception was based on the poor rate of survival of commotio cordis victims reported to the Commotio Cordis Registry before 1995 (1). Registry data reported in 2002 showed that survival had increased to 15% (11). More recent data from the Commotio Cordis Registry demonstrate that the survival rate has increased steadily over

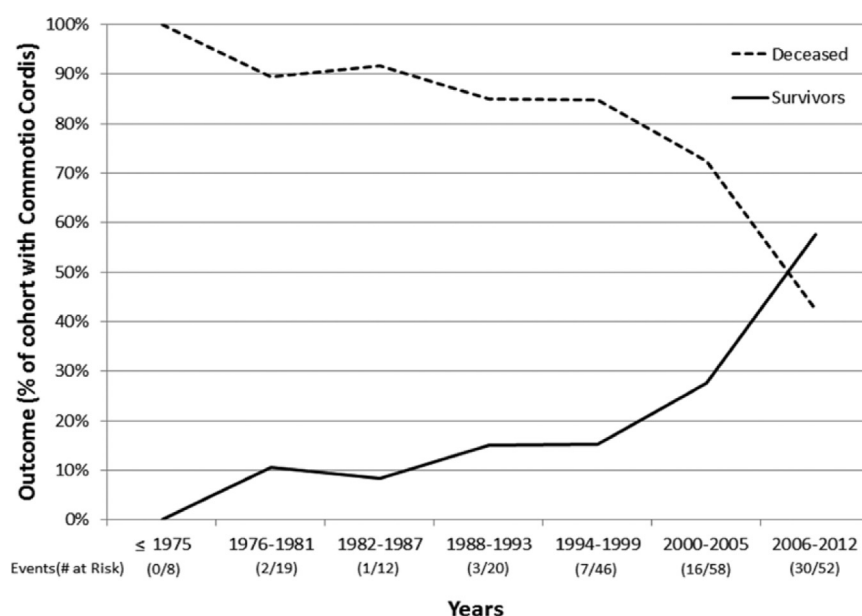
the past 15 years (Figure) (12), and survival in the most recent years is now >50%. The reasons for improved survival are multifactorial, including greater recognition of commotio cordis, which leads to a shorter time interval after collapse to cardiopulmonary resuscitation and defibrillation; more dissemination of automated external defibrillators in the community; and a greater number of people who have been trained and are willing to perform cardiopulmonary resuscitation and defibrillation.

Barriers remain to a successful outcome for victims of commotio cordis. In the Registry, blacks had a much lower survival rate than whites, and events that occur at home or during recreational sports are associated with lower survival than those in the setting of competitive sports, likely because of more rapid response times.

PREVENTION

Data from the Commotio Cordis Registry show that commotio cordis events can occur despite the use of safety baseballs and chest protectors (13). Although most baseball events have occurred with a standard baseball, there have been a small number that occurred with safety baseballs (11). Without knowing the relative number of chest impacts with standard versus safety baseballs, it is not possible to assess from these data whether safety baseballs are protective. In an

FIGURE Increasing Survival From Commotio Cordis Reported to the National Commotio Cordis Registry



Reprinted from Maron et al. (12) with permission. Copyright © 2013, Heart Rhythm Society. Published by Elsevier Inc. All rights reserved.

experimental model, the risk for commotio cordis decreased incrementally with softer balls (age-dependent safety baseballs), but safety baseballs were not absolutely protective against commotio cordis (6,8). This decrease was observed with chest wall impacts at both 30 and 40 mph.

Of the commotio cordis events that occurred in competitive sports, chest protectors were worn in 37% (12). Despite the use of these chest barriers, commotio cordis still occurs; in some sports, such as hockey, the chest protector can be raised with lifting of the arms, thereby uncovering the precordium and thus failing to provide protection. However, in other sports, such as baseball and lacrosse, the chest protectors have remained over the heart, and impact occurred through the barrier. Again, without knowing the relative number of impacts with or without chest protectors, these data cannot be interpreted with regard to risk. However, it is apparent that even with chest protection, prevention of commotio cordis is not absolute. In the swine model, commercial chest protectors for lacrosse and baseball did not lower the risk of commotio cordis (14). At impact velocities of 40 mph, the incidence of VF was similar among chest protectors and control impacts in which no chest protector was worn.

RETURN TO PLAY

Commotio cordis victims must undergo a complete cardiac workup to rule out structural heart disease. This includes but is not restricted to ECGs, echocardiograms, magnetic resonance imaging, ambulatory ECG monitoring, and stress testing. Pharmacological testing for Brugada and long-QT syndromes should also be considered in the presence of typical electrocardiographic features. Age-based electrocardiographic criteria should be applied, because T-wave abnormalities and QT intervals may be greater in the young (15). In those instances in which long-QT syndrome is a persistent concern, genetic testing could be considered. If underlying cardiac disease is absent, implantable cardioverter defibrillators are not recommended for survivors of commotio cordis.

Return-to-play decisions are largely dictated by the presence versus absence of underlying cardiac disease.

Given the large number of variables necessary to be confluent to trigger commotio cordis, a randomly occurring second event would be unlikely. Still, given some animal data for individual susceptibility to commotio cordis (16), it would be prudent to avoid sports that involve chest wall impact. Maturation of the chest wall with age also should lower the risk of recurrent commotio cordis.

CONCLUSIONS

Commotio cordis is an unusual event but still an important cause of morbidity and mortality in youth sports, as well as in many other circumstances. Absolute prevention will likely never be completely attainable, and thus, the most reasonable focus should be on recognition and resuscitation, including timely cardiopulmonary resuscitation and defibrillation.

Recommendations

1. Measures should be taken to ensure successful resuscitation of commotio cordis victims, including training of coaches, staff, and others to ensure prompt recognition, notification of emergency medical services, and institution of cardiopulmonary resuscitation and defibrillation (2,12,17) (Class I; Level of Evidence B).
2. A comprehensive evaluation for underlying cardiac pathology and susceptibility to arrhythmias should be performed in survivors of commotio cordis (2,4) (Class I; Level of Evidence B).
3. It is reasonable to use age appropriate safety baseballs to reduce the risk of injury and commotio cordis (6,8) (Class IIa; Level of Evidence B).
4. Rules governing athletics and coaching techniques to reduce chest blows can be useful to decrease the probability of commotio cordis (Class IIa; Level of Evidence C).
5. If no underlying cardiac abnormality is identified, then individuals can safely resume training and competition after resuscitation from commotio cordis (Class IIa; Level of Evidence C).

DISCLOSURES

Writing Group Disclosures

Writing Group Member	Employment	Research Grant	Other Research Support	Speakers Bureau/Honoraria	Expert Witness	Ownership Interest	Consultant/Advisory Board	Other
Mark S. Link	Tufts Medical Center	Unequal Technologies, a maker of sports equipment*; The National Operating Committee on Standards for Athletic Equipment (NOCSAE)*	None	None	None	None	None	None
N.A. Mark Estes III	Tufts Medical Center	None	None	None	None	None	Medtronic*; St. Jude Medical†; Boston Scientific†	None
Barry J. Maron	Minneapolis Heart Institute Foundation	None	None	None	None	None	None	None

This table represents the relationships of writing group members that may be perceived as actual or reasonably perceived conflicts of interest as reported on the Disclosure Questionnaire, which all members of the writing group are required to complete and submit. A relationship is considered to be "significant" if (a) the person receives \$10,000 or more during any 12-month period, or 5% or more of the person's gross income; or (b) the person owns 5% or more of the voting stock or share of the entity, or owns \$10,000 or more of the fair market value of the entity. A relationship is considered to be "modest" if it is less than "significant" under the preceding definition.

*Modest.

†Significant.

Reviewer Disclosures

Reviewer	Employment	Research Grant	Other Research Support	Speakers Bureau/Honoraria	Expert Witness	Ownership Interest	Consultant/Advisory Board	Other
George E. Billman	The Ohio State University	None	None	None	None	None	None	None
Ross A. Breckenridge	University College, London (England)	None	None	None	None	None	None	None
Charles B. Eaton	Memorial Hospital of Rhode Island	TBD	TBD	TBD	TBD	TBD	TBD	TBD
Bryan C. Cannon	Mayo Clinic, Rochester, MN	None	None	None	None	None	None	None
Michael S. Emery	Greenville Health System	None	None	None	None	None	None	None
Michael Lloyd	Emory University Hospital	Boston Scientific*	None	None	None	None	St. Jude Medical*; Medtronic*	None

This table represents the relationships of reviewers that may be perceived as actual or reasonably perceived conflicts of interest as reported on the Disclosure Questionnaire, which all reviewers are required to complete and submit. A relationship is considered to be "significant" if (a) the person receives \$10,000 or more during any 12-month period, or 5% or more of the person's gross income; or (b) the person owns 5% or more of the voting stock or share of the entity, or owns \$10,000 or more of the fair market value of the entity. A relationship is considered to be "modest" if it is less than "significant" under the preceding definition.

*Modest.

REFERENCES

- 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. <http://dx.doi.org/10.1056/NEJM199508103330602>.
- Maron BJ, Estes NA 3rd. Commotio cordis. *N Engl J Med.* 2010;362:917-27. <http://dx.doi.org/10.1056/NEJMr0910111>.
- Maron BJ, Ahluwalia A, Haas TS, Semsarian C, Link MS, Estes NA 3rd. Global epidemiology and demographics of commotio cordis. *Heart Rhythm.* 2011;8:1969-71. <http://dx.doi.org/10.1016/j.hrthm.2011.07.014>.
- Maron BJ. Sudden death in young athletes. *N Engl J Med.* 2003;349:1064-75. <http://dx.doi.org/10.1056/NEJMr022783>.
- Tenzer ML. The spectrum of myocardial contusion: a review. *J Trauma.* 1985;25:620-7.
- Link MS, Wang PJ, Pandian NG, Bharati S, Udelson JE, Lee MY, Vecchiotti MA, VanderBrink BA, Mirra G, Maron BJ, Estes NA 3rd. An experimental model of sudden death due to low-energy chest-wall impact (commotio cordis). *N Engl J Med.* 1998;338:1805-11. <http://dx.doi.org/10.1056/NEJM199806183382504>.
- Link MS, Maron BJ, VanderBrink BA, Takeuchi M, Pandian NG, Wang PJ, Estes NA 3rd. 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.
- Link MS, Maron BJ, Wang PJ, Pandian NG, VanderBrink BA, Estes NA 3rd. Reduced risk of sudden death from chest wall blows (commotio cordis) with safety baseballs. *Pediatrics.* 2002;109:873-7.
- Kalin J, Madias C, Alsheikh-Ali AA, Link MS. Reduced diameter spheres increases the risk of chest blow-induced ventricular fibrillation (commotio cordis). *Heart Rhythm.* 2011;8:1578-81. <http://dx.doi.org/10.1016/j.hrthm.2011.05.009>.
- Link MS, Maron BJ, Wang PJ, VanderBrink BA, Zhu W, Estes NA 3rd. Upper and lower limits of vulnerability to sudden arrhythmic death with chest-wall impact (commotio cordis). *J Am Coll Cardiol.* 2003;41:99-104.
- Maron BJ, Gohman TE, Kyle SB, Estes NA 3rd, Link MS. Clinical profile and spectrum of commotio cordis. *JAMA.* 2002;287:1142-6.

12. Maron BJ, Haas TS, Ahluwalia A, Garberich RF, Estes NA 3rd, Link MS. Increasing survival rate from commotio cordis. *Heart Rhythm*. 2013;10:219-23. <http://dx.doi.org/10.1016/j.hrthm.2012.10.034>.
13. Doerer JJ, Haas TS, Estes NA 3rd, Link MS, Maron BJ. Evaluation of chest barriers for protection against sudden death due to commotio cordis. *Am J Cardiol*. 2007;99:857-9. <http://dx.doi.org/10.1016/j.amjcard.2006.10.053>.
14. Weinstock J, Maron BJ, Song C, Mane PP, Estes NA 3rd, Link MS. Failure of commercially available chest wall protectors to prevent sudden cardiac death induced by chest wall blows in an experimental model of commotio cordis. *Pediatrics*. 2006;117:e656-62. <http://dx.doi.org/10.1542/peds.2005-1270>.
15. Rautaharju PM, Surawicz B, Gettes LS. AHA/ACCF/HRS recommendations for the standardization and interpretation of the electrocardiogram: part IV: the ST segment, T and U waves, and the QT interval: a scientific statement from the American Heart Association Electrocardiography and Arrhythmias Committee, Council on Clinical Cardiology; the American College of Cardiology Foundation; and the Heart Rhythm Society. *J Am Coll Cardiol*. 2009;53:1003-11. <http://dx.doi.org/10.1016/j.jacc.2008.12.016>.
16. Alsheikh-Ali AA, Madias C, Supran S, Link MS. Marked variability in susceptibility to ventricular fibrillation in an experimental commotio cordis model. *Circulation*. 2010;122:2499-504. <http://dx.doi.org/10.1161/CIRCULATIONAHA.110.955336>.
17. Travers AH, Rea TD, Bobrow BJ, Edelson DP, Berg RA, Sayre MR, Berg MD, Chameides L, O'Connor RE, Swor RA. Part 4: CPR overview: 2010 American Heart Association guidelines for cardiopulmonary resuscitation and emergency cardiovascular care. *Circulation*. 2010;122 suppl 3:S676-84. <http://dx.doi.org/10.1161/CIRCULATIONAHA.110.970913>.

KEY WORDS ACC/AHA Scientific Statements, athletes, cardiovascular abnormalities, commotio cordis, sudden cardiac death, ventricular fibrillation

AHA/ACC SCIENTIFIC STATEMENT

Eligibility and Disqualification Recommendations for Competitive Athletes With Cardiovascular Abnormalities: Task Force 14: Sick Cell Trait

A Scientific Statement From the American Heart Association and American College of Cardiology

Barry J. Maron, MD, FACC, *Chair**

Kevin M. Harris, MD, FACC*
 Paul D. Thompson, MD, FAHA,
 FACC*

E. Randy Eichner, MD*
 Martin H. Steinberg, MD*

Sickle cell trait (SCT), in which a normal hemoglobin gene and an abnormal mutated β -globin sickle gene (*HbS*) are inherited, occurs in 8% of blacks in the United States (0.08% of nonblacks) (1,2). SCT has been regarded as a benign condition that generally does not expose affected people to health risks, although for many years it has also been recognized as a potential cause of death in military training recruits during vigorous and intense physical exertion (3). More recently, evidence has been assembled proposing SCT as a cause of sudden death in competitive athletes,

usually during training and conditioning (4,5). In 2010, the National Collegiate Athletic Association (NCAA) mandated SCT screening (with solubility testing) for all student-athletes in division I sports (division II, 2012; division III, 2013). In addition, all newborns have been routinely tested for SCT shortly after birth since 1987 in accordance with a National Institutes of Health recommendation (1,6). Much of the controversy regarding SCT and athletes has focused on mandatory screening measures for the genetic defect, an issue that we have not addressed in this statement.

*On behalf of the American Heart Association Electrocardiography and Arrhythmias Committee of the Council on Clinical Cardiology, Council on Cardiovascular Disease in the Young, Council on Cardiovascular and Stroke Nursing, Council on Functional Genomics and Translational Biology, and the American College of Cardiology.

The American Heart Association and the American College of Cardiology make every effort to avoid any actual or potential conflicts of interest that may arise as a result of an outside relationship or a personal, professional, or business interest of a member of the writing panel. Specifically, all members of the writing group are required to complete and submit a Disclosure Questionnaire showing all such relationships that might be perceived as real or potential conflicts of interest. The Preamble and other Task Force reports for these proceedings are available online at www.onlinejacc.org (J Am Coll Cardiol 2015;XX:000-000; 000-000; 000-000; 000-000; 000-000; 000-000; 000-000; 000-000; 000-000; 000-000; 000-000; 000-000; and 000-000).

This statement was approved by the American Heart Association Science Advisory and Coordinating Committee on June 24, 2015, and the American Heart Association Executive Committee on July 22, 2015, and by the American College of Cardiology Board of Trustees and Executive Committee on June 3, 2015.

The American College of Cardiology requests that this document be cited as follows: Maron BJ, Harris KM, Thompson PD, Eichner ER, Steinberg MH; on behalf of the American Heart Association Electrocardiography and Arrhythmias Committee of the Council on Clinical Cardiology, Council on Cardiovascular Disease in the Young, Council on Cardiovascular and Stroke Nursing, Council on Functional Genomics and Translational Biology, and the American College of Cardiology. Eligibility and disqualification recommendations for competitive athletes with cardiovascular abnormalities: Task Force 14: sickle cell trait: a scientific statement from the American Heart Association and American College of Cardiology. J Am Coll Cardiol 2015;xx:●●●-●●●.

This article has been copublished in *Circulation*.

Copies: This document is available on the World Wide Web sites of the American Heart Association (<http://my.americanheart.org>) and the American College of Cardiology (www.acc.org). For copies of this document, please contact Elsevier Inc. Reprint Department via fax (212-633-3820) or e-mail (reprints@elsevier.com).

Permissions: Multiple copies, modification, alteration, enhancement, and/or distribution of this document are not permitted without the express permission of the American College of Cardiology. Requests may be completed online via the Elsevier site (<http://www.elsevier.com/about/policies/author-agreement/obtaining-permission>).

That SCT can be responsible for lethal sudden collapse (7), including on the athletic field, is based on evidence from the forensic-based US National Sudden Death in Athletes Registry (4,8,9) and other databases (10), as well as numerous case reports and considerable expert experience acquired in athletic venues (5). The epidemiology and characterization of SCT-related events in athletes are evolving. A large experience from the US Sudden Death in Athletes Registry documented SCT-associated collapse and death in 0.9% of 2462 athletes. This outcome occurred in 3.3% of the blacks in the registry (4). Ages of the victims were 12 to 22 years, and 90% were male. Events were most common in college football players during conditioning drills.

A distinctive clinical presentation has emerged that involves gradual deterioration over several minutes. Symptoms include cramping, dyspnea, muscle pain and severe weakness, and fatigue and exhaustion, provoked by vigorous physical exertion, often with sequential brief bursts of sustained maximal physical activity (e.g., interval training). Events typically occur early in the training season or after periods of deconditioning, often in ambient temperatures $\geq 80^{\circ}$, at high altitude, or associated with development of rhabdomyolysis (4,11-14). Notably, this scenario is in striking contrast to collapse caused by cardiovascular disease with ventricular tachyarrhythmias, which is typically virtually instantaneous (8,9).

Although the pathophysiology and clinical determinants of death in people with SCT participating in intense exercise are not fully understood, cardiovascular collapse likely occurs under conditions that (in laboratory studies) promote *HbS* polymerization and erythrocyte sickling. These include hyperthermia, dehydration, acidosis, and hypoxemia (11-14). It is possible that with intense exercise, a cascade of events ensues under unpredictable circumstances that recreates some of the laboratory conditions that lead to *HbS* polymerization and erythrocyte sickling, thereby triggering vascular occlusion, endothelial damage, and impaired muscular blood flow (12-16). This exertional sickling scenario could promote rhabdomyolysis and disseminated intravascular coagulation, which in turn could lead to hyperkalemia, lactic acidosis, worsening hypoxia, impaired cardiac and renal function, and lethal arrhythmias. However, widespread sickling in the heart and other organs identified at autopsy does not itself represent definitive evidence for SCT-related death, because postmortem *HbS* polymerization and erythrocyte sickling

are an expected consequence of the diminished oxygen environment at death.

These considerations have advanced specific precautionary recommendations for targeted and tailored measures during training for athletes with SCT to enhance the prevention of sudden death (15,16). These precautions, which can also benefit all athletes, include more gradual conditioning at the beginning of the training season (or after periods of deconditioning) with attention to modifying pace, providing adequate rest and hydration during conditioning drills, and promoting a high index of suspicion to immediately cease physical activity should muscle weakness, cramping or pain, fatigue, and disproportionately excessive dyspnea occur.

Indeed, collapse of an athlete with SCT is a medical emergency that requires support of vital signs, administration of supplemental oxygen, intravenous hydration, possibly cooling to protect against fulminating rhabdomyolysis, and likely rapid transport to a medical facility. A metabolic insult with lactic acidosis, hyperkalemia, and hypocalcemia can lead to pulseless electrical activity, so that the effectiveness of external defibrillation in this clinical setting is unpredictable. Such modified conditioning strategies and surveillance are now widely used by athletic trainers and coaching staffs in college athletic programs to prevent SCT-related complications and catastrophes.

SCT should now be included among the myriad of nontraumatic risks of sports participation capable of leading to the demise of some susceptible athletes, the vast majority of whom are black.

Recommendations

- 1. Recognition of SCT status is not itself a justification for disqualification from competitive sports (Class I; Level of Evidence C).**
- 2. Recommended preventive strategies (including adequate rest and hydration) should be performed to minimize the likelihood of an event occurring on the athletic field in a person known to have SCT (Class I; Level of Evidence B).**
- 3. It is critical to be prospectively aware of acute emergency medical strategies should suspicion of an emerging event arise in an athlete known to have SCT (Class I; Level of Evidence C).**
- 4. Particular caution should be exercised for athletes known to have SCT who are competing or training in high environmental temperatures or at extreme altitude (Class I; Level of Evidence C).**

DISCLOSURES

Writing Group Disclosures

Writing Group Member	Employment	Research Grant	Other Research Support	Speakers Bureau/Honoraria	Expert Witness	Ownership Interest	Consultant/Advisory Board	Other
Barry J. Maron	Minneapolis Heart Institute Foundation	None	None	None	None	None	None	None
E. Randy Eichner	University of Oklahoma	None	None	None	None	None	None	None
Kevin M. Harris	Minneapolis Heart Institute Foundation	None	None	None	None	None	None	None
Martin H. Steinberg	Boston University	None	None	None	None	None	None	None
Paul D. Thompson	Hartford Hospital	None	None	None	None	None	None	None

This table represents the relationships of writing group members that may be perceived as actual or reasonably perceived conflicts of interest as reported on the Disclosure Questionnaire, which all members of the writing group are required to complete and submit. A relationship is considered to be "significant" if (a) the person receives \$10,000 or more during any 12-month period, or 5% or more of the person's gross income; or (b) the person owns 5% or more of the voting stock or share of the entity, or owns \$10,000 or more of the fair market value of the entity. A relationship is considered to be "modest" if it is less than "significant" under the preceding definition.

Reviewer Disclosures

Reviewer	Employment	Research Grant	Other Research Support	Speakers Bureau/Honoraria	Expert Witness	Ownership Interest	Consultant/Advisory Board	Other
Michelle A. Grenier	University of Mississippi	None	None	None	None	None	None	None
Pranav Kansara	Christiana Care Health System	None	None	None	None	None	None	None
Mark S. Kindy	Medical University of South Carolina	None	None	None	None	None	None	None

This table represents the relationships of reviewers that may be perceived as actual or reasonably perceived conflicts of interest as reported on the Disclosure Questionnaire, which all reviewers are required to complete and submit. A relationship is considered to be "significant" if (a) the person receives \$10,000 or more during any 12-month period, or 5% or more of the person's gross income; or (b) the person owns 5% or more of the voting stock or share of the entity, or owns \$10,000 or more of the fair market value of the entity. A relationship is considered to be "modest" if it is less than "significant" under the preceding definition.

REFERENCES

- Steinberg MH. In the clinic: sickle cell disease. *Ann Intern Med.* 2011;155:ITC31-315. <http://dx.doi.org/10.7326/0003-4819-155-5-201109060-01003>.
- Goldsmith JC, Bonham VL, Joiner CH, Kato GJ, Noonan AS, Steinberg MH. Framing the research agenda for sickle cell trait: building on the current understanding of clinical events and their potential implications. *Am J Hematol.* 2012;87:340-6. <http://dx.doi.org/10.1002/ajh.22271>.
- Kark JA, Posey DM, Schumacher HR, Ruehle CJ. Sickle-cell trait as a risk factor for sudden death in physical training. *N Engl J Med.* 1987;317:781-7. <http://dx.doi.org/10.1056/NEJM198709243171301>.
- Harris KM, Haas TS, Eichner ER, Maron BJ. Sickle cell trait associated with sudden death in competitive athletes. *Am J Cardiol.* 2012;110:1185-8. <http://dx.doi.org/10.1016/j.amjcard.2012.06.004>.
- Eichner ER. Sickle cell trait in sports. *Curr Sports Med Rep.* 2010;9:347-51. <http://dx.doi.org/10.1249/JSR.0b013e3181fc73d7>.
- Bonham VL, Dover GJ, Brody LC. Screening student athletes for sickle cell trait: a social and clinical experiment. *N Engl J Med.* 2010;363:997-9. <http://dx.doi.org/10.1056/NEJMp1007639>.
- Taylor C, Kavanagh P, Zuckerman B. Sickle cell trait: neglected opportunities in the era of genomic medicine. *JAMA.* 2014;311:1495-6. <http://dx.doi.org/10.1001/jama.2014.2157>.
- Maron BJ. Sudden death in young athletes. *N Engl J Med.* 2003;349:1064-75. <http://dx.doi.org/10.1056/NEJMr022783>.
- Maron BJ, Doerer JJ, Haas TS, Tierney DM, Mueller FO. Sudden deaths in young competitive athletes: analysis of 1866 deaths in the United States, 1980-2006. *Circulation.* 2009;119:1085-92. <http://dx.doi.org/10.1161/CIRCULATIONAHA.108.804617>.
- Harmon KG, Drezner JA, Klossner D, Asif IM. Sickle cell trait associated with a RR of death of 37 times in National Collegiate Athletic Association football athletes: a database with 2 million athlete-years as the denominator. *Br J Sports Med.* 2012;46:325-30. <http://dx.doi.org/10.1136/bjsports-2011-090896>.
- Admanski C. Steelers' Ryan Clark Prominent in Sickle Cell Initiative Launch. *CBSports.com* Website. 2012. <http://www.cbssports.com/nfl/blog/nfl-rapidreports/20049491/steelers-ryan-clark-prominent-in-sickle-cell-initiative-launch>. Accessed March 12, 2013.
- Jones SR, Binder RA, Donowho EM Jr. Sudden death in sickle-cell trait. *N Engl J Med.* 1970;282:323-5. <http://dx.doi.org/10.1056/NEJM197002052820607>.
- Eichner ER. Sickle cell considerations in athletes. *Clin Sports Med.* 2011;30:537-49. <http://dx.doi.org/10.1016/j.csm.2011.03.004>.
- Tsaras G, Owusu-Ansah A, Boateng FO, Amoateng-Adjepong Y. Complications associated with sickle cell trait: a brief narrative review. *Am J Med.* 2009;122:507-12. <http://dx.doi.org/10.1016/j.amjmed.2008.12.020>.
- The National Athletic Trainers' Association (NATA) releases "Sickle Cell Trait and the Athlete" consensus statement [news release]. Dallas, TX: National Athletic Trainers' Association. June 27, 2007. <http://www.nata.org/NRO62107>. Accessed April 17, 2012.
- Klossner D. 2009-10 NCAA Sports Medicine Handbook. Indianapolis, IN: National Collegiate Athletic Association; 2009. <http://www.ncaapublications.com/productdownloads/MD10.pdf>. Accessed May 3, 2011.

KEY WORDS ACC/AHA Scientific Statements, athletes, cardiovascular abnormalities, sickle cell trait, sudden death

AHA/ACC SCIENTIFIC STATEMENT

Eligibility and Disqualification Recommendations for Competitive Athletes With Cardiovascular Abnormalities: Task Force 15: Legal Aspects of Medical Eligibility and Disqualification Recommendations

A Scientific Statement From the American Heart Association and American College of Cardiology

Matthew J. Mitten, JD, *Chair**

Douglas P. Zipes, MD, FAHA, MACC*

Barry J. Maron, MD, FACC*

William J. Bryant, JD*

From a legal perspective and medical perspective, protection of the health and safety of an athlete (as well as that of others potentially endangered by his or her participation) and avoidance of exposure to a significant risk of sudden cardiac death during competitive athletics should be the primary factors determining the exercise of clinical judgment and the making of medical recommendations regarding athletic participation by those with a cardiovascular

abnormality. A physician's general legal duty is to conform to accepted, customary, or reasonable medical practice providing medical sports participation recommendations consistent with an athlete's medical best interests from both a short- and long-term perspective (1,2). Courts generally have recognized that guidelines established by national medical associations are evidence of good medical practice, but they are not conclusive evidence of the medical or

*On behalf of the American Heart Association Electrocardiography and Arrhythmias Committee of the Council on Clinical Cardiology, Council on Cardiovascular Disease in the Young, Council on Cardiovascular and Stroke Nursing, Council on Functional Genomics and Translational Biology, and the American College of Cardiology.

The American Heart Association and the American College of Cardiology make every effort to avoid any actual or potential conflicts of interest that may arise as a result of an outside relationship or a personal, professional, or business interest of a member of the writing panel. Specifically, all members of the writing group are required to complete and submit a Disclosure Questionnaire showing all such relationships that might be perceived as real or potential conflicts of interest. The Preamble and other Task Force reports for these proceedings are available online at www.onlinejacc.org (J Am Coll Cardiol 2015;XX:000-000; 000-000; 000-000; 000-000; 000-000; 000-000; 000-000; 000-000; 000-000; 000-000; 000-000; 000-000; 000-000; 000-000; 000-000; 000-000; 000-000; 000-000; 000-000).

This statement was approved by the American Heart Association Science Advisory and Coordinating Committee on June 24, 2015, and the American Heart Association Executive Committee on July 22, 2015, and by the American College of Cardiology Board of Trustees and Executive Committee on June 3, 2015.

The American College of Cardiology requests that this document be cited as follows: Mitten MJ, Zipes DP, Maron BJ, Bryant WJ; on behalf of the American Heart Association Electrocardiography and Arrhythmias Committee of the Council on Clinical Cardiology, Council on Cardiovascular Disease in the Young, Council on Cardiovascular and Stroke Nursing, Council on Functional Genomics and Translational Biology, and the American College of Cardiology. Eligibility and disqualification recommendations for competitive athletes with cardiovascular abnormalities: Task Force 15: legal aspects of medical eligibility and disqualification recommendations: a scientific statement from the American Heart Association and American College of Cardiology. J Am Coll Cardiol 2015;xx:•••-•••.

This article has been copublished in *Circulation*.

Copies: This document is available on the World Wide Web sites of the American Heart Association (<http://my.americanheart.org>) and the American College of Cardiology (www.acc.org). For copies of this document, please contact Elsevier Inc. Reprint Department via fax (212-633-3820) or e-mail (reprints@elsevier.com).

Permissions: Multiple copies, modification, alteration, enhancement, and/or distribution of this document are not permitted without the express permission of the American College of Cardiology. Requests may be completed online via the Elsevier site (<http://www.elsevier.com/about/policies/author-agreement/obtaining-permission>).

legal standard of care (3-5). Avoidance of the unnecessary restriction of competitive athletic activity is a legitimate objective, but a physician's medical judgment should not be compromised by an athlete's strong desire to play a sport and willingness to assume a medically unreasonable risk, or by the team's need for an athlete's talents (6,7).

Knapp v Northwestern University (8), a 1996 federal appellate court case brought by a student-athlete claiming the legal right to play intercollegiate basketball contrary to a university team physician's medical recommendation (which was consistent with the then-current 26th Bethesda Conference guidelines) (9), established the current legal framework for resolving athlete challenges to medical disqualification based on cardiovascular abnormalities or events (10). Nicholas Knapp sued Northwestern University, claiming that its refusal to allow him to play on its basketball team violated the Rehabilitation Act, a federal law prohibiting educational institutions that receive federal funds from discriminating against people with covered disabilities. Although Northwestern agreed to honor Knapp's full athletic scholarship (which had been awarded before his incident of cardiac arrest), the university prohibited him from playing on its intercollegiate basketball team on the basis of its team physician's medical recommendation.

Knapp experienced sudden cardiac arrest while playing recreational basketball during the summer before his senior year in high school, which required cardiopulmonary resuscitation and defibrillation to restore sinus rhythm. Thereafter, he had an implantable cardioverter-defibrillator inserted and resumed playing recreational basketball without any subsequent cardiovascular events, although he did not play interscholastic basketball during his senior year. Northwestern's team physician refused to clear Knapp to play intercollegiate basketball on the basis of his medical records and history, the then-current 1994 26th Bethesda Conference recommendations, and the opinions of 2 consulting cardiologists who concluded that Knapp would expose himself to a medically unacceptable risk for ventricular fibrillation during competitive athletics, although 3 other cardiologists medically cleared him to play college basketball.

The Chicago, Illinois-based United States Court of Appeals for the Seventh Circuit held that a university has the legal right to establish legitimate physical qualifications for its intercollegiate athletes and that Northwestern did not violate the Rehabilitation Act by following its team physician's reasonable medical advice. It ruled that an intercollegiate athlete may be medically disqualified and excluded from a sport if necessary to avoid a "significant risk of personal physical injury" (which requires consideration of both the probability and severity of potential harm, including the 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" (8).

The court recognized that one of the factors a physician may rely on is then-current consensus medical guidelines:

"Although the Bethesda Conferences were not convened by public health officials and such guidelines should not substitute for individualized assessment of an athlete's particular physical condition, the consensus recommendations of several physicians in a certain field do carry weight and support the Northwestern team doctors' individualized assessment of Knapp" (8).

Consistent with the *Knapp* case, although some specialists provided medical clearance, another court also declined to "substitute its judgment" for a university team physician's "conservative" medical opinion that is "reasonable and rational" and consistent with other specialists' recommendations in federal disability discrimination litigation by a medically disqualified intercollegiate athlete against a university (11). These 2 cases hold that the federal disability discrimination laws (the Americans With Disabilities Act and the Rehabilitation Act) require only that a student-athlete's exclusion from an interscholastic or intercollegiate sport be based on an individualized medical evaluation and that disqualification must have a reasonable medical basis (8,11-13). Even if other physicians disagree, these laws are not violated if an educational institution accepts its team physician's reasonable medical judgment that a student-athlete should not be permitted to participate in a sport.

On the other hand, in *Mobley v Madison Square Garden LP* (14), a New York federal district court ruled that Cutino Mobley, a former NBA (National Basketball Association) basketball player, may have a valid state law

disability discrimination claim against the New York Knicks for refusing to allow him to play basketball with hypertrophic cardiomyopathy during the 2008 to 2009 season based on his medical disqualification by 2 cardiologists. In his complaint, Mobley alleged that he had been medically cleared to play NBA basketball from 1999 to 2008 (subject to his signing a liability waiver) and that 3 other cardiologists had examined him and concluded there was no material change in his heart condition and that he was as fit to play basketball in the fall of 2008 as he had been in 1998 and 2012. The court held that Mobley pled sufficient facts to contradict the medical opinions of the 2 cardiologists who had disqualified him and that it was “plausible that he was qualified to perform safely the essential functions of a professional basketball player,” which he ultimately had to prove to prevail on his New York disability discrimination law claim against the Knicks.

Mobley suggests that some courts may be willing to adopt an “athlete informed consent model” for professional athletes, in contrast to the *Knapp* court’s “team physician medical judgment model, which requires only that there be an individualized and reasonable medical basis for medically disqualifying college or high school athletes from participation in a sport” (12). By contrast, an “athlete informed consent model” would enable a professional athlete to choose to participate in a sport despite an individualized and reasonable medical disqualification by the team physician, if other competent medical authority clears him to play. However, it is important to understand that cases that apply federal and state disability discrimination laws such as *Knapp* and *Mobley* do not address or alter a physician’s legal duty to provide athletic participation recommendations consistent with good medical practice and necessary to protect an athlete’s health and safety, nor does either case rule that a liability waiver is enforceable and will immunize a physician from tort liability for failing to conform to acceptable, customary, or reasonable medical practice when making medical clearance recommendations for athletes at any level of competition (1,6). It is important to understand that *Mobley* does not hold that a physician’s “conservative” medical disqualification of an athlete with a cardiovascular abnormality constitutes malpractice, or that the decision of a professional team (or an educational institution) to exclude the athlete from participation based thereon necessarily violates federal or state disability discrimination laws.

To date, there is no legal precedent holding a physician liable for refusing to medically clear an athlete with a known or probable cardiovascular abnormality or implantable cardioverter-defibrillator consistent with consensus guidelines, or declining to do so based on a medically reasonable belief that participation in a sport would expose the athlete or others to a significantly

enhanced risk of sudden cardiac death or serious injury. The most similar case is *Penny v Sands*, a 1989 lawsuit in which Anthony Penny alleged that a cardiologist was negligent for misdiagnosing his heart condition as cardiomyopathy and medically disqualifying him to play college basketball when other cardiologists had medically cleared him (6). Penny died while playing professional basketball in England before the court decided the merits of his medical malpractice claim, so this case does not establish any legal precedent. To avoid interfering with a physician’s medical judgment and recommendations to protect athletes’ health and safety, it is unlikely that a court would impose malpractice liability for refusing to provide medical clearance to an athlete to participate in a competitive sport with a properly diagnosed cardiovascular abnormality or implantable cardioverter-defibrillator (6).

Like the 26th Bethesda Conference guidelines in 1994 (9) and the 36th Bethesda Conference guidelines in 2005 (15), the updated 2015 American Heart Association/American College of Cardiology recommendations regarding the medical appropriateness of participation in particular competitive sports for a person with a confirmed or probable cardiovascular abnormality are “generally conservative,” although some of them are less restrictive on the basis of additional data and athletic participation experiences since 2005. As stated in the Preamble (16), the current recommendations in this document “are not intended to establish absolute mandates” that must be followed in all cases or the medical standard of care. Rather, it is “a consensus reference document that is potentially helpful in resolving predictably difficult clinical dilemmas.”

In specific cases, it may be consistent with accepted, customary, or reasonable medical practice for a physician to deviate from the American Heart Association/American College recommendations by providing medical clearance based on individualized factors evidencing that participation by an athlete with a cardiovascular abnormality in a particular sport would not create a significant risk of sudden cardiac death or other serious injury to the athlete or others. If a physician does so, it is important to fully inform the athlete of the potential material risks of participating in a competitive sport, preferably in writing, even if they are deemed to be medically reasonable (1,6). It also would be legally permissible for a physician to medically disqualify an athlete consistent with the 36th Bethesda Conference guidelines in individualized situations if there is a reasonable medical, scientific, or clinical basis for doing so. In other words, although the current American Heart Association/American College guidelines could permit athletic participation in a sport with the subject cardiovascular abnormality or an implantable cardioverter-defibrillator, or although some athletes (including Nicholas Knapp, who played intercollegiate basketball for 2 years at Ashland University after he left

Northwestern) have done so without serious adverse health consequences (17-20), the current guidelines do not require that medical clearance be provided in such

cases. Rather, these guidelines are one of the factors a physician should consider in exercising medical best judgment in individual situations.

DISCLOSURES

Writing Group Disclosures

Writing Group Member	Employment	Research Grant	Other Research Support	Speakers Bureau/Honoraria	Expert Witness	Ownership Interest	Consultant/Advisory Board	Other
Matthew J. Mitten	Marquette University	None	None	None	None	None	None	None
William J. Bryant	Dominick, Feld Hyde PC	None	None	None	None	None	None	None
Barry J. Maron	Minneapolis Heart Institute Foundation	None	None	None	None	None	None	None
Douglas P. Zipes	Indiana University	None	None	None	None	None	None	None

This table represents the relationships of writing group members that may be perceived as actual or reasonably perceived conflicts of interest as reported on the Disclosure Questionnaire, which all members of the writing group are required to complete and submit. A relationship is considered to be "significant" if (a) the person receives \$10,000 or more during any 12-month period, or 5% or more of the person's gross income; or (b) the person owns 5% or more of the voting stock or share of the entity, or owns \$10,000 or more of the fair market value of the entity. A relationship is considered to be "modest" if it is less than "significant" under the preceding definition.

Reviewer Disclosures

Reviewer	Employment	Research Grant	Other Research Support	Speakers Bureau/Honoraria	Expert Witness	Ownership Interest	Consultant/Advisory Board	Other
Michael S. Emery	Greenville Health System	None	None	None	None	None	None	None
Timothy F. Feltes	Nationwide Children's Hospital/The Ohio State University	None	None	None	None	None	None	None
Dave Herbert	David L Herbert & Associates, LLC	None	None	None	None	None	None	None
David T. Hardman	The Canberra Hospital, Canberra (Australia)	None	None	None	None	None	None	None

This table represents the relationships of reviewers that may be perceived as actual or reasonably perceived conflicts of interest as reported on the Disclosure Questionnaire, which all reviewers are required to complete and submit. A relationship is considered to be "significant" if (a) the person receives \$10,000 or more during any 12-month period, or 5% or more of the person's gross income; or (b) the person owns 5% or more of the voting stock or share of the entity, or owns \$10,000 or more of the fair market value of the entity. A relationship is considered to be "modest" if it is less than "significant" under the preceding definition.

REFERENCES

- Mitten MJ. Emerging legal issues in sports medicine: a synthesis, summary, and analysis. *St John's Law Rev.* 2002;76:5-86.
- Champion WT Jr. Fundamentals of Sports Law. Deerfield, IL: Clark Boardman Callaghan; 2000. Chapter 4, §4.1:91-8.
- Stone v Proctor, 131 SE2d 297, 299 (NC 1963).
- Pollard v Goldsmith, 572 P2d 1201, 1203 (Ariz Ct App 1977).
- Swank v Halivopoulos, 260 A2d 240, 242-3 (NJ Super Ct App Div 1969).
- Mitten MJ. Team physicians and competitive athletes: allocating legal responsibility for athletic injuries. *Univ Pitt Law Rev.* 1993;55:129-60.
- Di Luca TR. Medical malpractice and the modern athlete: a whole new ballgame ...or is it? *Westchester County Bar Assoc J.* 2008;35:17-26.
- Knapp v Northwestern University, 101 F3d 473 (7th Cir 1996), cert denied, 520 US 1274 (1997).
- Maron BJ, Mitchell JH. 26th Bethesda Conference: recommendations for determining eligibility for competition in athletes with cardiovascular abnormalities [published correction appears in *Med Sci Sports Exerc.* 1994;26:followi]. *J Am Coll Cardiol.* 1994;24:845-99.
- 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. <http://dx.doi.org/10.1056/NEJM199811263392211>.
- Pahulu v University of Kansas, 897 F Supp 1387 (D Kan 1995).
- Mitten MJ. Enhanced risk of harm to one's self as justification for exclusion from athletics. *Marquette Sports Law Rev.* 1998;8:189-223.
- Weston MA. The intersection of sports and disability: analyzing reasonable accommodations for athletes with disabilities. *St Louis Univ Law J.* 2005;50:137-63.
- Mobley v Madison Square Garden LP, 2013 US Dist LEXIS (SD New York 2012).
- Maron BJ, Zipes DP. Introduction: eligibility recommendations for competitive athletes with cardiovascular abnormalities: general considerations. *J Am Coll Cardiol.* 2005;45:1318-21. <http://dx.doi.org/10.1016/j.jacc.2005.02.006>.
- Maron BJ, Zipes DP, Kovacs RJ, on behalf of the American Heart Association Electrocardiography and Arrhythmias Committee of the Council on Clinical Cardiology, Council on Cardiovascular Disease in the Young, Council on Cardiovascular and Stroke Nursing, Council on Functional Genomics and Translational Biology, and the American College of Cardiology. Eligibility and disqualification recommendations for competitive athletes with cardiovascular abnormalities: preamble, principles, and general considerations: a scientific statement from the American Heart Association and American College of Cardiology. *J Am Coll Cardiol.* 2015 In Press. <http://dx.doi.org/10.1016/j.jacc.2015.09.046>.
- Kranhold K, Helliker K. Cardiologist helps athletes get back in game. *Wall Street Journal.* May 12, 2012. <http://www.post-gazette.com/news/health/2006/07/25/cardiologist-helps-athletes-get-back-in-the-game/stories/200607250235>. Accessed August 25, 2015.
- Katz A. Negedu moves on after brush with death. *ESPN Website.* May 18, 2010. http://sports.espn.go.com/ncb/columns/story?columnist=katz_andy&id=5197641. Accessed August 25, 2015.
- Deleted in proof.
- Medcalf M. Taking a closer look at heart issues. *ESPN Website.* August 29, 2012. http://espn.go.com/mens-college-basketball/story/_/id/8313100/when-hearts-young-athletes-fail-college-basketball. Accessed August 25, 2015.

KEY WORDS ACC/AHA Scientific Statements, athletes, cardiovascular abnormalities, disqualification, eligibility, legal