

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

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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 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

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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 (eg, 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 (eg, 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 (eg, 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 States¹⁸ and Israel⁷ 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 routinely¹⁸; 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 5000-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 athletes¹¹ 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 ($\approx 30\%$ – 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 (ie, ≈ 10 – 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 (ie, annual) ECG screening during adolescence, given the possibility of developing phenotypic evidence of cardiomyopathies during this time period or later³³; (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 position¹ 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 (eg, 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 non-athletes 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*).

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*Modest.

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*Modest.

†Significant.

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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

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