Special Communication

Cardiac Screening of Young Athletes Prior to Participation in Sports
Difficulties in Detecting the Fatally Flawed Among the Fabulously Fit

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Deaths of young athletes from cardiac disease are uncommon but receive considerable media attention and intermittently galvanize debates about cardiac screening prior to participation in sports. Both the American Heart Association (AHA) and European Society of Cardiology (ESC) endorse preparticipation screening in athletes; however, there is disagreement about the best approach. The AHA recommends history and physical examination; this approach is pragmatic and relatively inexpensive but has poor sensitivity because most athletes are asymptomatic and physical examination identifies only a minority of those at risk of sudden cardiac death. The inclusion of the electrocardiogram in accordance with the recommendations of the ESC improves sensitivity for detection of serious cardiac disease but is associated with an unacceptably high false-positive rate, in part because of the overlap between the electrical manifestations of athletic training and the cardiomyopathies. For young athletes with normal electrocardiogram results, echocardiography contributes minimally to the diagnosis of serious cardiac diseases. Given all the complexities, cardiac screening of young athletes should be voluntary not mandatory and conducted by highly experienced physicians who fully understand the cardiovascular adaptation to intensive exercise.

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The benefits of regular exercise on the cardiovascular system are established, and young athletes are considered to represent the healthiest segment of society. Therefore, when a young athlete dies, his or her death is frequently publicized through the news media and captures the hearts of society as a whole. The majority of deaths in athletes are due to inherited or congenital cardiac diseases that may cause serious ventricular arrhythmias during exercise but generally have low adverse event rates (0.2%-0.8% per annum) and are compatible with a normal quality and quantity of life.

Whereas the actual incidence of sudden cardiac death in athletes is low, ranging from 1 in 50 000 to 1 in 200 000,[1,2] several electrocardiographic studies worldwide have demonstrated that 1 in 300 young athletes harbor a cardiac condition that has the potential to cause exercise-related sudden death.[1,3] The unpredictability of such catastrophes is often considered a compelling justification for a structured preparticipation screening program including a health questionnaire, physical examination, and possibly other cardiac investigations to identify those at risk. This obligation is magnified when consideration is given to the fact that most cardiac conditions are detectable during life and there are therapeutic strategies available to minimize the risk of that such conditions will have catastrophic consequences. The American Heart Association (AHA)[4] and the European Society of Cardiology (ESC)[5] advocate cardiac screening of young athletes prior to participation in sports. On the basis of the premise that sudden cardiac death is preventable, both groups aspire to identify most athletes with cardiac disease using relatively practical and cost-effective methods.

Current Screening Models

The inclusion of the electrocardiogram is associated with a significant increase in sensitivity through its ability to detect congenital accessory pathways and ion channelopathies. Whereas the cardiomyopathies rely on cardiac imaging studies for diagnosis, between 80% and 95% of young patients with a cardiomyopathy have an abnormal electrocardiogram result, which may provide the first index of suspicion of disease.

It is debatable whether the detection of disease equates to a reduction in sudden cardiac deaths. The most persuasive evidence for the efficacy of electrocardiography-based screening in reducing sudden cardiac death is derived from a large prospective Italian study.[1] Over a 25-year period, the investigators demonstrated that electrocardiographic screening in athletes reduced the incidence of sudden cardiac death by 90%, from 3.6 deaths per 100 000 person-years to 0.43 deaths per 100 000 person-years.[1] These results are contradicted by a study from Minneapolis, which showed similar mor-

tality rates in young athletes with comparable demographic characteristics using the approach recommended by the AHA.[6] Another study from Israel failed to demonstrate any reduction in death rates in young athletes during a 12-year period prior to mandatory electrocardiography screening compared with a 12-year period of electrocardiography screening.[7] The Italian study based its conclusions on a systematic sudden cardiac death registry in athletes, which included data on cardiac pathology. In contrast, the documentation of sudden cardiac death in the United States and Israeli studies relied on media, web-based search engines and insurance claims documents. It is possible that the US and Israeli studies failed to capture a substantial proportion of deaths.[8]

Pitfalls of Electrocardiographic Screening

The electrocardiogram does not identify all athletes at risk of death, particularly those with anomalous coronary origins or premature atherosclerotic coronary disease. In our experience, these entities account for at least one-sixth of all sudden cardiac deaths in young athletes. The overlap between the electrical manifestations of athletic training and the cardiomyopathies is associated with a false-positive rate of 9% to 25% for electrocardiograms, which is unacceptably high. On the basis of studies in college athletes, Baggish et al[9] reported that the addition of electrocardiography to history and physical examination improved the sensitivity of detecting a cardiac abnormality from 45% to 90% but reduced the specificity from 94% to 83%, with no certainty that the abnormalities detected by electrocardiography would ultimately cause a cardiac event. In this study, the false-positive rate for history and physical examination alone was 5.5% compared with 16.9% after electrocardiography was included. Despite major advances in the interpretation of the electrocardiogram in athletes, the false-positive rate in white athletes is 5% among the most experienced physicians. The situation is worse in athletes of African/Afro-Caribbean ancestry; false-positive rates still exceed 10%.[3] The cost of follow-up investigations to confirm or refute the presence of a serious cardiac disorder and the potential to unfairly disqualify athletes from participating in sports are causes for concern.

The positive predictive value of an abnormal electrocardiogram result to detect a serious cardiac disorder is poor. In a large study of more than 5000 British athletes, the positive predictive value of the 12-lead electrocardiogram was 7.4% in white athletes and 3.1% in black athletes.[3] Chandra et al[10] evaluated 7764 nonathletes aged 14 to 35 years with electrocardiography screening. Among these individuals, 784 (10%) exhibited electrocardiographic changes that would have prompted an echocardiogram to confirm or refute an underlying cardiomyopathy. Subsequent echocardiography in all 784 individuals showed that only 2% had either left ventricular hypertrophy or left and/or right ventricular dilatation that overlapped with the dimensions observed in patients with cardiomyopathy.

How Useful Is Echocardiography?

Echocardiography is considered the most practical method for detecting structural cardiac defects. Its use as a screening tool is generally confined to the most elite echelons of sport. Such practice reflects the requirement for clubs or teams to have sufficient financial resources to offer comprehensive assessment for their professional athletes. The routine use of echocardiography, however, is debatable when cost is a consideration. Echocardiographic studies in athletes have revealed the presence of minor congenital abnormalities even when the electrocardiogram result is normal[9]; however, these abnormalities are rarely implicated in sudden cardiac death. Although echocardiography may identify serious diseases such as anomalous coronary arteries, aortic root dilatation, and mild cardiomyopathy in the absence of an abnormal electrocardiogram result, the diagnostic yield in asymptomatic athletes with normal physical examination and electrocardiogram findings is low. Magalski et al[11] studied 964 consecutive US collegiate athletes with medical history, physical examination, electrocardiography, and echocardiography. Ninety-five athletes (10%) had a distinctly abnormal electrocardiogram result, and 9 athletes were subsequently diagnosed as having a potentially serious cardiac condition. Echocardiography did not identify any additional abnormalities in the context of a normal electrocardiogram and physical examination results.

The structural changes due to physiological adaptation of the heart to long-term intensive exercise frequently overlap with changes observed in cardiomyopathy, particularly in male athletes. Of male athletes, 14% exhibit a left ventricular cavity size resembling dilated cardiomyopathy. In addition, 12% of endurance athletes show a reduced baseline ejection fraction, 13% to 18% of black male athletes have a left ventricular wall thickness compatible with morphologically mild hypertrophic cardiomyopathy, and 37% of male athletes have a right ventricular cavity dimension that fulfils a criterion in the revised "task force criteria" for diagnosing arrhythmogenic right ventricular cardiomyopathy.[12-14] More recently, echocardiographic techniques to improve tissue characterization have led to 8% of athletes fulfilling echocardiographic criteria for isolated left ventricular noncompaction.[15] This is a recently recognized cardiomyopathy that is characterized by increased trabeculations within the left ventricular myocardium and is associated with progressive heart failure, a predilection to fatal arrhythmias, and an increased risk of systemic thromboembolism.[16] In a recent case, expert opinions differed regarding the cessation of competitive sport in an athlete with increased left ventricular trabeculation who was judged to have left ventricular noncompaction.[17] In summary, echocardiography is an excellent test at excluding cardiomyopathy in relatively sedentary individuals. Paradoxically, however, the morphological alterations associated with athletic training have a greater potential to lead to erroneous diagnoses, particularly when cardiologists with insufficient expertise in evaluating athletes perform the studies.

Perspective

The prevention of sudden cardiac death in young athletes is important for public health; however, screening young athletes for cardiac disease is less straightforward than the screening is often portrayed. History and physical examination are important but alone are insensitive, electrocardiography has a relatively low specificity, and echocardiography frequently reveals cardiac dimensions that are normal for athletes but that overlap with those for primary cardiomyopathies, leading to false-positive results. More comprehensive tests including an exercise stress test, 24-hour electrocardiog-

raphy and cardiac magnetic resonance imaging are cost prohibitive. Among inexperienced physicians, current screening strategies have the potential to cause more harm than good. The long-term physical and psychological impact for athletes disqualified because of an erroneous diagnosis has not been examined.

The electrocardiogram is probably the most cost-effective method for detecting athletes with cardiac disease, with the caveat that the physicians interpreting the studies should have expertise in evaluating the electrocardiographic appearance of athletes' hearts. In such circumstances, it is reasonable to provide rapid clearance and limit anxiety by performing on site echocardiography in athletes with abnormal physical examination or electrocardiogram results. In our experience and in other studies, such practice provides clearance for up to 97% of athletes in 1 evaluation.[18] Still, the impact of electrocardiography on preventing sudden cardiac death in athletes requires further study to compare death rates in young competitive athletes who have been screened with unscreened athletes from the same population.

Although screening the most elite athletes is feasible, the practicalities associated with the development of new screening programs for all young athletes are difficult to ignore. Most countries do not possess the infrastructure, personnel, or the expertise to offer a cardiac evaluation to all young individuals who exercise. The argument to confine screening to elite athletes is ironic and invalid because the presence of a cardiomyopathy likely selects out most individuals capable of competing at the elite level but may be conducive to less-intensive exercise among recreational athletes.

Conclusions

The detection of potentially serious cardiac disease in athletes is important. Given all the complexities, cardiac screening of young athletes should be voluntary and not mandatory, but should be conducted by highly experienced physicians who fully understand the cardiovascular adaptation to intensive exercise.

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REFERENCES

1. 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(13):1593-1601.

2. Roberts WO, Stovitz SD. Incidence of sudden cardiac death in Minnesota high school athletes 1993-2012 screened with a standardized pre-participation evaluation. *J Am Coll Cardiol*. 2013;62(14):1298-1301.

3. Sheikh N, Papadakis M, Ghani S, et al. Comparison of electrocardiographic criteria for the detection of cardiac abnormalities in elite black and white athletes. *Circulation*. 2014;129(16):1637-1649.

4. Maron BJ, Thompson PD, Ackerman MJ, et al; American Heart Association Council on Nutrition, Physical Activity, and Metabolism. Recommendations and considerations related to preparticipation screening for cardiovascular abnormalities in competitive athletes: 2007 update: a scientific statement from the American Heart Association Council on Nutrition, Physical Activity, and Metabolism: endorsed by the American College of Cardiology Foundation. *Circulation*. 2007;115(12):1643-1655.

5. Corrado D, Pelliccia A, Heidbuchel H, et al; Section of Sports Cardiology, European Association of Cardiovascular Prevention and Rehabilitation. Recommendations for interpretation of 12-lead electrocardiogram in the athlete. *Eur Heart J*. 2010; 31(2):243-259.

6. Maron BJ, Haas TS, Doerer JJ, Thompson PD, Hodges JS. Comparison of US and Italian experiences with sudden cardiac deaths in young competitive athletes and implications for preparticipation screening strategies. *Am J Cardiol*. 2009;104(2):276-280.

7. Steinvil A, Chundadze T, Zeltser D, et al. Mandatory electrocardiographic screening of athletes to reduce their risk for sudden death proven fact or wishful thinking? *J Am Coll Cardiol*. 2011;57(11):1291-1296.

8. Drezner JA, Harmon KG, Marek JC. Incidence of sudden cardiac arrest in Minnesota high school student athletes: the limitations of catastrophic insurance claims. *J Am Coll Cardiol*. 2014;63(14): 1455-1456.

9. Baggish AL, Hutter AM Jr, Wang F, et al. Cardiovascular screening in college athletes with and without electrocardiography: a cross-sectional study. *Ann Intern Med*. 2010;152(5):269-275.

10. Chandra N, Bastiaenen R, Papadakis M, et al. Prevalence of electrocardiographic anomalies in young individuals: relevance to a nationwide cardiac screening program. *J Am Coll Cardiol*. 2014; 63(19):2028-2034.

11. Magalski A, McCoy M, Zabel M, et al. Cardiovascular screening with electrocardiography and echocardiography in collegiate athletes. *Am J Med*. 2011;124(6):511-518.

12. Zaidi A, Ghani S, Sharma R, et al. Physiological right ventricular adaptation in elite athletes of African and Afro-Caribbean origin. *Circulation*. 2013;127(17):1783-1792.

13. Marcus FI, McKenna WJ, Sherrill D, et al. Diagnosis of arrhythmogenic right ventricular cardiomyopathy/dysplasia: proposed modification of the task force criteria. *Circulation*. 2010;121(13): 1533-1541.

14. Sheikh N, Sharma S. Impact of ethnicity on cardiac adaptation to exercise. *Nat Rev Cardiol*. 2014;11(4):198-217.

15. Gati S, Chandra N, Bennett RL, et al. Increased left ventricular trabeculation in highly trained athletes: do we need more stringent criteria for the diagnosis of left ventricular non-compaction in athletes? *Heart*. 2013;99(6):401-408.

16. Thavendiranathan P, Dahiya A, Phelan D, Desai MY, Tang WHW. Isolated left ventricular non-compaction controversies in diagnostic criteria, adverse outcomes and management. *Heart*. 2013;99(10):681-689.

17. Peritz DC, Vaughn A, Ciocca M, Chung EH. Hypertrabeculation vs left ventricular noncompaction on echocardiogram: a reason to restrict athletic participation? *JAMA Intern Med*. 2014;174(8):1379-1382.

18. Asif IM, Johnson S, Schmieg J, et al. The psychological impact of cardiovascular screening: the athlete's perspective. *Br J Sports Med*. 2014;48 (15):1162-1166.

19. 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(13):1593-1601.

20. Vetter VL, Dugan NP. A discussion of electrocardiographic screening and sudden cardiac death prevention: evidence and consensus. *Curr Opin Cardiol*. 2013;28(2):139-151.

21. Wilson MG, Basavarajaiah S, Whyte GP, Cox S, Loosemore M, Sharma S. Efficacy of personal symptom and family history questionnaires when screening for inherited cardiac pathologies: the role of electrocardiography. *Br J Sports Med*. 2008;42 (3):207-211.