

Saving Athletes' Lives

A Reason to Find Common Ground?

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Although both U.S. and European guidelines recommend screening athletes for suspected heart disease before participation in competitive sport, they disagree on which findings are associated with increased risk as well as those athletes whose activities should be restricted. Recent Italian data suggest that screening programs can save lives, but few countries have fully adopted such practices. It is time to develop an international consensus on who should receive pre-participation screening, who should perform the screening, what tests the screening should consist of, and what criteria should be used to restrict participation. (J Am Coll Cardiol 2008;52:1997-9) © 2008 by the American College of Cardiology Foundation

Extraordinary physical fitness does not confer immunity to the tragedy of sudden cardiac death (SCD). Although the combination of some forms of heart disease with exercise can be lethal and there is universal agreement about the need to screen athletes for suspected heart disease before participation in competitive sport, recommendations as to how best to accomplish this differ from one side of the Atlantic to the other (1,2), and actual practice is even more variable. Disagreement also exists on which findings are associated with increased risk as well as the guidelines for who should not participate in competitive sport (3-5). Perhaps not surprisingly, given this lack of consensus, much of what we know is based on expert opinion rather than substantive prospective research, and few cardiologists have extensive experience with athletes at risk.

In this issue of the *Journal*, Corrado et al. (6) provide details on one such extensive experience through a retrospective observational analysis of a regional Italian pre-participation screening program. Their data show a remarkable drop in the rate of SCD in athletes after program initiation, from approximately 4/100,000 to <1/100,000, whereas the rate of athlete disqualification due to suspected cardiomyopathy doubled. The SCD rate for athletes is now the same as the general Italian population, but it is also the same as the rate in the athletic population in the U.S., which has less rigorous pre-participation screening programs. Although some of these data have been previously presented (7,8), the present article emphasizes the implications of this unique program, which have not been universally accepted, and the lessons we might learn from them. It is a timely message in this Olympic year.

One of the first lessons is to more clearly define the goals of screening. Corrado et al. (6) emphasize that they seek to identify a population of individuals who might be at elevated risk rather than single individuals with heart disease. This subtle shift in focus leads to a higher rate of subsequent testing (usually echocardiography) than has been reported in at least 1 American series (9% vs. 3%) (9) and mitigates the impact of any lack of specificity in the initial screening. However, the diseases responsible for causing SCD might differ in this region with its higher prevalence of right ventricular cardiomyopathies and higher death rate present before beginning the screening program, possibly affecting these results and their "translatability" to other countries. Such issues are difficult to resolve; the underlying prevalence of disease is still in dispute, even for well-studied entities such as hypertrophic cardiomyopathy (10).

Additional lessons can be learned regarding the Italians' screening methodology and personnel (6). Most important of these is the use of electrocardiograms (ECGs) as part of their screening program in addition to a cardiac history and physical, a strategy endorsed by the European Society of Cardiology but not the American Heart Association guidelines (1,2) and that remains controversial in the U.S. (11,12). In spite of this, screening methods have become significantly more rigorous in the U.S. over the past decade, and although still not requiring an ECG, more states now require screening, and the mandated questionnaires more completely explore cardiovascular risk (13). The Italian screeners are sports physicians, with 4 years of post-graduate training in sports medicine and cardiology, including the recognition of ECG abnormalities in athletes, and all were in sports medicine practice full time. In contrast, in the U.S., over time screeners are becoming less likely to be physicians or even trained in sports medicine (13).

**Abbreviations
and Acronyms**

ECG = electrocardiogram

SCD = sudden cardiac
death

In addition to the size of athletic programs in the U.S. and the limited availability of trained sports medicine physicians, the costs of implementing a similar program might be prohibitive. In Italy, screening costs about €30/athlete and, along with any needed subsequent testing, is at least partially supported by the National Health System. Furthermore, a favorable environment regarding possible liability of the screening program for missed diagnoses is also present (6). Other incompletely resolved methodologic questions include the competitive level of athletes who should be screened, the frequency of repeat screening, and the criteria used to determine the need for additional testing. Every screening program must explicitly or implicitly wrestle with these issues, and the experience of Corrado et al. (6) can provide some guidance in the absence of clear guidelines.

Another lesson is related to the link between the effectiveness of screening and reduced death, which can only be through the actions taken in response to screening results. Once again European and American recommendations from an American College of Cardiology Bethesda Conference differ, as nicely dissected by Pelliccia et al. (3-5,14). Although both documents advocate restricting activity in athletes deemed to be at risk, definitions of who is at risk vary significantly, and the level of exercise limitation recommended is not uniform. The European guidelines are more restrictive in many cases, tilting the balance toward proscription of activity rather than individual freedom to pursue sport. The result of this recommendation is demonstrated in practice by the high exclusion rate in the Italian programs of 2% of those screened, despite only 0.2% having "potentially lethal" conditions (7). The "pursuit of happiness" is a characteristically American concept, and the existence of a U.S. legal precedent that allows athletes to choose to participate despite elevated risk is relevant to the content of both screening and participation guidelines and the ability to implement them in practice.

In addition to such cultural differences, the European but not the American guidelines advise against participation for genotype-positive, phenotype-negative athletes with hypertrophic cardiomyopathy, Brugada syndrome, long-QT syndrome, and Marfan syndrome. Extrapolation of these recommendations suggests that consideration should be given to genotyping asymptomatic athletes for mutations known to be associated with these diseases as part of pre-participation screening. The implications are staggering in terms of the required expense and expertise (not coincidentally, these same objections are raised to support screening without using ECGs) and in terms of ethics: could the opportunity to participate in sport be reasonably withheld for a positive genotype with no other manifestations of disease? Furthermore, if one were to accept this hypothetical approach, the possibility of prejudice based on genetic

information is substantial. Perhaps fortuitously, Van Driest et al. (15) found that only 30% to 61% of individuals with phenotypic hypertrophic cardiomyopathy had previously reported and therefore detectable mutations, suggesting that the yield of genetic screening would be low at present. Even if genotyping were performed only in those who have positive clinical features (i.e., a pre-clinical phenotype or family history), it is unclear how the extra expense of testing for genetic risk and the uncertainty with regard to acting upon its results would be handled, especially in light of the lack of agreement on the proper use of a time-honored and inexpensive test such as an ECG. However, it is only a matter of time until advances in genetic science and technologies make this approach more accurate and perhaps even feasible. Until such questions can be answered, one must assume that the European guidelines are to be followed with a "don't ask, don't tell" approach, whereby genetic testing is only performed once other parameters indicate the possible presence of disease.

Whereas issues such as genetic testing will continue to evolve, it is critical to ask what level of evidence would be required to harmonize current approaches to pre-participation screening. At present, although most countries in Europe have slightly different policies, most do require an ECG as part of pre-participation testing. In the U.S. a growing consumer movement is increasingly bypassing the ECG in favor of screening with an echocardiogram (16). Although the echocardiogram is arguably more sensitive and certainly more specific than the ECG for diagnosis of hypertrophic cardiomyopathy, its accuracy for other abnormalities causing cardiac arrest, such as the "channelopathies," is arguably worse and certainly untested. In the likely continued absence of randomized trials testing alternative screening strategies, despite calls for such research (11,12), are the observational results of Corrado et al. (6) powerful enough to alter U.S. guidelines and, most importantly, screening practices? If not, what level of evidence would be required, and how can we develop it? Similar questions revolve around testing the validity of differing participation recommendations where even less evidence is available.

Few things are more universally mourned than the sudden death of an athlete. Yet our scientific and clinical community has responded in a fragmented manner, which can leave the sports community confused as to how to ensure the health of athletes, clinicians uncertain how to care for an individual patient, and parents of young athletes in fear for their offspring's health. To develop a single international consensus, experts would have to reach agreement on the cardiovascular risks of competitive athletics and the recommended procedures and personnel involved in screening for possible disease, and adopt similar policies restricting participation in those at risk. Perhaps this vision is too ambitious, and we can only hope that physicians and others can learn from the Italian "experiment." But don't our athletes deserve more than this?

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