Protecting Athletes From Sudden Cardiac Death

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THE CARDIOVASCULAR BENEFITS OF REGULAR PHYSICAL activity have been well documented,[1] but vigorous exercise can also transiently increase the risk of sudden cardiac death.[2-4] Exercise-related sudden death in adults is primarily due to coronary artery disease,[2,5] whereas such events in younger individuals are due to a variety of congenital and genetic cardiovascular disorders, including inherited cardiomyopathies and arrhythmias, anomalies of the coronary arteries or to acquired cardiomyopathy.[6-8]

By virtue of their regular participation in vigorous physical activity, athletes are potentially vulnerable to exercise-related sudden death. Such deaths among athletes are unexpected, dramatic, and often elicit community calls for preventive measures. Cardiac evaluation of athletes before participation is intuitively attractive to identify athletes at risk. Both the American Heart Association[9] and the Sports Cardiology Study Group of the European Society of Cardiology[10] recommend screening high school and college athletes before athletic participation. Both guidelines recommend a personal and family history as well as a physical examination, but the European guidelines recommend obtaining routine electrocardiograms (ECGs). Both recommendations are based primarily on consensus opinion because there are few, if any, prior large studies of screening protocols that provide mortality data.

In this issue of *JAMA*, Corrado and colleagues[11] report findings supporting the cardiovascular screening of athletes and the European approach of routinely requiring ECGs. Italy has had a national mandated preparticipation screening program for athletes since 1982. The present study provides incidence data for sudden cardiac deaths in Italian athletes and nonathletes aged 12 to 35 years in the Veneto region of Italy before and during this screening program. The annual incidence of sudden cardiac death in athletes decreased from 3.6 deaths per 100 000 person-years (1 death per year per 27 777 athletes) in 1979-1980 to 0.4 deaths per 100 000 person-years (1 death per year per 250 000 athletes) in 2003-2004, an 89% reduction. There was no change in deaths among nonathletes, suggesting that this reduction was not due to changes in the population death rate. Most of the decrease in death was due to fewer deaths attributable to cardiomyopathies, whereas the number of athletes disqualified because of cardiomyopathies increased. Both the decrease in deaths and the increase in disqualifications were primarily due to changes in the frequency of arrhythmogenic right ventricular cardiomyopathy (ARVC). Of the 42 386 screened athletes, 3914 (9%) required additional cardiovascular testing and 879 (2%) were ultimately prohibited from athletic participation.

Although these results are provocative, they do not definitively prove the value of screening or establish the importance of routine ECGs in the screening process. First, this study was not a controlled comparison of the screening vs nonscreening of athletes, but rather is a population-based observational study. Other concurrent changes in treating the athletic population over time are possible and could have contributed to the reduction in events. For example, the term ARVC was proposed in 1977 and recognized as a cause of exercise sudden death in the 1980s.[12] Physicians newly aware of ARVC but not involved in the screening process could have detected ARVC at an earlier age among affected family members, and thereby contributed to the decline in deaths from this disease. It is also possible that an increased awareness of exercise-related sudden death, prompted by the Italian screening requirement or by emerging recognition of the problem,[5,13,14] prompted physicians not involved in the screening process to restrict athletes or to more carefully evaluate symptomatic athletes or those with a strong family history.

Second, this study did not evaluate the routine use of ECGs compared with more limited screening based on history and physical examination. The authors attribute their success to the routine use of ECGs, but this component was not examined separately and directly, making it impossible to determine if the ECG added to the other components of the examination.

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Third, there are differences in disease prevalence between Italy and other countries. Hypertrophic cardiomyopathy is the predominant cause of exercise-related sudden death in the United States,[6,7] whereas ARVC is the predominant cause of exertion-related sudden death in Italy.[15] In addition, the screening of Italian athletes is performed by specifically trained physicians. Screening performed by other clinicians who may be less well-trained in the process may miss subtle ECG changes in at-risk individuals, such as those with ARVC and long QT, or may increase the rate of false-positive results and unnecessary disqualifications.

Fourth, the annual death rate prior to the initiation of the mandatory screening program was 1 per year for 27 000 athletes, which is high compared with other studies.[7] The death rate did decrease progressively over time, but this initial death rate accounts for much of the reduction over the course of the study.

And fifth, the lowest annual death rate achieved with screening was 0.4 deaths per 100 000 person-years. This rate is similar to the 0.44 sudden deaths per 100 000 person-years reported for high school and college athletes in the United States between 1983 and 1993.[7] This rate is based on information from the National Center for Catastrophic Sports Injury Research and is the best data available for nontraumatic deaths in US athletes. These participation rates were estimated and noncardiac deaths were included, making the risk estimates less certain than the Italian study; however, this latter limitation probably would have, if anything, overestimated the sudden cardiac death risk in the United States. Nevertheless, such results suggest that the less formal screening process practiced in the United States at that time may have been as effective as the more formal Italian program.

The public[16] and many physicians are enamored with screening, and to many individuals the concept that any well-intentioned screening program could actually have risk is an anathema. This enthusiasm is often encouraged by organizations and companies who sell the screening equipment or the materials to treat detected "disease" and who may overestimate the benefits and minimize the risks and costs of screening. In the study by Corrado et al,[11] none of the cases of sudden death in the nonathlete group reportedly were former athletes who had been previously screened and disqualified from competition. This suggests that screening reduced cardiac deaths and did not simply reclassify sudden deaths from the athletic to the nonathletic population. However, the absence of deaths in disqualified athletes also raises the question as to whether the 2% of athletes who were disqualified were truly at risk for an exercise-related cardiac event. It seems unlikely that all of these screened-out athletes immediately adopted a sedentary lifestyle to avoid the risk of sudden death. The observation that 2% of apparently healthy, young athletes have potentially lethal cardiac conditions is surprising and higher than that in screening studies of US high school and college athletes using ECG (0.4%)[17] or echocardiography (0.5%).[18]

Disqualifications for certain conditions are prudent given the present understanding of disease prognosis, but prognosis for most cardiac conditions is based on experience with individuals whose symptoms lead to the diagnosis. Data are not available on asymptomatic individuals detected during cardiac screening, a group whose prognosis may be far different from that in symptomatic individuals. This also raises the possibility that "lifesaving" interventions documented as effective in patients who have symptoms may have a higher risk-benefit ratio in asymptomatic athletes detected by screening alone. However, it should be recognized that applying a system designed to reduce the risk of death as low as possible during competitive sports will probably exclude some athletes who were not truly at increased risk and deny them the opportunity to participate in competitive athletics.[19]

Despite these limitations, Corrado et al have rigorously collected data from over 2 decades to examine the possible benefits of screening athletes. Their results add important data to the debate on the role of screening and its components, and insight into how to improve the screening process. Their findings support a screening process, which should at a minimum follow established guidelines.[9,11] Moreover, as noted previously,[7] women were underrepresented among athletes experiencing sudden death during sports, with only 4 of the 55 deaths occurring in women.[11] Thus, elaborate testing schemes may be more effective in men. In addition, since 10 of the 55 athletes who died had non-ECG symptoms or findings prior to their demise,[7] physicians should carefully evaluate possible cardiac symptoms in athletes. The evaluation of symptomatic athletes may be one of the most effective sudden death prevention strategies.[20]

The study by Corrado et al provides the best evidence to date supporting the preparticipation screening of athletes and provocative evidence for including ECGs in this process. However, cardiologists and other physicians involved in the evaluation of athletes can take a valuable lesson from Corrado et al, and collaborate to develop a rigorous, comprehensive regional or national registry to study the preparticipation screening process prospectively and directly, and to determine how to implement such programs most effectively and how to manage asymptomatic athletes with cardiac abnormalities detected by the screening process.

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Complexity of the Cerebral Palsy Syndromes
Toward a Developmental Neuroscience Approach

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OVER THE PAST 2 DECADES, MAJOR ADVANCES IN OBstetrics, genetics, maternal fetal medicine, neonatology, developmental neurosciences, and reproductive epidemiology[1-12] have resulted in unprecedented low rates of infant mortality. In 2004, the overall US infant mortality rate was 7 per 1000, with 90% survival of children born very prematurely at 28 to 32 weeks of gestation and survival as high as 80% for children born extremely prematurely at 24 to 28 weeks.[13,14] In addition, a new consensus definition of cerebral palsy (CP) has been proposed,[15] advances in neuroimaging[16-20] have allowed for the examination of central nervous system structure, and a gross motor function classification system[21,22] has given neurodevelopmental pediatricians, orthopedic surgeons, and physical therapists a common language for interdisciplinary collaboration.

In 2000, Stanley et al[23] proposed that etiologic research on single factors needed a more comprehensive framework of causal pathways in order to understand the complexity of children with CP syndromes. One population at known risk of CP that has not been systematically examined is children who have survived very preterm or extremely preterm birth. Recent data from the 14-center National Institute of Child Health and Human Development Neonatal Network of children born between 1993 and 1998 found rates of CP of 19% in survivors born at 22 to 26 weeks of gestation with birth weights of less than 1000 g, and 12% for children who survived after delivery at 27 to 32 weeks of gestation and weighed less than 1000 g.[24] Subsequent analysis from this same group examined outcomes for 1016 infants at the threshold of viability.[25] These infants had a mean birth weight of less than 750 g, a gestational age of less than 24 completed weeks, and a 10-minute Apgar score of 3 or less. Furthermore, 75.8% of these infants died. Among the survivors, 30% had a CP syndrome and almost 1 in 2 had cognitive developmental disability as indicated by a Bayley Mental Development Index score of less than 70 in early childhood.

The report by Bax and colleagues[26] in this issue of *JAMA* is a major advance. This multicenter collaboration investigated clinical correlates of CP in a population sample and compared clinical findings with information available from magnetic resonance imaging (MRI). A cross-sectional population involving more than 500 children with CP born between 1996 and 1999 was assembled from 8 major European centers. Four hundred thirty-one children with CP syndromes were clinically assessed using a structured history and a systematic neurodevelopmental evaluation that included topography (diplegia, hemiplegia, and quadriplegia), physiology (spasticity, dyskinesia, dystonia, and ataxia),

See also p 1602.

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