

Features:

Universal ECG Screening: The Advocate's Perspective

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For patients and families at risk for sudden cardiac death (SCD) due to heart rhythm disorders, an abnormal 12-lead electrocardiogram often yields the first critical clues of serious underlying disease. While it is true that some life-threatening cardiac conditions may not be readily identified by ECG alone, the majority of undiagnosed cases can be found with this simple, relatively inexpensive test.

Studies have shown that the ECG will identify abnormalities in up to 95% of patients with hypertrophic cardiomyopathy (HCM), 90% with long QT syndrome (LQTS), and 80% with arrhythmogenic right ventricular dysplasia (ARVD).^{1,2} Most cases of Wolff-Parkinson White (WPW) and Brugada syndrome are also discovered on ECG. Though the prevalence of these potentially lethal cardiac abnormalities is low, the benefits of early diagnosis and preventative treatment are enormous. Given the high sensitivity of the ECG to presage the risk for sudden death, it seems intuitive that every child be tested at some point in their young life.

Figure 1.



A 4-year-old gets her first ECG at a Philadelphia screening program.

Cardiac screening in the young — particularly newborn infants and competitive athletes — is a long-running, often polarizing point of controversy in cardiology. The debate over the feasibility of cardiovascular screening programs in the United States has intensified in response to recent international recommendations to identify those at risk for sudden death.

In 2005, the European Society of Cardiology (ESC) released its consensus statement on *Cardiovascular Pre-participation Screening of Young Competitive Athletes* recommending a 12-lead electrocardiogram in addition to the focused health history and physical exam.³ In 2004, the International Olympic Committee (IOC) directed that cardiovascular screening be included, along with family history and physical exam, for all its athletes.⁴

The EOC and IOC recommendations are well-founded on decades of European and Japanese experience in cardiovascular screening of the young. Starting in 1979, the Italian government's *Medical Protection of Athletic Activities Act* mandated screening for all athletes aged 12–35 wishing to participate in organized team or individual sports. Between 1979 and 2004, the Italians screened 42,386 athletes of the ages between 12–35, and demonstrated that the annual incidence of SCA in athletes decreased by 89%.⁵

Since 1973, Japan has required that school children be screened by ECG in the 1st, 7th, and 10th grades.⁶ The Japanese experience has demonstrated that the ECG is far more sensitive than medical history or physical exam in identifying high-risk cardiac abnormalities such as LQTS, HCM, WPW, and dilated cardiomyopathy.⁷

The largest reported screening study to date in the United States evaluated 5,615 student athletes in Nevada. The ECG demonstrated greater sensitivity to identifying those at risk for serious heart disorders compared to the medical history and physical exam alone, 70% vs. 3%. The specificity of the ECG was 97.4%.⁸

There are approximately 10 million student athletes competing annually in a variety of sports in the United States. The incidence of SCD in this population is estimated to be less than 300 per year, although there is no mandatory reporting system in place to precisely track the number and causes of these events. Studies

suggest that as many as 70–90% of these deaths were due to pre-existing disorders that could have been identified by ECG screening.

In March 2007, the American Heart Association, in its *Recommendations and Considerations Related to Preparticipation Screening for Cardiovascular Abnormalities in Competitive Athletes*, reaffirmed its long-standing opinion that routine ECG screening of competitive athletes is not warranted.⁹ In spite of the current data confirming the benefits of such testing following decades of Japanese and European studies, as well as the state of Nevada's screening program, the AHA position has remained unchanged from its original 1996 statement.

AHA recommends a 12-point evaluation to help reduce sudden cardiac death in young athletes. The process includes eight medical history questions and four physical tests. For example, the young athlete is to report any personal history of chest pain or discomfort during exercise, any unexplained fainting or near-fainting, episodes of excessive and unexplained fatigue with exercise, a heart murmur and/or high blood pressure.

Questions regarding family history look for any recollection of sudden or unexpected death in one or more relatives before age 50 or in any close relative under age 50 with a disabling heart condition. Furthermore, the young athlete is asked to report any known cardiac conditions, such as HCM or dilated cardiomyopathy, LQTS, Marfan syndrome or serious arrhythmias, in family members. The AHA's confidence in this protocol assigns tremendous responsibility to both the parent and athlete to understand the serious nature of these questions and their ability to provide accurate and objective responses.

Per the AHA recommendations, examiners should test for the presence of heart murmur, the quality of femoral pulses to exclude narrowing of the aorta, any obvious physical characteristics of Marfan syndrome, and a brachial blood pressure. Parents are expected to participate in completing their children's 12-point screening questionnaire. If any of these elements has a "yes" answer, the child should be referred for additional cardiac evaluation.

In a perfect world, a system would exist to guarantee testing compliance and require standardized skills and qualifications for all authorized health examiners. However, current testing is conducted by a wide range of physician and non-physician examiners with varying degrees of training and experience. Referring an athlete for additional testing is at the discretion of the examiner and dependent on their expertise to differentiate between benign conditions and more serious indicators of potentially lethal cardiac abnormalities. This concession at the outset challenges the validity of the AHA panel's recommended guidelines and does not engender confidence that the 12-point evaluation will effectively reduce the incidence of sudden cardiac death in student athletes.

While the scientific community continues to debate points for and against universal screening, patient advocates, parent groups, schools and healthcare professionals have taken it upon themselves to conduct grass-roots screening programs in hundreds of locales around the United States. Surveying dozens of these community-based programs, we found a range of testing methods, including 12-lead ECG with or without 2D echocardiography, stand-alone echocardiograms, medical and family history, brachial blood pressure, and cholesterol and blood glucose checks. The authors of the AHA recommendations appreciate the benevolent intentions of these volunteer efforts and grant that there may be incremental benefits to these initiatives. However, they suggest that these non-standardized programs may create medical liability concerns and are neither financially or technically sustainable on a regional or national scale.⁹

Though large-scale cardiovascular screening programs with 12-lead ECG have been successfully instituted in other countries, the AHA panel feels that similar

practices would be impractical and costly to implement in the United States⁹ specific to the following concerns:

- **Cost-effectiveness:** the panel estimates a total annual program cost of \$2 billion for 10 million athletes. Per athlete costs include \$25 for history and physical exam, and \$50 for an ECG. Additional testing for positive results on history, physical exam or ECG (~15%/1.5 million) is estimated at \$500 per athlete. Program administration costs could be as much as \$500 million.⁹

The AHA's proposed 12-point evaluation has a minimum cost of \$25 per history and physical exam. With or without an ECG, as many as 15% of those examined will require additional testing at \$500 per athlete. The basic premise of the 12-point evaluation is to identify those who may be at risk for sudden death, and the added cost of further study should not be viewed as unwarranted. Administering the 12-point evaluation program has minimum fixed costs as well. Therefore, using the raw figures suggested by the panel, the addition of ECG screening to an already existing cost structure would not be \$2 billion, but more likely in a range of \$500–\$750 million. It should be noted that the panel's financial analysis does not take into consideration the opportunity for cost reductions based on economies of scale in a large, nationalized program.

- **Lack of existing infrastructure:** the panel suggests that the addition of ECG to the existing sports physical exam would require the creation of a new system with significant resources to obtain these recordings. Furthermore, new legislation would be necessary to enforce compliance and set standards for disqualification. A program of this magnitude would require more physician examiners qualified to acquire and interpret ECG results and understand when further testing is warranted.⁹

The current system in place for physical and history exams could be expanded to include acquisition of ECGs. There is a clear opportunity for industry to improve the acuity of computerized interpretive algorithms with more age-specific criteria. Though not an immediate solution, considering a centralized analysis and storage system modeled on the Federal Aviation Administration's ECG database may provide a solution for acquisition and interpretation of recordings from areas lacking manpower. Readings and information directed to a central database could be available for study to increase efficiency and effectiveness of the ECG screening process.

- **Medical-legal implications of false negative results:** concern stems from reality that the ECG is not 100% predictive of all conditions that may cause sudden cardiac death. However, as mentioned previously, the ECG is far more sensitive at identifying those at risk than medical history and physical exam alone. Examining physicians may be concerned about potential litigation if they clear an athlete who later manifests symptoms or suffers sudden cardiac death.

It is unrealistic to expect 100% sensitivity from any diagnostic test. The high sensitivity of the ECG to diagnose most cardiac conditions has been demonstrated, and its utility should not be dismissed. Examiners have a duty to inform their patients of the risks and benefits of any testing they plan to perform, and the patient has the responsibility to consent or decline testing based on this information. The medical informed consent process is well-defined and should be applied to the preparticipation exam just as it is to other testing and therapeutic options.

- **Impact of false positive results:** the panelists are primarily concerned with the financial burden of additional testing that may in the end reveal the good news that no cardiac abnormalities are present. They also express concern for the patient and

their family who may be overwhelmed by the possibility of a life-threatening disorder that could unfairly restrict their participation in organized sports. Of particular note is the burden this additional testing might place on those with low socioeconomic status.⁹

Recent Italian data indicates that the incidence of false positives is far less than reported by the panel.¹⁰ Regarding the emotional impact of an overly conservative result that may or may not indicate a problem on advanced testing, most patients and parents surveyed would rather accept the temporary inconvenience and psychological strain than be forced to cope with the tragic consequences of a missed diagnosis, which could result in death or disability.

To answer the real economic problems faced by those without the financial means to seek additional testing and treatment, there are already federal and state programs in place to screen and provide therapy for low-income children from birth to age 21.¹¹ Not only are these existing programs available to assist those in true need, but the models of these programs may also provide insights on how we might approach funding and implementation to the broader population of all school-aged children.

It is important to also mention that screening efforts focused primarily on student athletes miss the other 25 million U.S. school children who do not participate in organized sports. It is estimated that sudden cardiac death takes the lives of 7,000 children and adolescents each year in this country. Because the majority of these deaths typically occur outside public venues and there is no central registry for these events, the actual numbers are difficult to determine. Many experts believe that the number of deaths due to sudden cardiac arrest in children, adolescents and young adults is vastly underestimated.

As compassionate citizens, we are horrified when avoidable mass casualties take innocent lives, and we vow to commit whatever resources are necessary to prevent similar tragedies in the future. Why then would we not demand a program to prevent the unnecessary deaths of thousands of children and young adults each year due to undiagnosed heart disorders? Are these events any less shocking because they occur incrementally and often privately without major media coverage?

Reliable diagnostic tools and effective treatment options exist to prevent SCD due to undiagnosed heart abnormalities. However, some experts believe it is more cost-effective to reserve advanced testing for only those with a strong family history or who have exhibited obvious symptoms of a possible heart disorder. Considering that cardiac arrest is the sentinel event in more than 55% of sudden deaths, does it make sense to wait and see who manifests symptoms sufficient to justify additional testing?¹² Furthermore, the genetic basis for many rhythm disorders multiplies the risk of SCD to as many as 50% of the index patient's family.

Patient organizations like the C.A.R.E. Foundation hear countless stories of children and young adults dying suddenly, with no small number of these deaths occurring even after prescient episodes of syncope, chest pain or shortness of breath were unfortunately attributed to more benign conditions such as anxiety, growing pains or asthma. Sadly, there are also far too many missed opportunities to prevent sudden death in young athletes who had in fact affirmed episodes of shortness of breath, dizziness or chest pain on their pre-participation questionnaire, though further cardiac evaluations were not performed. Admittedly, these reports are anecdotal, lacking scientific validation to support evidence-based medicine. Nonetheless, one at a time these deaths add up to significant numbers and are devastating to those who have lost loved ones for no fault other than they were unaware that their symptoms might be warning of a serious underlying heart problem.

Advocates for universal ECG screening to prevent sudden death in the young appreciate the enormous challenges to be faced in implementing such a program in the United States. We also believe that the time is right to build on lessons learned by our European and Japanese colleagues to assess what qualities can be emulated in a U.S. program and where inconsistencies require additional investigation. To that end, a next logical step would be to begin screening programs while simultaneously initiating a prospective nationwide ECG screening study to better understand issues regarding methods, costs, effectiveness, and infrastructure requirements.

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