SCREENING ATHLETES FOR HEART DISEASE

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The goals of evaluating individuals for exercise and sports participation are shown in table 1. The most important reason to screen for heart disease is to prevent sudden, unexpected death. Heart disease may also lead to sudden incapacity which may result in injuries, and pre-existing heart disease may be exacerbated by exercise. Also, almost all states in the USA require some type of pre-participation screening of participants in organised sports. Since there are more athletes at educational institutions than in amateur organisations and professional organisations, most physicians encounter this issue in association with high school and university sports. It has been estimated that there are 5 million active athletes at the high school, university, professional and master's levels in the United States.¹ The sudden death rate among high school athletes is 1:100–200 000; among marathon runners 1:50 000; and among recreational joggers 1:15 000.² Thus, athletic sudden cardiac death is a rare event.

One could argue that, given this low frequency of sudden athletic death, no screening programme could possibly be cost effective. The idea of preventing such a low frequency event is challenging and the efficacy of any screening programme would be difficult to prove. Despite these clear realities, public interest in this problem is high. The reason for this was succinctly summarised by Maron and Zipes in the 36th Bethesda Conference Report: “Sudden cardiac deaths in competitive athletes continue to be highly visible, compelling emotional events with significant liability concerns. These catastrophes are frequently subjected to intense public scrutiny largely because of their occurrence in young otherwise healthy-appearing individuals, including elite participants in collegiate and professional sports”.³ Consequently, the purpose of this article is to acquaint the physician with the issue of screening athletes for heart disease and the current recommendations for this activity.

CAUSES OF SUDDEN DEATH IN ATHLETES

In athletes above the age of 35, especially men, the most common cause of sudden death is atherosclerotic coronary artery disease. In younger individuals atherosclerotic coronary artery disease is much less common and other diseases predominate (table 2). The most common cause of sudden death in young athletes is hypertrophic cardiomyopathy. The second most common is coronary artery anomalies, and the third is abnormal left ventricular hypertrophy.² Other diseases fall at 5% or less of cases of sudden death. Some diseases are more prevalent in certain groups of individuals such as the higher frequency of arrhythmogenic right ventricular cardiomyopathy found in northern Italy. Although most athletic sudden death episodes are caused by cardiovascular disease, it must be remembered that there are other causes of sudden death such as commotio cordis, asthma, heat stroke, drug abuse and trauma that causes structural cardiac injury. Commotio cordis is a unique cause of sudden death where a projectile such as a hockey puck or baseball hits the anterior chest with enough force to induce ventricular tachyarrhythmias, but not cause cardiac contusion.⁴ This has to be distinguished from more severe chest trauma, which causes actual structural cardiac injury such as the sudden deceleration of a skier hitting a tree. One can appreciate that there are a number of unusual congenital and acquired heart diseases of children that could make them susceptible to sudden cardiac death. Any screening programme to accommodate such diverse diagnoses is challenging. Thus, most screening programmes have focused on the most common entities such as hypertrophic cardiomyopathy and arrhythmogenic right ventricular cardiomyopathy in Italy. Also, athletic sudden death is 5–9 times more common in men than women, yet screening programmes usually are not different for the two sexes.²

DIFFERENTIATING CARDIAC DISEASE FROM ATHLETE'S HEART

Physiologic adaptation to exercise training results in cardiac structural and functional alterations that can be confused with heart disease.⁵ Dynamic exercise training leads to increases in cardiac chamber

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Table 1 Goals of pre-participation screening of athletes

- ▶ Prevent sudden unexpected death
- ▶ Prevent injuries
- ▶ Prevent disease exacerbations
- ▶ Meet legal and insurance requirements

sizes and left ventricular mass. Static exercise training increases left ventricular wall thickness and left ventricular mass. These changes in cavity size and hypertrophy can alter the ECG in ways that resemble pathologic chamber enlargement and may resemble patterns seen in myocardial inflammation or infarction. Dynamic exercise training also reduces resting heart rate and may lead to excessive vagal tone at rest, resulting in abnormalities such as atrioventricular block and wandering atrial pacemaker. In addition bradycardia may increase the frequency of premature ventricular contractions and short runs of ventricular tachycardia. All of these rhythm disturbances normally disappear with exercise in the trained athlete.⁶

The degree of chamber enlargement and hypertrophy is roughly correlated to the level of aerobic training as represented by the maximum oxygen consumption ($\dot{M}\dot{V}O_2$) at peak exercise. Thus, athletes with the highest $\dot{M}\dot{V}O_2$ such as bicycle racers and long distance runners have very large hearts. By contrast athletes with little aerobic exercise training, such as golfers, have hearts that are in the normal range of size. Athletes with considerable static exercise training such as weight lifters often have thick left ventricular myocardial walls which may exceed the upper limits of normal. One study recorded wall thickness up to 16 mm in highly trained athletes.⁷

Highly trained athletes with cardiac chamber enlargement may have abnormal physical findings such as an enlarged apical impulse, systolic flow murmurs, and third and fourth heart sounds. Such findings increase the challenge of distinguishing cardiac adaptations to exercise from those of heart disease. The ECG may help distinguish cardiac disease from exercise training adaptations, but there are considerable false positives that have to be dealt with. Pelliccia and colleagues, who evaluated over 1000 athletes by ECG, showed that 17% of men and 8% of women had distinctly abnormal ECGs, and a further 28% of men and 14% of women had mildly abnormal ECGs.⁸ When those with distinctly abnormal ECGs were evaluated further it was found that the majority of them were in the high aerobic conditioning sports such as cycling as

Table 2 Causes of sudden cardiac deaths in young athletes

46%	Hypertrophic cardiomyopathy and unexplained left ventricular hypertrophy
19%	Coronary artery anomalies
≤5%	Myocarditis
	Ruptured aortic aneurysm (Marfan syndrome)
	Arrhythmogenic right ventricular cardiomyopathy
	Tunnelled (bridged) coronary artery
	Aortic valve stenosis
	Atherosclerotic coronary artery disease
	Dilated cardiomyopathy
	Mitral valve prolapse
<1%	Long QT syndromes

compared to lower aerobically trained athletes such as equestrians, 89% of whom had normal ECGs. Among the 145 patients with distinctly abnormal ECGs, 14 were found to have cardiovascular disease. Among the other 131, two thirds had enlarged cavities or thickened ventricular walls by echocardiogram and the other third were completely normal on echocardiogram. Thus, most athletes with ECG abnormalities do not have heart disease.

RECOMMENDATIONS FOR SCREENING

The most important time to screen athletes is pre-participation.⁹ Accordingly, potential medical problems that would preclude athletic activity or that need further evaluation can be identified before the individual starts participating in sporting activities. The American Heart Association recommends that high school and college athletes have a pre-participation examination and that it be repeated every two years.¹ A new medical history only is recommended in the intervening years. Many schools fully evaluate their athletes yearly, but this frequency of pre-participation examinations was felt to be a potential hardship on less affluent schools. The American Heart Association recommends a 12 point screening procedure that is outlined in table 3. As can be seen, eight of the 12 points are related to the history and the remaining four are physical examination elements. These points should be viewed as minimal criteria and a complete physical examination should be encouraged when feasible. For example, palpation of the carotid pulses and precordium are not included in the 12 points, but they may provide important information. Also, a history of any medication or recreational drug use should be sought. Many physicians use the occasion of a pre-participation evaluation to address adolescent health issues in general.

The American Heart Association did not recommend other testing on a routine basis, but rather that other testing be done on an individual basis when there are unexplained abnormal findings or suspicion of cardiovascular disease.¹ The European Society of Cardiology has taken a different approach and specified that an ECG should be done with each evaluation.¹⁰ They believe that this will detect most cases of hypertrophic cardiomyopathy and the majority of cases of arrhythmogenic right ventricular cardiomyopathy. Although the American

Table 3 American Heart Association 12 point screening procedure

Personal medical history

1. Exertional chest pain/discomfort
2. Syncope/near syncope
3. Excessive unexplained exertional dyspnoea or fatigue
4. Prior recognition of a heart murmur
5. Elevated blood pressure

Family medical history

6. Premature death related to cardiovascular disease
7. Disability from cardiovascular disease at age <50 years
8. Hypertrophic cardiomyopathy, dilated cardiomyopathy, Marfan syndrome, arrhythmias, channelopathy (eg, long QT)

Physical examination

9. Heart murmur
10. Femoral vs radial pulses to exclude aortic coarctation
11. Physical stigmata of Marfan syndrome
12. Brachial artery blood pressure

Heart Association did not disagree with this assertion, they believe that the cost of doing ECGs versus the yield is prohibitive and that the cost of evaluating false positives, both in terms of financial cost and psychological impairment, are too great to make this practice cost effective.¹ Currently there are no comparative data using the two approaches and there are little data to support that either approach significantly reduces the risk of sudden athletic death. No matter what screening procedure is used it is impossible to achieve zero risk in sports.

If cardiovascular or other abnormalities are found when evaluating an individual, an estimation should be made of how much physical exercise can be safely tolerated. This requires knowledge of the type of exercise the individual will be doing; how much static and dynamic exertion is required; and how vigorous the training programme is. In some sports training is often more vigorous than the actual competition—for example, boxing. The 36th Bethesda Conference report has a chart of all major sporting activities and what level of dynamic and static exercise are generally required to participate in the sports.³ The report also discusses many cardiovascular diseases, including congenital heart diseases, and details what levels of exertion are acceptable at various severities of the disease. Combining this information will allow for a decision as to whether an individual patient can safely participate in a particular sport or not. For example, athletes with mild aortic stenosis can participate in all competitive sports, but moderate aortic stenosis would restrict an athlete to low to moderate static and dynamic sports such as diving or volleyball.

The primary obligation of a physician to the athlete is their best medical interest, but the physician must avoid unnecessary exclusion from sports. Thus, when abnormalities are detected that may disqualify a person from the sport they are interested in, it behoves the general physician to request specialty consultation or testing. Also, the athlete should be temporarily withdrawn from activities until the issue can be resolved. If the general physician and the specialist both agree that the patient's condition requires disqualification, then they should not hesitate to disqualify the individual from participation. Such decisions, if based on a reasonable pre-participation evaluation following the usual and customary medical practices of the region, have generally been upheld in court cases. Also, there seems to be little liability risk if an asymptomatic condition is missed.⁸

The physician should resist pressure from competing interests such as the athlete, the family, the coach, the administrative officials of the educational institution, and the alumni. Such individuals may be interested in having the individual compete athletically for reasons that are not in the individual's best interest medically. Once a decision has been made, the physician should report only to the patient, or the patient's parents if the patient is a minor, the referring doctor, and in some cases the institutional officials when an institution is paying for the medical evaluation. It is unwise to make any public pronouncements or discuss cases with the press unless the above-mentioned people all wish for this to be done.

Evaluating the older athlete who wants to embark on a fitness programme or participate in competitive master's level sports is a different challenge. Here one must weigh the health benefits of exercise versus the risk of triggering a cardiovascular event in those with unsuspected cardiovascular disease.¹¹

Regular exercise decreases the overall risk of sudden death, but if sudden death occurs, it is more likely to occur during exercise. The focus in these athletes is coronary artery disease risk. They should be evaluated for coronary artery disease risk factors and if one risk factor beyond male sex is present an exercise test or some other screening procedure for coronary artery disease would seem warranted. Also an exercise test would determine the individual's overall fitness for athletic participation. If a patient has known coronary artery disease, even if they have been revascularised, high intensity competitive sports should not be permitted. Other contraindications to high intensity competitive sports are listed in table 4.

SCREENING CONTROVERSIES

Due to the potential costs involved, many institutions and government bodies have permitted or encouraged non-physicians to do the screening of athletes.¹² Consequently, nurse practitioners, physician's assistants, nurses and trainers are doing the pre-participation evaluation in many institutions. Some institutions or states have a form that must be filled out on each athlete. There is no generally agreed upon form, and many are incomplete with regards to history or physical exam elements that may identify diseases known to cause sudden death. The Italian Sports Authority requires that a certified sports medicine physician do the evaluations.¹¹ The American Heart Association has taken a more inclusive approach and recognises that other well trained health professionals can do this actively.¹ The concern is how well trained some of these non-physicians are. How accurately could a health technician pick out someone with Marfan syndrome from the rest of the basketball players? Also, trainers and nurses employed full-time by the institution may not be as objective about disqualifying a star player. The Italian approach is close to ideal, but the cost of such a high level evaluation for preventing such rare events would be difficult to sell in other countries.

How much testing should be required is controversial. The Italian Sports Authority requires an ECG.¹⁰ This may not be much of an added cost in their system, but it would be in others. The higher prevalence of arrhythmogenic right ventricular cardiomyopathy in Italy, which almost always is associated with an abnormal ECG, and their interest in other conditions such as Brugada's syndrome, may make this practice cost effective. Also, Corrado and colleagues have shown that among 33 735 athletes screened in Italy, 22 were ultimately found to have hypertrophic cardiomyopathy by echocardiography (0.07%).¹³ Of these 22, 16 had an abnormal ECG, only two had a heart murmur on examination, and only three had a

Table 4 Contraindications to high intensity competitive sports

- ▶ Left ventricular ejection fraction <50%
- ▶ Exercise tachyarrhythmias
- ▶ Exercise hypotension
- ▶ Exercise ischaemia
- ▶ Syncope
- ▶ Moderately severe or more valvular disease
- ▶ Prior embolic event

family history of hypertrophic cardiomyopathy. Since not everyone had an echocardiogram, we do not know how many hypertrophic cardiomyopathies were missed and whether this approach prevented any sudden deaths. Nevertheless, it suggests that the ECG may be far superior to history and physical examination for detecting hypertrophic cardiomyopathy.

This raises the issue of echocardiographic screening. Some have recommended a limited echocardiogram to assess for left ventricular hypertrophy, right ventricular enlargement and an enlarged aortic root, perhaps using one view such as a parasternal long axis view.^{14 15} Proponents claim this could be done at low cost by trained technicians who would refer any abnormal cases for further evaluation or a more complete echocardiogram. This would detect many with abnormalities, but it still would not cover some causes of sudden death such as a tunnelled coronary artery. Even a limited echocardiogram would have some cost, which could be prohibitive. Also, the cost effectiveness of this approach has not been proven. At this time no major sports or medical organisation recommends routine echocardiography.

THE VALUE OF PRE-PARTICIPATION SCREENING

There are few data on the value of pre-participation screening, especially with regard to the prevention of sudden death, and the data that do exist are not particularly encouraging. For example, a retrospective study of 134 high school and college athletes, who died suddenly of cardiovascular causes, evaluated the results of their pre-participation evaluation.² Four of them declined an evaluation, and 115 underwent history and physical examination. Of these, four were suspected of having some sort of cardiovascular abnormality which was correctly identified in only one case. All four were cleared to play despite the abnormality finding. This study suggests that pre-participation screening in the USA is not sensitive for discovering important cardiovascular disease in young athletes (sensitivity 3%).

Experience with screening in Italy of more than 33 000 athletes over seven years resulted in the identification of 22 (0.07%) with hypertrophic cardiomyopathy using their protocol in which an ECG is done in all individuals.¹³ This frequency is similar to that observed in a US study, using echocardiographic screening in all individuals, of 0.1%. These data suggest that the Italian screening programme, which uses licensed sports medicine physicians and ECG, performs as well as echocardiography based screening for detecting hypertrophic cardiomyopathy. Also, in the same Italian study there were 269 juvenile sudden deaths, of which 49 died during athletic events and had undergone pre-participation screening.¹³ Of these 49, only one had hypertrophic cardiomyopathy (2%) which is much lower than the 7.3% incidence in the 220 non-athletes. This suggests that screening in Italy does decrease athletic deaths caused by hypertrophic cardiomyopathy. The most common causes of juvenile sudden death were arrhythmogenic right ventricular cardiomyopathy, atherosclerotic coronary artery disease, and anomalous origin of a coronary artery; none of which were reduced in frequency in the athletes. This indicates that screening, even with ECG, is not particularly effective for identifying subjects with these conditions.

Screening has many problems.¹⁴ The history and physical examination have a low sensitivity for detecting problems that

Screening athletes for heart disease: key points

- ▶ Pre-participation athlete screening is mainly targeted at preventing tragic sudden deaths in young athletes
- ▶ Athlete sudden deaths are usually caused by unrecognised cardiovascular disease
- ▶ Cardiovascular disease can be difficult to distinguish from the compensatory adaptations of the heart and circulation to athletic training
- ▶ Although it is difficult to prove the efficacy of screening, it is recommended by major health and sports organisations
- ▶ The extent of screening procedures and tests should be decided based upon each individual or in mass screenings the most frequent causes of sudden death in the population

could lead to sudden cardiac death. For example, ventricular arrhythmias, premature coronary artery disease, and coronary artery anomalies usually have no physical findings. Although sensitivity can be increased with non-invasive testing, it will never be 100%. Diseases such as coronary artery anomalies are very difficult to detect, even with non-invasive imaging. The specificity of the ECG is poor. The ability to differentiate the athlete's heart from disease by echocardiography is often challenging. Even at autopsy, the cause of death is not always clear. In a study of non-traumatic military recruit deaths that were studied pathologically, 51% had cardiovascular disease, but in 35% the cause of death was unexplained. The major problem in screening is that there is a low prevalence of disease in the athletic population, so many of those suspected of having disease are false positives. Also, there is a low risk of sudden death even in athletes with disease.

RECOMMENDATIONS FOR SCREENING

Ideally screening should be done pre-participation by a physician trained in this activity, but other well-trained healthcare workers are acceptable as long as they do not have a conflict of interest. Screening should be done yearly just before training for the sports activity begins. Every two years is acceptable, with a new health history in the intervening years. The evaluation should include a history of symptoms suggestive of heart disease, a family history of premature death or specific cardiac diseases, and questions about substance abuse. The physical examination should include blood pressure, femoral pulses, auscultation of the heart in the standing position and inspection for Marfan syndrome features. There should be a low threshold for delaying clearance to play to evaluate further suspected cardiovascular disease. Further evaluation may include ECG, echocardiography and referral to a cardiologist. Only after this evaluation is complete should an athlete be cleared to participate. In some situations restrictions will need to be advised or alternative sports activities that are safer for the individual recommended. If the physicians involved in this process follow guidelines from major medical and sports organisations and treat the athlete in the usual and customary way, legal risk should be minimal.

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