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Non-Traumatic Sudden Death In Sports

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INTRODUCTION

An athlete can be defined as someone who participates in competitive individual or team sports. Athletes are commonly perceived as being very healthy. Nevertheless, on rare occasions, athletes may die suddenly and unexpectedly, mainly from a cardiac cause. The overwhelming cause of sudden death (SD) is due to cardiac reasons, and this is called sudden cardiac death (SCD). SCD has been defined as a natural death due to a cardiac cause, occurring either instantaneously or up to 1 hour after the onset of symptoms in someone who may or may not have known heart disease, but in whom the death occurs unexpectedly.

EPIDEMIOLOGY

The prevalence of SD in athletes is low. It is estimated to be about 1 in 200,000 in high school aged athletes per year, although it is thought that this is an under-estimate. About 90% of these deaths occur in men. This could be because more men participate in sports, and also in higher risk sports. Also, men have a higher risk of cardiovascular (CV) disease.

PHYSIOLOGICAL EFFECTS OF EXERCISE

Depending on natural ability and training, strenuous dynamic exercise increases cardiac output by 4 to 6 times from baseline. However, generally, there is minimal change in mean blood pressure (BP) because while systolic BP rises, vascular resistance falls. On the other hand, static isometric exercise increases mean BP. Still, both types of exercises increase myocardial oxygen needs. If there is any abnormality of blood supply or ventricular structure, this may cause relative ischaemia during exercise, leading to fatal arrhythmia. Exercise also increases sympathetic discharge, which affects heart electrophysiology.

Regular exercise causes normal adaptive structural and electrophysiological changes in the heart. The changes are sometimes referred to as 'athlete's heart'. Structural changes include an increase in left ventricular (LV) end-diastolic end volume, increased LV wall thickness, increased LV mass, and increased ventricular cavity diameters. Electrophysiological changes include sinus bradycardia, high QRS voltages, and early re-polarisation.

CAUSES OF SUDDEN CARDIAC DEATH IN ATHLETES

In young (<35 years old) competitive athletes who die suddenly, the majority have some inherited functional or structural CV lesion, which predisposes to fatal arrhythmia. US studies indicate that the most common lesion in this group is hypertrophic cardiomyopathy (HCM). Other lesions include congenital coronary artery anomalies, idiopathic LV hypertrophy, arrhythmogenic right ventricular dysplasia (ARVD), myocarditis, Marfan syndrome with aortic dissection and rupture, valvular

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disease (mitral valve prolapse or MVP, aortic stenosis), electrophysiological disorders (Wolff-Parkinson-White syndrome or WPW, long QT syndrome or LQTS), drug use (cocaine or anabolic steroids), and atherosclerotic coronary artery disease (CAD). In the older age group, CAD is the major lesion.

HCM – This is characterised by inappropriate asymmetrical LV hypertrophy (LVH) without any obvious cause. The prevalence is about 0.2% in those aged 18 to 30 years (US studies). The majority of cases are inherited as an autosomal dominant trait, with over 100 gene mutations for sarcomere proteins having been identified. Familial studies suggest that polymorphism, other gene mutations and environmental factors play a role. Most persons with HCM have no symptoms and unfortunately, SCD is the first presentation. There are usually ECG changes, but up to 15% of cases can have normal ECG. Echocardiography is the mainstay for diagnosis. It may be difficult to distinguish HCM from athlete's heart in borderline LVH, and genetic studies may be needed. Persons with HCM have increased likelihood for exercise related SCD, but the relative risk is unknown, because there appears to be other potential mechanisms acting which cause SCD. The extent of myocardial disarray appears to be the most important determinant of electrical instability. Risk factors with greater disarray include younger age, family history of SCD, known gene mutations associated with increased SCD prevalence, previously aborted SCD, sustained ventricular or supra-ventricular tachycardias, recurrent syncope and non-sustained ventricular tachycardia during Holter monitoring and bradyarrhythmias. It has been suggested by some expert groups that athletes with unequivocal HCM should be disqualified from competitive sports.

IDIOPATHIC LVH – Some young athletes who die suddenly have non-dilated, mildly enlarged hearts showing symmetrical LVH with no disarray and no known genetic abnormalities. It is possible that this lesion is a HCM variant or due to undetected severe hypertension.

CONGENITAL CORONARY ARTERY ABNORMALITIES – This is thought to be the second most common cause of SCD in young athletes. In this circumstance, it is usually an isolated abnormality, rather than part of a complex malformation. The most common abnormality found in relation to SCD is that of the left main artery, arising from the right sinus, resulting in high take-off of the artery from the ascending aorta. Exercise stress testing usually does not induce ischaemia, and coronary angiography is required to come to a definitive diagnosis, although coronary magnetic resonance angiogram is an alternative investigative technique.

ARVD – This is a cardiomyopathy of unknown origin, characterised by electrical instability due to fibro-fatty infiltration of the RV. The ECG changes of prolongation of the QRS duration (>100 ms in V1-V3) and persistent T inversion in V1-V3 in athletes beyond adolescence are suggestive of ARDV. An epsilon wave that points to late RV activation just after the QRS complex is only seen in about 5-30% of cases.

MYOCARDITIS – Myocarditis refers to inflammation of the heart, usually of an infectious (viral) aetiology. Most cases of myocarditis are self-limiting and are clinically unrecognised because there are no symptoms or the symptoms are trivial. Tachycardia out of proportion to the fever may be present. ECG changes are usually transient. Athletes with a febrile, common cold-like illness should be rested until symptoms abate. If a diagnosis of presumed myocarditis has been made, athletes should not train until further cardiological evaluation has been done. If the diagnosis of myocarditis is confirmed, athletes should not engage in competition for at least 6 months, after which they should undergo cardiological evaluation again.

MARFAN SYNDROME (MS) WITH AORTIC DISSECTION AND RUPTURE – MS is an inherited connective tissue disorder involving a defect in fibrillin-1, an extra-cellular glycoprotein. The main pathological changes in the CV system are MVP (see page 28), and ascending aorta dilatation due to cystic medionecrosis. The dilatation may give rise to aortic incompetence. Weakening of the media of the aorta predisposes to a tear of the intima and aortic dissection. The dissection may extend along the entire aorta, and may rupture, a calamity that can cause SD. Athletes with MS without aortic dilatation should have periodic echocardiographic evaluation. Based on cardiological advice, they might be able to participate in non-contact, low-level intensity competitive sports. Athletes with aortic dilatation should not participate in competitive sports. Depending on the extent of the aortic dilatation,

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persons with MS and aortic dilatation may require prophylactic surgery.

MVP – MVP has been identified as a lesion associated with SCD in athletes. However, the associated risk is not known, and it is not thought to be an important cause of SCD. This is because cases of SCD with identifiable MVP usually do not occur during exercise, and there are often other cardiac disorders present (thus MVP in these cases of SCD could be an incidental finding). Athletes with asymptomatic MVP and who have no evidence of connective tissue disease can probably engage in competitive sports. All other athletes with MVP (e.g. with history of syncope, family history of SCD or mitral regurgitation) should be evaluated.

AORTIC STENOSIS – This accounts for a small proportion of SCD in athletes, probably because the presence of a murmur usually leads to further evaluation and disqualification from competitive sports.

ELECTROPHYSIOLOGIC DISORDERS – Patients with WPW will need evaluation for risk stratification and consideration for catheter ablation. LQTS is a familial disorder, usually transmitted as an autosomal dominant trait. It is caused by mutations to ion channel genes. A female gender, corrected QT >500 ms and a history of syncope are risk factors for SCD. Athletes with LQTS should be prohibited from competitive sports.

NON-PENETRATING IMPACT TO THE CHEST

Non-penetrating impact of the chest without injury to the heart or great vessels can cause SCD in athletes. This is sometimes called commotio cordis or cardiac concussion. It usually occurs when a projectile (e.g. a baseball) hits the chest. It can also occur when a blow is delivered to the chest during martial arts. The velocity of impact is usually not high and death occurs remarkable quickly, sometimes almost instantaneously. The usual rhythm seen when medical attention becomes available is ventricular fibrillation and less commonly, bradyarrhythmia. The mechanism is unknown. Animal experimental studies and clinical studies suggest that the mechanisms include the impact inducing an ectopic beat during re-polarisation, extreme vaso-vagal response, conduction system damage only detectable by special histochemical techniques, coronary vasospasm and K+-ATP channel activation. No proven predictive clinical indicators or preventive measures are known.

NON-CARDIOVASCULAR CAUSES OF SD IN ATHLETES

These include exertional hyperthermia (heatstroke), exertional rhabdomyolysis, status asthmaticus and exercise-induced anaphylaxis.

PRE-PARTICIPATION SCREENING IN COMPETITIVE ATHLETES

CV screening of competitive athletes, especially pre-participation, is important as it may detect otherwise occult CV abnormalities that can lead to SCD during extreme physical exertion. A history of palpitations, exertional chest discomfort, giddiness or syncope would require further evaluation. Detailed family history from the patients or parents of young athletes would be necessary to detect family history of heart disease or SD. Once an abnormality is detected, there is consensus that further referral for a full cardiological evaluation is necessary. However, only a few cardiac abnormalities such as severe HCM and congenital aortic stenosis may be detected clinically.

Whilst there is agreement for a detailed history (especially family history) of heart disease and detailed physical examination to be taken, there is controversy regarding the need for routine ECG screening. Occasionally, athletes with HCM, pre-excitation syndrome, Brugada syndrome, LQTS and short QT syndromes can be detected on the routine ECG. However, other important abnormalities such as coronary artery anomalies and cathecholaminergic polymorphic VT will usually not be detected by routine 12-lead ECG screening alone. CAD, which is the most common cause of SCD in older athletes (>35 years of age) will also usually not be detected by a routine 12-lead ECG.

The substantial number of young athletes who require evaluation with only a very small percentage (estimated to be <0.5%) who are truly abnormal, together with the cost involved have been the major reasons for not implementing routine ECG screening in the US Guidelines.

The European Society of Cardiology consensus statement in 2005, however, recommends including a 12-lead ECG. They argue that it is ethically and clinically justifiable to make every effort to recognise any disease that can put athletes at risk and to reduce fatalities.

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In Singapore, as recommended by the Sports Safety Committee in 2007, selected groups undergo full CV history, physical examination and ECG testing.

Routine exercise stress testing and echocardiograms are currently not indicated, not just because of the cost involved, but also because these tests lead to too many false positive and false negatives in an unselected population. This may then lead to further unnecessary tests which may themselves be associated with other risks (e.g. cardiac CT and radiation exposure).

CONCLUSION

It is probable that the majority of causes of SCD in athletes have been identified. SCD may be prevented by disqualification of high-risk individuals from competitive sports and commencing specific therapy where available. Identification of high-risk individuals is the main rate-limiting step to the implementation of effective preventive measures, due to low prevalence and a lack of practical cost-effective screening methods with adequate sensitivity and specificity. ■

References

1. ***Corrado D, Pelliccia A, Bjørnstad HH, et al. Cardiovascular preparticipation screening of young competitive athletes for prevention of sudden death: proposal for a common European protocol. Eur Heart J 2005; 26:516-524.***
2. ***Maron BJ, Douglas PS, Graham TP, et al. Task Force 1: preparticipation screening and diagnosis of cardiovascular disease in athletes. J Am Coll Cardiol 2005; 45:1322-6.***
3. ***Maron BJ, Poliac AC, Kaplan JA, et al. Blunt impact to the chest leading to sudden death from cardiac arrest during sport activities. N Engl J Med 1995; 333:337-42.***
4. ***Maron BJ, Robert WC, McAllister HA, et al. Sudden death in young athletes. Circulation 1980; 62:218-29.***
5. ***Maron BJ, Shirani J, Poliac LC, et al. Sudden death in young competitive athletes. Clinical, demographic and pathological profiles. JAMA 1996; 276:199-204.***
6. ***Maron BJ, Thompson PD, Ackerman MJ, et al. Recommendations and considerations related to preparticipation screening for cardiovascular abnormalities in competitive athletes: 2007 update. Circulation 2007; 115:1643-1655.***