PERSPECTIVE

Sudden cardiac death in the athlete

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Physiological stress may trigger fatal cardiac events in athletes with an unknown cardiac condition.

Athletes are generally considered to be healthy as well as fit; however, they do have an incidence of sudden death. The GP may be the patient's only previous medical contact, so it is important that warning signs or family history are detected and acted on at the earliest opportunity.

The sudden death of an athlete or sportsperson is usually reported in the media, as in the case of Fabrice Muamba, the soccer player who had a ventricular fibrillation arrest during a televised UK FA cup match in March this year. Following more than an hour of cardiopulmonary resuscitation, Muamba survived to make a good recovery. However, not all patients are so fortunate. The literature suggests a sudden death rate of between 0.35 and 2.3 per 100,000 sporting participants per year, with a marked male preponderance, which may be due to the higher rate of male participation in addition to higher disease expression.

Presentation and the role of the GP

Most cases of sudden death on the sports field are completely unheralded. A minority will have had symptoms and sought medical advice previously, usually from a GP. It is important not to disregard these presentations solely on the grounds that the patient is young and fit. The major features of importance, suggested by the American Heart Association, are listed in the box on page 23.[1] Strenuous training or competitive participation should be ceased on finding a positive risk feature, and the patient should be referred to a specialist. Modern

Key points

- **The presence of exertional symptoms such as syncope, chest pain or palpitations should trigger referral of the patient for specialist assessment.**
- **Apparent good health and exercise capacity provide no reassurance regarding potentially serious inheritable conditions.**
- **Life-saving treatments such as implantable defibrillators, catheter ablations and medications are available and appropriate for people at high or moderate risk of sudden cardiac arrest.**

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treatments, including implantable defibrillators and catheter ablations, are appropriate for some people and can save their lives.

Specific conditions causing sudden cardiac arrest

Cardiac disease accounts for most sudden death on the sports field; associated conditions and other main causes are listed in the boxes on page 24. Cardiac diagnoses can be separated into the morphologically abnormal, with the cause being clearly identified at postmortem, and the morphologically normal, with death being from a presumed arrhythmia. The latter 'primary arrhythmias' are often due to one of several ion-channel defects or 'channelopathies'. These may be detected by genetic testing or ECG abnormalities. A prospective national registry, the Australian Genetic Heart Disease Registry, is aiming to record and characterise all episodes of potential cardiac sudden death and 'molecular autopsy' findings. Physicians, patients or relatives can contact the registry directly through its website (http://www.heartregistry.org.au).

Hypertrophic cardiomyopathy (HCM)

The most common of the genetic heart diseases (prevalence, 1:500) and the most common cause (30%) of sudden death in the athlete is hypertrophic cardiomyopathy. This has an autosomal dominant pattern of inheritance, with sporadic mutations also being common. Variable penetrance leads to a highly variable expression of disease severity. Abnormal thickening of the ventricular myocardium with disorganised muscle fibres leads to risk of malignant ventricular arrhythmias. ECG changes are frequently seen (Figure 1).

Arrhythmogenic right ventricular cardiomyopathy (ARVC)

Arrhythmogenic right ventricular cardiomyopathy is an autosomal dominant condition that results in fibro-fatty replacement of the right ventricle. T-wave changes in the right precordial ECG leads are often seen. Right ventricular dilation or reduced function may be detected on echocardiogram. Right ventricular loading and stretch during exercise may cause worsening of the disease and arrhythmia burden.

Long QT syndrome (LQTS)

Syncope, polymorphic ventricular tachycardia and sudden death are typical features of long QT syndrome. For some subtypes of long QT syndrome, swimming is a frequent arrhythmic trigger. Penetrance is incomplete and the QT interval may be labile, so detection is enhanced by obtaining ECGs while the patient is supine and standing, and during exercise and recovery. β-blockers are usually effective in patients with long QT1.

Catecholaminergic polymorphic ventricular tachycardia (CPVT)

Catecholaminergic polymorphic ventricular tachycardia is characterised by palpitations and syncope due to exercise-related ventricular tachycardia. Symptoms usually develop by early adolescence. Diagnosis is by exercise ECG, which may show a characteristic 'bidirectional ventricular tachycardia' or polymorphic ventricular tachycardia.

Congenital structural heart disease

An abnormally posterior origin of the left main coronary artery has been associated with sudden death on exertion. The artery may have a slit-like lumen and a highly angulated proximal course, and can become 'squeezed' between the aorta and pulmonary artery during episodes of high cardiac output. This can lead to sudden and catastrophic ischaemia and ventricular fibrillation, accounting for a significant proportion of sudden cardiac death in athletes.

Marfan syndrome or congenital bicuspid aortic valve may be associated with a significant abnormality of the aortic wall. Under strenuous exertion, spontaneous dissection of the aortic wall may occur, with a rapidly fatal outcome.

Commotio cordis

A direct nonpenetrating impact to the chest wall, usually from a ball or less frequently a collision with another player, can initiate ventricular fibrillation if critically timed to the T-wave. Prompt defibrillation is required to restore cardiac rhythm.

The role of routine screening

It is generally accepted that athletes with one of the cardiac conditions listed above are at significant risk of a fatal event triggered by

Twelve-point screening for the detection of cardiac risk in the athlete[1]

The presence of any of the following factors in an athlete would indicate a risk of cardiac disease.

Personal medical history

1. Chest pain or discomfort on exertion
2. Syncope/near syncope (especially related to exertion)
3. Excessive and unexplained dyspnoea or fatigue associated with exercise
4. Prior recognition of heart murmur
5. High blood pressure

Family medical history

6. One or more relatives who died (sudden or otherwise) of heart disease before age 50 years
7. A close relative under the age of 50 years with disability from heart disease
8. Specific knowledge of hypertrophic or dilated cardiomyopathy, ion channelopathies such as long QT syndrome, Marfan syndrome or clinically important arrhythmias

Physical examination

9. Heart murmur
10. Femoral pulses to exclude aortic coarctation
11. Physical appearance of Marfan syndrome
12. Brachial artery blood pressure (taken in a sitting position)

Cardiac conditions associated with sudden death in the athlete

Frequent causes

- Hypertrophic cardiomyopathy
- Aberrant coronary artery course

Rarer causes

- Arrhythmogenic right ventricular cardiomyopathy
- Catacholaminergic polymorphic ventricular tachycardia
- Long QT syndrome
- Myocarditis
- Marfan syndrome/aortic dissection/bicuspid aortic valve stenosis

Main causes of death on the sports field

- Asthma
- Brain injury
- Cervical spine injury
- Diabetes
- Exertion heat stroke
- Exertional hyponatraemia
- Exertional sickling
- Head down contact (American football)
- Lightning strike
- Sudden cardiac arrest

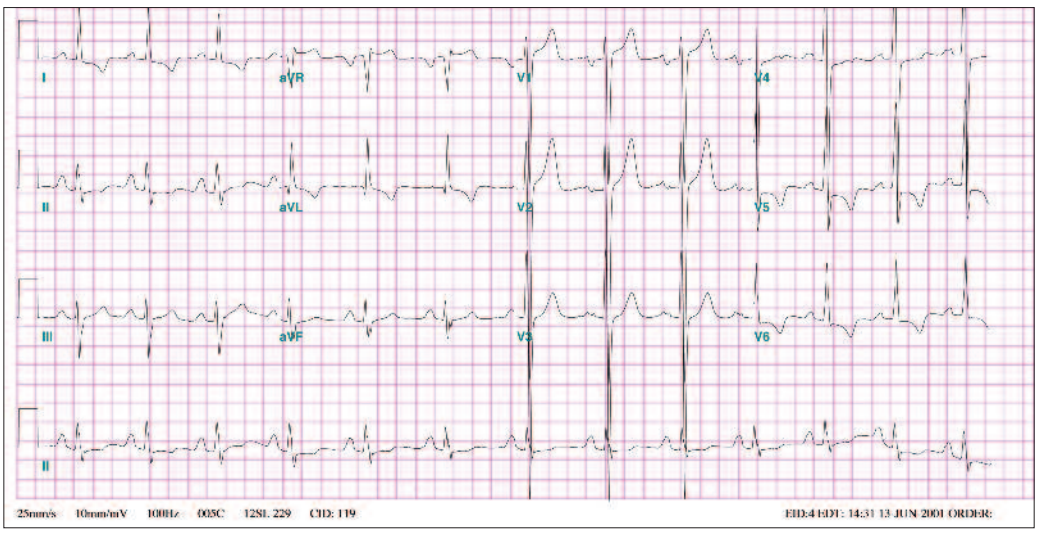

Figure 1. Marked ECG abnormalities such as ST/T-wave changes with left ventricular hypertrophy criteria are seen in 90% of patients with hypertrophic cardiomyopathy by the end of adolescence.

physiological stress and should not participate in high level or competitive sport. For professional athletes, this often has significant financial and lifestyle implications.

Screening, however, is a controversial issue, due to the low sensitivity and specificity of testing. A normal cardiac response to regular exertion (as little as 20 minutes, three times per week) is hypertrophy, which may be confused with hypertrophic cardiomyopathy. ECGs can demonstrate physiological bradycardia with U waves, mimicking the changes of long QT syndrome. Nonetheless, a large screening program in Italy has shown a marked reduction in sporting deaths over a period of 25 years by using a patient's history, a physical examination and a resting 12-lead ECG to assess cardiac risk.[2] A minority of participants needed a more detailed assessment. The American Heart Association suggests that screening programs are justifiable, but not cost-effective.[1] This has led to a rare divergence of guidelines where the European Society of Cardiology promotes pre-participation screening with an ECG,[3] but the American Heart Association does not. In Australia, The Cardiac Society of Australia and New Zealand and the Heart Foundation have not published or endorsed any screening guidelines. Certainly symptomatic people should be investigated, and asymptomatic people with a family history of heritable cardiac conditions should be screened. The GP can play an important role in facilitating this. **CT**

Further reading

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References

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