BACKGROUND
Participation in regular exercise has appreciable cardiovascular benefits and there is evidence that exercising individuals have a greater lifespan. However, athletes harbouring structural or electrical cardiac anomalies may die suddenly during exercise. The public cardiac arrest of Bolton professional football player Fabrice Muamba in March of 2012 was a testament of this counter-intuitive fact, although he was fortunate enough to survive. Indeed, the risk of a young athlete dying from a structural cardiac abnormality is almost three-times greater than in young sedentary individuals, indicating that the stresses of exercise, notably high levels of circulating catecholamines, coupled with dehydration, high body temperature and minor electrolyte and pH shifts are triggers for fatal cardiac arrhythmias in affected individuals.

The sudden cardiac death (SCD) of an athlete is a highly emotional and devastating event for families of victims, peers, clubs and the sporting community. Unsurprisingly, such tragedies receive considerable media attention; headlines frequently allude to the youth of the individual involved, the number of life years lost and the circumstantial paradox, evoking intense emotion within the community at large.

SUDDEN CARDIAC DEATH IN ATHLETES
Up to 90% of all non-traumatic sudden deaths in sport are due to disorders of the cardiovascular system. In young athletes (aged < 25 years) deaths are most commonly from hereditary or congenital cardiac abnormalities whereas, in older athletes, the commonest cause of death is from coronary artery disease.

The most common causes of SCD in young athletes are the cardiomyopathies (heart muscle disorders) which account for almost 40% of all deaths reported in the literature. The main cardiomyopathies implicated in SCD during sport are:
- hypertrophic cardiomyopathy (HCM)
- arrhythmogenic right ventricular cardiomyopathy¹.

Coronary artery anomalies, especially the origin of a coronary artery from the aberrant sinus, are the second commonest cause of SCD in athletes. Other causes are: premature coronary disease, aortic rupture from Marfan’s syndrome, aortic stenosis, Wolff-Parkinson-White syndrome and long QT syndrome (Figure 1).

Football has claimed the lives of several high profile athletes in the past decade
MAGNITUDE OF THE PROBLEM AND DEMOGRAPHICS OF THOSE AFFECTED

Sudden cardiac death in sport is rare, however the exact incidence is debatable. Reliable data from the Italian pathology registry in the Veneto region (where pathologists have a particular interest in conditions causing SCD in the young) indicates a prevalence of 1 in 50,000.

Most deaths occur in males. The most vulnerable group of athletes at risk of SCD are adolescent athletes and athletes in early adulthood. Between 40 to 60% of SCDs occur in athletes under 18 years of age and 33% overall in athletes under 16 years.

Deaths are most frequent in high intensity dynamic sports of a start-stop nature. Football is the most commonly implicated sport in Europe and has claimed the lives of several high profile athletes in the past decade (Table 1). The higher death rates in football compared with other sporting disciplines can almost certainly be attributed to its popularity and high rate of participation. Of all cardiac deaths in sport, 90% occur during, or immediately after, exercise (Table 1).

WHAT ARE THE SYMPTOMS OF CARDIAC DISEASE?

Recognised symptoms of cardiac disease include central chest pain on exertion that may radiate to the arms, neck or back, breathlessness that is disproportionate to the amount of exercise being performed. Dizziness or loss of consciousness during exertion and palpitations (an awareness of an unusually rapid heartbeat) may also occur. Unfortunately, retrospective interviews with family members and sporting clubs suggest that such warning symptoms are absent in approximately 85% of cases and underscore the need for the inclusion of a simple cardiac test to facilitate the diagnosis of a heart defect.

CONTROVERSIES IN PRE-PARTICIPATION SCREENING

The sudden and unexpected death of an athlete during sport usually galvanises discussions relating to pre-participation screening (PPS) programmes to prevent further tragedies. However, PPS has been a long-standing controversial issue with several antagonists.

Arguments against screening include the low event rate and low prevalence of conditions causing SCD which means that several thousand athletes would have to

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<th>ATHLETE</th>
<th>YEAR</th>
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<tr>
<td>Piermario Morosini</td>
<td>2012</td>
<td>25</td>
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<td>20</td>
<td>Brazil</td>
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<td>Marc Vivian Foe</td>
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Figure 1: Causes of sudden cardiac death in young athletes.
be screened to identify a handful of those who may become victims of SCD. There are also concerns that electrocardiographic changes associated with physiological cardiac enlargement in response to exercise training overlap with those of pathological cardiac disorders. This may generate false positive results causing unnecessary anxiety to an athlete, or worse still, an erroneous pathological diagnosis in an otherwise perfectly healthy individual. No single cardiac investigation can identify all conditions capable of causing SCD. Although the sensitivity and specificity of identifying all conditions increases with the number of cardiac investigations performed, the cost is undoubtedly prohibitive.

The counter-argument is that the death of a young athlete results in the loss of many life years. For example, the death of an 18 year old means 65 years of life years lost (if one assumes an average United Kingdom’s national lifespan of 83 years). Furthermore, there is a definite link between exercise and SCD. Thus, timely identification of conditions causing SCD pre-mortem may prevent deaths by lifestyle modification, pharmacological therapy or implantation of an internal cardioverter defibrillator.

PRE-PARTICIPATION SCREENING MODELS

Based on several considerations, the American Heart Association and the European Society of Cardiology both agree that there should be an obligation to screen young athletes for conditions causing SCD on ethical, medical and legal grounds. These scientific organisations also agree that the most cost-effective method should be employed given the low fatality rate.

The USA has traditionally offered a long-standing low cost PPS programme, which is limited to medical history taking and physical examination. Unfortunately, the efficacy of the programme for detecting potentially lethal cardiac disease is questionable since most athletes are asymptomatic and physical examination is rarely helpful. In a series of 134 deaths in the USA, of which 115 had been investigated with a health questionnaire and a cardiovascular physical examination, a correct diagnosis was made in only one (0.9%) athlete.

In Italy, a unique state-sponsored mandatory national screening programme has been operating for 30 years. All competitive athletes are required to undergo physical examination as well as additional 12-lead ECG and limited exercise testing for the identification of predominantly hereditary or congenital abnormalities causing SCD. Athletes with abnormalities on the initial evaluation undergo further investigations and those diagnosed with a potentially serious cardiac disorder are disqualified from sport (Figure 2). The programme generates a small number of false positives (7%) creating the need for further investigation, however it has been shown to be effective in identifying individuals with Wolff-Parkinson-White syndrome, long QT syndrome and HCM, the leading cause of SCD in young athletes worldwide. Screening with ECG is ineffective for the diagnosis of anomalous coronary arteries and most cases of premature coronary artery disease do not usually manifest with abnormalities on the resting ECG (Figure 2).

WHAT IS THE IDENTIFICATION RATE WITH ECG SCREENING?

Data from the Italian programme reveals that 0.2% (1 in 500) of athletes harbour a cardiac disorder capable of causing sudden death. Studies elsewhere including the UK, Qatar, Netherlands and USA reveal figures ranging from 0.3 to 0.7%.

DOES SCREENING WITH ECG PREVENT SUDDEN CARDIAC DEATH?

Until recently, the true benefit of such a screening programme in reducing SCD in sport has been unresolved. However, a recent report on the 25-year experience of

![Figure 2: The Italian Screening Model.](image-url)
screening in Italian athletes has shown a reduction in death rate from 3.6/100,000 athletes to 0.4/100,000 athletes⁶. These figures represent a 90% reduction in SCD (predominantly due to reduced cardiomyopathy deaths) and provide the most compelling evidence available to date that cardiovascular screening does indeed reduce death rates in sport. The findings of the study have led to the endorsement of a cardiovascular screening programme for all highly trained athletes - with additional 12-lead ECG testing - by major sporting bodies including FIFA, UEFA and the International Olympic Committee.

THE QATARI EXPERIENCE OF SCREENING FOOTBALL PLAYERS
Aspetar - Qatar Orthopaedic and Sports Medicine Hospital is a sports medicine hospital built in 2007 to provide world-class sports medicine services to athletes of the region. Unencumbered by historical biases and bureaucracy, Aspetar recognises the critical role sports cardiology plays in the comprehensive care of the athlete and set about establishing a pre-eminent sports cardiology service. The goal was clear: to minimise adverse cardiac events in athletes in Qatar and around the world. As a FIFA accredited medical centre of excellence, Aspetar has been following a modified European Society of Cardiology cardiac screening protocol since 1 January 2009. As of November 2012, it has cardiovascularly examined over 5,000 athletes, including every professional football player competing in the Qatar Stars League (Qatar’s top flight competition).

Aspetar’s PPS consists of a physical examination, health questionnaire, 12-lead ECG and a cardiac ultrasound – or echocardiogram (Figure 3). A unique feature of this sports medicine hospital is its advanced diagnostic/investigative capability for the athlete that demonstrates abnormal features on their baseline PPS. These on-site modalities include exercise testing, ambulatory monitoring, cardiac MRI and cardiac genetic testing. Accordingly, it has identified 10 athletes with HCM (a prevalence of 1 in 500; in line with established data from the USA), one athlete with arrhythmogenic right ventricular cardiomyopathy, three athletes with Wolff-Parkinson-White syndrome and one athlete with Long QT syndrome.

To reach this point, Aspetar has invested heavily in high quality sports medicine physicians, sports cardiologists, physiologists, nurses and administrative staff. It has recognised the need for high quality research, yet has developed auditable clinical pathways to ensure the highest level of care in the screening of athletes with Aspetar as a ‘one-stop-shop’. Aspetar has also invested in training and resourcing its medical and associated staff to react appropriately should there be an adverse cardiac event. Finally, through the National Sports Medicine Programme, Aspetar ensures appropriate quality assurance and control procedures are in place in all sporting clubs and federations in Qatar, with particular attention paid to emergency response management in the event of a cardiac arrest.

THE BRITISH EXPERIENCE OF SCREENING FOOTBALL PLAYERS
There are no state-sponsored provisions for PPS, or any medical or legal obligations to prevent SCD in athletes, in the United Kingdom. Some sporting organisations have implemented self-funded screening that is confined to the most elite athletes. The English Football Association offers screening to all young academy players (16 years old) considered talented enough to be offered contracts to play for large football clubs. The programme provides screening for between 600 to 650 youths each year and consists of a health questionnaire, physical examination, ECG and a cardiac ultrasound. The results are interpreted by designated cardiologists. The FA pays a designated fee of £250 per player which includes the cost of the tests and the cardiologist’s report. Over 21,000 football players had been evaluated by 2009 and 0.3% (1 in 333) young academy players exhibited a cardiac disorder implicated in SCD. There is less data on screening adult professional players since most are only assessed during club transfers, however results from over
Disqualification from sport at between 14 to 16 years still leaves the player with several options to pursue an alternative career.

Football players should be screened at least once before signing a professional contract or during a transfer if a previous club has not performed a cardiac evaluation. Football governing bodies such as FIFA and UEFA are more stringent and recommend two yearly screening of all athletes competing in European football tournaments.

WHO SHOULD PERFORM THE PRE-PARTICIPATION SCREENING?

Conditions causing SCD are rare and may manifest in many different ways. Furthermore, regular exercise training may result in electrical and structural changes within the heart that are beneficial to increase cardiac output but may resemble those seen in patients with cardiomyopathies. The differentiation between athlete’s heart and cardiomyopathy is prudent since an erroneous diagnosis has the potential for serious consequences.

Based on these considerations it is essential that screening is performed in an expert setting by individuals with excellent knowledge of the conditions predisposing to SCD and the spectrum of changes seen in the normal athlete’s heart. Such practice minimises the risk of an erroneous diagnosis and the number of false positive results. For example, at St Georges Hospital’s sports cardiology clinic London, UK, the false positive rate following screening of football players is now as low as <4%. A serious disorder is identified in 0.3% of players screened.

IS THERE A ROLE FOR THE AUTOMATED EXTERNAL DEFIBRILLATOR?

As with any screening programme, the use of ECG is not fool-proof. There are certain conditions that cannot be identified with ECG (e.g. coronary artery anomalies), and some athletes may die of acquired disorders such as myocarditis which may not have been present during the PPS. Therefore, measures should be taken to protect all athletes if they experience a cardiac arrest during a game. The use of automated external defibrillators (AED) has emerged as a useful intervention against SCD during cardiac arrest in the athletic arena. Early and effective use of AEDs has shown to improve survival after cardiac arrest by 64%. However, an effective emergency response plan is required which involves adequate training of personnel, availability of AED, excellent communication systems and regular review of the emergency response protocol.

The outcome in the case of Fabrice Muamba could have been so tragically different had it not been for the actions of the well-practised medical team at pitch side.
CONCLUSION

The deaths of numerous high-profile football players within the sporting community have focused attention on the phenomenon of ‘the athlete’s heart’. Early recognition of cardiac pathology can potentially save young lives. PPS of young competitive athletes is recommended by both the American Heart Association and the European Society of Cardiology. The European recommendations have been endorsed by the IOC and the football governing bodies, with FIFA and UEFA advocating mandatory screening of all players participating in European championships. These recommendations are based on the 25-year Italian experience, which has shown that PPS of young athletes with a 12-lead ECG is effective in reducing SCD predominantly from cardiomyopathies and electrical disorders of the heart, at the expense of a small number of false-positive tests.

References

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Image: Cameroonian midfielder Marc-Vivien Foe receives aid during the Confederations Cup semi-final in 2003. The 28-year-old was stretchered off the pitch in the 72nd minute unconscious. He was later pronounced dead.