Sudden cardiac death
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Sudden death is an unexpected death that occurs instantaneously or within a few minutes of an abrupt change in a person’s previous clinical state. In the sporting world it has become an all too familiar spectacle. A seemingly healthy, vibrant young athlete collapses and dies for no apparent reason. These deaths have far reaching effects not only on family, team members, and classmates, but also on the community as a whole.

Sudden death can be categorized as either traumatic or nontraumatic in etiology. In athletes, traumatic causes include deaths from blunt force or penetrating trauma, such as may be experienced in collision sports (eg, subdural hematoma, cervical spine fractures, etc). Nontraumatic deaths may be further subdivided into cardiovascular and noncardiovascular events. Leading causes of noncardiovascular sudden death include hyperthermia, rhabdomyolysis, and asthma [1]. Noncardiovascular causes of sudden death are very real risks that face today’s athletes. Recent events involving the deaths of two high-level athletes have illustrated the potentially devastating consequences of hyperthermia and asthma. Fortunately, these conditions are often recognized by their clinical manifestations and symptoms, thereby avoiding potential life-threatening situations. In contrast, cardiovascular mechanisms of sudden death often have an initial presentation that results in death, with a diagnosis made only at autopsy. The unfortunate fact that postmortem diagnosis is the rule rather than the exception places a heavy burden on the sports medicine community; namely, that identifying individuals at risk for sudden cardiac death (SCD) is of paramount importance but is obviously extremely difficult. This task has generated debate regarding the most appropriate screening methods to evaluate athletes for SCD. To further complicate matters, many athletes develop cardiovascular adaptations (the so-called “athletes heart”) to physical exercise that can further cloud the diagnostic process.

Despite the potential pitfalls in the prevention of SCD, sports medicine professionals must have an understanding of the normal cardiovascular adaptations...
to exercise, and the epidemiology, etiology, diagnostic evaluations, management, and prevention strategies of the most common causes of sudden cardiac death.

**Athletic heart syndrome**

The athletic heart syndrome (AHS) is a constellation of physiologic adaptations to exercise that include, but are not limited to, cardiac chamber enlargement, increased ventricular wall thickness, and increased resting vagal tone. These physiologic changes may mimic pathologic cardiovascular findings on physical examination and diagnostic studies.

The first documented report of AHS was in 1899, when Henschen recognized that competitive cross-country skiers had larger hearts than sedentary controls [2]. He further demonstrated a positive correlation between an athlete’s cardiac size and his race performance, and therefore considered this a beneficial adaptation to training. In the early 1900s, technologic advances afforded more extensive evaluation of this cardiac enlargement. Using radiological and electrocardiographic data, these cardiac adaptations were thoroughly examined and found to be similar to the cardiac changes of prolonged untreated hypertension. This supported the theory that the increased cardiac size in athletes was a pathologic response to the increased cardiac stress of exercise. Currently the opinion of the medical community has come full circle regarding the athletic heart. It is now known that the athletic heart syndrome represents a constellation of normal physiologic adaptations to training that allow normal or improved cardiac function in contrast to the cardiac dysfunction of pathologic hypertrophy [3].

In order to understand AHS it is essential to understand the normal physiologic response of the cardiovascular system to exercise. Exercise training affects the cardiovascular system both peripherally and centrally to increase maximal oxygen consumption (VO₂max). The peripheral response occurs mainly in the skeletal muscle, where adaptations improve oxygen extraction. These changes include increased numbers of capillaries, mitochondria, and oxidative enzymes that allow improved uptake and utilization of oxygen. The central adaptations occur in the heart to maximize oxygen delivery to the exercising skeletal muscle. The primary adaptation to exercise is an increase in stroke volume that directly improves cardiac output. This increase in stroke volume occurs due to physiologic cardiac dilation and hypertrophy. Associated with this increase in stroke volume (SV) is a decrease in resting heart rate (HR). Because the cardiac output needed at rest remains constant before and after training, the increase in stroke volume necessitates a comparable decrease in resting heart rate (CO = SV × HR). These physiologic changes may occur in sufficient magnitude to alter the normal physical examination, electrocardiograph (ECG), and other diagnostic studies, and may, as a result, mimic pathologic cardiac conditions. Collectively, these changes are termed the athletic heart syndrome.

Although athletic heart syndrome is a physiologic marvel, the associated physiologic changes can make it difficult to differentiate this benign condition.
from the potentially fatal cardiac abnormalities that occasionally occur in young athletes. Numerous physical examination findings are associated with AHS, including alterations in cardiac rate and rhythm. This is likely due to the increased vagal tone that accompanies extreme cardiac conditioning. Other than bradycardia, the rest of the vital signs are usually within normal limits. Left ventricular hypertrophy can frequently be documented through percussion or a laterally displaced point of maximal impulse (PMI). Cardiac auscultation reveals normal S₁ and S₂ commonly with a systolic ejection murmur. This physiologic murmur must be differentiated from the characteristic pathologic murmurs—namely, the subaortic outflow murmur of obstructive hypertrophic cardiomyopathy. Typically, the physiologic murmur intensity will increase and decrease proportionally with left ventricular filling. Therefore in the supine position the physiologic murmurs are the most intense, due to increased left ventricular filling; when standing (or with Valsalva maneuver) the murmur will become less intense, due to decreased left ventricular filling. Diastolic murmurs are not normally associated with athletic heart syndrome and should be thoroughly evaluated. Electrocardiographic abnormalities are common in athletic individuals. Bradycardia, sinus arrhythmia, and first-degree heart block are frequently reported in resting athletes [4]. When associated with AHS, these abnormalities cause no symptoms and resolve with exercise. If athletes with these abnormalities complain of syncope or presyncope, a full work-up should be pursued.

Echocardiography has significantly enhanced the understanding of athletic heart syndrome. Currently this diagnostic tool can be used to accurately discern between benign and pathologic cardiac conditions [5]. Because of the inordinately low pretest probability for congenital/acquired cardiac abnormalities in a young athlete, no diagnostic study will significantly modify the diagnosis. Therefore, the only true candidates for echocardiographic evaluations at this time are those athletes with an abnormally high pretest probability of having a cardiac abnormality (ie, symptomatic athletes).

**Epidemiology of sudden cardiac death**

Because nontraumatic sudden death in young athletes is a rare event, incidence estimates vary significantly. The population sampled, age of athlete, and definition of sudden death will all affect incidence numbers. In a ten-year study by the National Center for Catastrophic Sports Injury, 160 cases of nontraumatic sudden death were identified. One hundred cases were determined to have a cardiovascular cause. Based on athletic participation data, estimated incidence rates of sudden death in high school and collegiate athletes were calculated, with an overall incidence of approximately 1 in 180,000 per year [1]. Males were noted to have an estimated death rate five times that of females, with the highest rates seen in basketball and football players. Maron et al examined 158 cases of sudden death among competitive athletes over a ten-year period and found a similar increased risk among males (120 deaths) compared with females (38 deaths). Basketball and
football were again the most common sports involved, accounting for 68% of the deaths. Overall, 85% of the deaths were attributable to a cardiovascular cause [6]. Although these studies indicate an increased death rate among males, the numbers may be misleading. Males may participate at a higher level of intensity and may be more apt to hide or deny symptoms, putting them in a higher risk category for sudden death events [1]. It must also be remembered that these studies looked solely at young competitive athletes, so caution must be used when defining risk for the general population.

The incidence of sudden death in the general population is difficult to define. Studies regarding sudden death in this setting are scarce and limited both methodologically and by a small number of cases. Nonetheless, they provide some insight into the scope of sudden death in the general population. In a study of exercise-related deaths in Rhode Island, previously healthy joggers (aged 30 to 65) were estimated to have a sudden death incidence rate of 1 in 15,240 annually. When joggers with known coronary artery disease (CAD) were included in this estimate, the incidence rate doubled (1 in 7620) [7]. In a separate study of physically active men in Seattle, the incidence of sudden death was calculated to be approximately 1 in 18,000 [8]. In a 6-year prospective study in Marion County, Indiana, the authors recorded a total of 44,481 deaths. Based on necropsy results, 16 of these deaths occurred in athletes dying suddenly from nontraumatic causes. This resulted in an incidence rate of approximately 1 in 3600 per year [9]. The discrepancies among these three studies in estimating incidence of sudden death demonstrates how difficult it is to get an accurate incidence measurement.

One constant among sudden death incidence studies is the role that age plays on incidence. Specifically, these studies show that sudden cardiac death in individuals younger than 30 to 35 is rare, and that individuals older than 30 to 35 have a higher incidence of sudden death [1,6,7,10,11].

**Etiology of sudden cardiac death**

Several studies have elucidated the most common causes of sudden cardiac death in young athletes [6,12,13]. Based on these studies, it is clear that below the ages of 30 to 35 a variety of primarily congenital cardiovascular diseases are responsible for the majority of sudden deaths. For those athletes 35 years of age and older, atherosclerotic coronary artery disease is clearly the leading cause of sudden cardiac death, accounting for up to 80% of such events [14,15] (Fig. 1).

**Cardiovascular disease and sudden death in the older athlete**

Although atherosclerotic coronary disease may occur in individuals younger than 30 to 35, it is a rare cause of sudden death in this age group. As noted above, past the age of 35, coronary artery disease becomes the predominant etiology. Sudden death during exercise in these individuals is thought to result from rupture of an atherosclerotic plaque, with subsequent coronary thrombosis [16]. This
mechanism differs from that operative in individuals who die suddenly at rest. In a study of 141 sudden death cases, Burke et al [17] compared the coronary arteries of men who died suddenly during exercise with those who died at rest. In the exercise group, 68% of 25 cases had rupture of an atherosclerotic plaque, compared with 23% of the 116 men who died at rest. The study also showed that the exercise group had plaques that were “unstable” and more prone to rupture.

Fig. 1. Estimated prevalence of causes of sudden cardiac death in trained athletes. <35 years compared with those >35 years of age. AS = aortic stenosis; LAD = left anterior descending coronary artery; C-M = cardiomyopathy; ARVD = arrhythmogenic right ventricular dysplasia; MVP = mitral valve prolapse; CAD = coronary artery disease; HCM = hypertrophic cardiomyopathy. An additional 2% of autopsy cases showed no evidence of cardiovascular disease sufficient to attribute a cause of death. (From Maron BJ, Epstein SE, Roberts WC. Causes of sudden death in the competitive athlete. J Am Coll Cardiol 1986;7:204–14; with permission.)
Given the above data, investigators have increasingly sought to define how exercise may precipitate sudden death in people with coronary artery disease. Numerous theories on this subject have been postulated. Among the possible mechanisms suspected are that physical activity: (1) produces mechanical shear stresses on the coronary arteries leading to plaque rupture [18]; (2) increases platelet aggregation, leading to thrombus formation [19]; or (3) increases systolic blood pressure, which may destabilize pre-existing fissures in coronary plaques [20]. Although each of these mechanisms has supporting evidence, it is likely that a combination of these factors is ultimately responsible.

Noncoronary causes of sudden cardiac death in the mature athlete include acquired valvular disease, mitral valve prolapse, and hypertrophic cardiomyopathy [10]. The number of sudden deaths attributed to these cardiac problems combined, however, still remains small in comparison with those resulting from CAD.

**Sudden death in the young athlete**

In young athletes, coronary artery disease plays little role in the pathogenesis of sudden cardiac death. In the United States, hypertrophic cardiomyopathy (HCM) and coronary anomalies account for over 50% of all cases of SCD [1,6]. Other causes include increased cardiac mass, myocarditis, ruptured aortic aneurysm (often from intrinsic aortic weakness with Marfan’s syndrome), arrhythmogenic right ventricular dysplasia (ARVD), valvular disease, and conduction system abnormalities (long QT syndrome) (Table 1).

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Hypertrophic cardiomyopathy

Hypertrophic cardiomyopathy is a complex primary cardiac disease that is diverse in its genetic, phenotypic, and clinical manifestations. It is typically acquired as an autosomal dominant genetic disorder caused by one of several mutations in genes that encode proteins of the cardiac sarcomere. The etiologic genes presently identified encode for (1) beta-myosin heavy chain on chromosome 14, (2) alpha tropomyosin on chromosome 15, (3) cardiac troponin T on chromosome 1, and (4) myosin-binding protein C on chromosome 11 [21–24]. Although there are ongoing efforts to characterize the specific genetic mutations responsible for HCM, DNA diagnostic techniques are not readily available for clinical practice.

Prevalence estimates have indicated that HCM is a relatively uncommon disease. Echocardiographic screening of a cohort of asymptomatic young adults noted HCM to be present in 0.17% of studied patients [25]. Electrocardiogram screening of 12,000 adults in Japan was followed by echocardiography in a subpopulation of this group. This process resulted in HCM being diagnosed in 0.2% of the study participants [26]. Given the similar prevalence data of these studies, HCM is generally estimated to occur in roughly 1 in 500 individuals. The fact that HCM (the leading cause of SCD in young adults) occurs at this rate and yet the incidence of sudden death is only approximately 1 in 200,000 underscores the heterogeneity and variable presentation of the disease process.

The hallmark cardiac finding in a HCM is that of a nondilated, hypertrophied left ventricle. The hypertrophy is usually asymmetric and unaccounted for by other disease processes. Significant structural variation exists, however, with patients exhibiting different patterns of, and wide variations in, extent of ventricular thickening [27]. Histological features of HCM include myocyte disorganization, cardiac tissue fibrosis, and the presence of intramural coronary arteries.

Clinical manifestations of HCM are often absent until a sudden death episode occurs. A major reason for this scenario is that the majority of patients with HCM have a nonobstructive form [28]. Thus the disease is often clinically silent, making detection difficult. Patients with HCM, however, may present with dyspnea on exertion, chest pain, or syncope. Dyspnea on exertion is due to restricted filling of the thickened left ventricle during exercise. Decreased diastolic filling time further exacerbates this condition. Chest pain is thought to be secondary to subendocardial ischemia, as a result of the thickened myocardium and abnormal intramural coronary arteries limiting blood supply and subsequently oxygen flow to the myocardium. Syncope can occur with obstruction of the left ventricular outflow tract, leading to a loss of cerebral blood flow. The mechanism of sudden death is thought to be secondary to malignant arrhythmias generated by abnormal conduction through thickened heart muscle [29].

A diagnosis of HCM should be suspected in any athlete with exertion-related cardiac symptoms. Other historical clues include a family history of sudden death or unexplained syncope. Detection of a systolic heart murmur that increases in intensity with Valsalva maneuver or in a standing position (decreased venous return, increased degree of outflow obstruction) is an important diagnostic clue, and should prompt further evaluation. Echocardiography has remained an impor-
tant imaging tool aiding the diagnosis of HCM. Findings suggestive of HCM on echocardiography include demonstration of thickening of the left ventricular wall, with or without systolic anterior motion of the anterior leaflet of the mitral valve (causing outflow obstruction). Although the degree of left ventricular thickening averages 21 mm to 22 mm in patients with HCM, this number can vary widely [28]. Asymptomatic patients with ventricular wall thickening on the order of 13 mm to 15 mm represent a gray area, as well-trained athletes may demonstrate similar findings that are physiologic [4]. In addition, individuals with HCM who have not reached physical maturity may have normal left ventricular thickness on echocardiography. This should not be interpreted as a sign that they are safe from developing pathologic hypertrophy in the future. Consequently, routine echocardiography should be employed with individuals suspected of having HCM until they reach physical maturity [30].

**Coronary artery abnormalities**

After HCM, coronary artery malformations are the next most frequent cause of sudden cardiac death in young athletes. This group of congenital vascular anomalies accounts for approximately 12% to 20% of sudden cardiac deaths in individuals younger than 35 [1,6,12]. One of the most frequently encountered coronary anomalies associated with sudden death is anomalous origin of the left main coronary from the right sinus of Valsalva [6,13]. In these individuals, the left main coronary artery is forced to take an oblique route between the aorta and the pulmonary trunk (Fig. 2). Restricted blood flow in this anomalous coronary is most likely to occur during exertion, as the aorta expands with greater stroke volume. At this point, the coronary ostium is thought to be compressed, with subsequent limitation in the coronary circulation. In conjunction with the restricted blood flow is an increased oxygen demand of the myocardium. This perfusion-demand mismatch can result in ischemia or infarction [10]. These ischemic episodes are believed to occur sporadically, but may be cumulative over time, resulting in patchy myocardial necrosis or fibrosis. This injured myocardium may then become a nidus for life-threatening ventricular tachyarrhythmias [31]. A similar situation exists in which the right coronary artery arises from the left sinus of Valsalva. This abnormality presents the same potential consequences, with compromised flow in the right coronary artery [32]. The combined prevalence of anomalous origins of the coronary arteries has been estimated at 0.17% [33].

Other less common coronary artery malformations that have been observed in sudden death cases include tunneling of the left anterior descending artery, hypoplasia of the right coronary or circumflex arteries, and congenital absence of the left coronary artery [6,13].

Premortem diagnosis of coronary artery anomalies is extremely difficult, because sudden death is frequently the presenting symptom. Some individuals may experience chest pain, syncope, or palpitations, which are commonly exertion related. Physical examination is typically normal, as are resting electrocardiograms [10]. Although echocardiography (both transthoracic and transesophageal) may be useful [33], coronary angiography remains the con-
Fig. 2. Top: anomalous origin of the left coronary artery from the right sinus of Valsava is one of the more common congenital coronary artery anomalies capable of producing sudden death. The unusual and circuitous course traveled by the left main coronary artery between the aorta and pulmonary artery is depicted. Bottom: drawing demonstrating normal anatomy and course of the left coronary artery. R.C.A. = right coronary artery; L. Circ = left circumflex coronary artery; Pul. A. = pulmonary artery; L.A.D. = left anterior descending coronary artery. (From Maron BJ, Epstein SE, Roberts WC. Causes of sudden death in the competitive athlete. J Am Coll Cardiol 1986;7:204–214; with permission.)
firmatory diagnostic test of choice. For those individuals fortunate enough to be diagnosed before a fatal event, most coronary anomalies are amenable to surgical correction.

Myocarditis

In 1993, the unexpected deaths of several highly recognized athletes were attributed to myocarditis [34]. Although these cases are uncommon, myocarditis is implicated in up to 6% of sudden cardiac deaths among young athletes [6,12]. Sudden cardiac death may occur in the actively infected individual or in the “healed” phase of myocarditis.

Myocarditis is usually caused by a viral infection, with Coxsackie B virus as the most frequently identifiable pathogen [35]. The viral infection results in an inflammatory process of the myocardium, characterized by monocyte infiltrate and myocyte necrosis. Viral myocarditis may present with symptoms of fatigue, dyspnea on exertion, and exercise intolerance. In other cases, patients may experience syncope, presyncope, or palpitations. Signs of heart failure with systolic dysfunction may accompany these symptoms. Individuals may also remain asymptomatic. In this latter group, sudden death may be the initial presentation.

Sudden cardiac death occurs when a fatal arrhythmia is generated in the irritated or scarred myocardium. Echocardiography in affected individuals is likely to demonstrate a decrease in left ventricular ejection fraction, and may show wall-motion defects. Recovery from myocarditis can take months. During this healing phase, individuals continue to be at risk for potentially lethal arrhythmias. Therefore, before returning to physical activity, individuals must be thoroughly screened and cleared, if appropriate, by a cardiologist. Guidelines for athletic participation in patients recovering from myocarditis are detailed in the 26th Bethesda Conference papers regarding eligibility for competition in athletes with cardiovascular abnormalities [36].

Aortic rupture

Aortic dissection and rupture may be associated with Marfan’s syndrome, an autosomal dominantly inherited disease that affects the connective tissue. Specifically, affected individuals have abnormal cross-linking of collagen and elastin. Skeletal, ophthalmologic, and cardiovascular manifestations are evident in affected people (Table 2). Pathologic changes in the aorta (cystic medial necrosis and degeneration of elastic elements) lead to aortic root dilatation. Subsequent aortic dissection and rupture is the etiologic mechanism for sudden cardiac death [6].

Screening of athletes is recommended for men taller than six feet and women taller than five feet, ten inches who have two or more physical manifestations, or who have a family history of Marfan’s. An electrocardiogram and a slit-lamp exam can be helpful initial screening steps. Echocardiography can then be used to measure the degree of aortic root dilatation. Affected patients should
have echocardiography approximately every six months to monitor aortic root dimensions.

**Arrhythmogenic right ventricular dysplasia (ARVD)**

Many other cardiovascular conditions have been associated with sudden cardiac death in young athletes but are relatively uncommon. ARVD deserves special mention because it has been implicated as the leading cause of sudden cardiac death in competitive athletes in a specific region of Italy [12,37]. This discrepancy between the American and Italian literature may be due to genetic or geographic predisposition, or may be the result of the methodology used in the studies. Specifically, the systematic use of echocardiography as a part of the athletic screening process in parts of Italy may have identified and restricted a significant number of athletes with hypertrophic cardiomyopathy. Because ARVD seems to be more difficult to identify premortem, the number of resultant ARVD sudden deaths would be relatively high compared with those caused by HCM, because many of the HCM athletes had already been identified and disqualified prior to any complications.

ARVD is a heart-muscle disorder characterized pathologically by myocyte death and subsequent fibro-fatty tissue replacement of the right ventricular myocardium. The extent of infiltrated myocardium is variable, and may cause sudden death by functional failure or by producing fatal arrhythmias. Currently the cause of ARVD is unknown. The noninvasive diagnostic study of choice is magnetic resonance imaging [38]. This study is more likely to be able to differentiate HCM from ARVD, which may not be possible with echocardiography alone.

**Valvular disease**

Both aortic stenosis and mitral valve prolapse have been found to be rare causes of sudden cardiac death in young athletes [1,6]. Aortic stenosis is an unlikely source of sudden cardiac death among athletes, primarily because it is readily identified on clinical exam by its characteristically loud systolic crescendo-decrescendo murmur. As a result, these individuals are likely to be restricted from participation in athletic activities [10]. Mitral valve prolapse (MVP) is a rather
common finding among the general population, occurring in approximately 5% of individuals. Despite this high prevalence, it has been directly implicated as a source of SCD among young competitive athletes in only a handful of instances [10]. As a result, a diagnosis of MVP by itself does not require restriction from athletic participation at any level, unless there is evidence of significant cardiac functional compromise [36].

**Long QT syndrome**

Long QT syndrome (LQTS) is a disorder characterized by lengthening of the repolarization phase of the ventricular action potential, leading to torsades de pointes, polymorphic ventricular tachycardia, and sudden cardiac death. LQTS may be congenital or acquired. Congenital LQTS is a heritable ion-channel disease caused by a number of genetic mutations of the sodium-potassium pump. Acquired factors can predispose certain athletes to LQTS through electrolyte abnormalities (low potassium, magnesium, calcium), marked bradycardia (which occurs with athletes at rest), intracranial pressure changes (subarachnoid hemorrhage, stroke), and HIV [39,40].

The diagnosis of LQTS is primarily clinical and should be pursued if athletes complain of unexplained symptoms varying from dizziness to syncope. If a family history of sudden death is known or the patient is symptomatic, then an ECG should be evaluated. Up to 94% of congenital LQTS patients have corrected QT intervals (QTc) >440 ms; therefore the ECG will be a useful diagnostic study. Immediate treatment includes intravenous fluids with potassium and magnesium to prevent any torsades recurrences. If necessary, temporary transvenous cardiac pacing can also be employed. Long-term treatment involves attempts to normalize the QT interval in order to prevent torsades. This may be accomplished pharmacologically with the use of beta-adrenergic blockers, mechanically by an implantable pacemaker/cardioverter-defibrillator, or surgically by left thoracic sympathectomy.

**Commotio cordis**

The term commotio cordis is used to describe sudden death as a result of low-impact blunt trauma to the precordial region of the chest wall. Sudden death by this mechanism has been documented in young individuals participating in a variety of recreational and organized activities [41,42]. The US Commotio Cordis Registry has catalogued 128 cases as of September 2001 [41]. Review of this database reveals that the majority of cases occurred in young individuals (mean 13.6 years old) who were engaged in competitive sports (62%). Ninety-five percent of the affected individuals were male. The most encountered mechanism of injury was from a projectile striking the chest of the individual. Projectiles included baseballs (41% of all cases), softballs (11%), hockey pucks (7.8%), and lacrosse balls (3.9%). Other less frequent projectiles noted were a soccer ball and a cricket ball [41].

Postmortem analyses of individuals dying from commotio cordis typically reveal no structural damage to the heart or overlying protective structures.
(sternum, ribs), but soft-tissue contusions of the left chest wall are commonly visualized [42]. The absence of cardiac structural damage suggests that sudden death in these cases results from blunt force-induced conduction abnormalities. Link et al developed a swine model to simulate the mechanism of sudden death in commotio cordis. In this study they inflicted a low impact blunt trauma to the chest wall of anesthetized pigs, using a wooden object that approximated the size and shape of a baseball. They found that there was a window in the cardiac cycle 30 to 15 milliseconds before the peak of the T-wave where ventricular fibrillation was inducible by the force of impact from the wooden object (velocity of the ball was 30 mph). Ventricular fibrillation was not inducible by impacts at other times of the cardiac cycle [43]. Link et al later found that the chest-wall impacts occurring over the center of the heart had the greatest propensity to trigger ventricular fibrillation, whereas impacts over noncardiac sites of the chest wall do not result in ventricular fibrillation [44]. Given the accuracy and timing required to produce ventricular fibrillation in these experimental models, it is understandable why commotio cordis is a fortunately rare clinical event.

Commotio cordis is usually a fatal event. Of the 128 confirmed cases on file with the US Commotio Cordis Registry, only 21 athletes survived. Early cardiopulmonary resuscitation and defibrillation offer the best chance of survival [41]. Because the overwhelming majority of these cases end in death, focus has been aimed at prevention. Possible prevention strategies include the use of protective padded equipment that covers the area of the chest over the heart (through a full range of sport-appropriate body positions) and switching to balls or pucks made of softer materials. Although these interventions may be feasible, unless they are employed on a national scale for a significant time period it will be difficult to ascertain any potential benefit because of the extremely low incidence rate of commotio cordis.

Role of automatic external defibrillators

Because dysrhythmias, particularly ventricular fibrillation, play an important role in mechanism of sudden cardiac death across a variety of etiologies, the availability of automatic external defibrillators (AEDs) has been postulated as a way to increase the chance of survival from a sudden-death episode. In the athletic environment, anecdotal evidence suggests that early defibrillation with an AED may help improve survival. Maron et al reported two subjects who experienced commotio cordis with resultant ventricular fibrillation that was terminated by the prompt use of an AED [41]. These two subjects accounted for 10% of all survivors of commotio cordis in the study population. Although fortuitous case examples such as these exist, there are no published studies that evaluate the usefulness of AEDs in aborting sudden death in the athletic setting. Such a study may not even be feasible, given the low incidence of sudden death in young individuals. For example, with a sudden death incidence of 1/180,000 per year, a university with 600 competitive athletes can expect to encounter an episode of nontraumatic sudden death once every 300 years.
In the general population, cardiac arrests in high-traffic public places may warrant the strategic placement of AEDs to improve prehospital cardiac care [45]. AEDs have also been recommended to be available on airplanes, in health clubs, and on emergency medical service vehicles. A meta-analysis of six studies in which first responders used AEDs with out-of-hospital cardiac arrest patients determined that survival to hospital discharge was improved over the control group (odds ratio 1.74) [46]. Whether or not access to an AED at sporting events becomes a commonplace occurrence is yet to be determined. Certainly in covering mass participation events where thousands of athletes of all ages and unknown medical histories are participating, the availability of an AED makes intuitive sense. The need for an AED on the sideline of an athletic contest is less clear.

Summary

Sudden cardiac death is a rare but devastating event. The majority of cases in young athletes are caused by congenital cardiac abnormalities that are routinely clinically silent before causing sudden death. An optimal screening practice to help identify underlying asymptomatic cardiac abnormalities has met with much debate. Beyond the American Heart Association’s recommendations for cardiovascular screening guidelines for the preparticipation physical examination [47], there are conflicting views regarding the use of more advanced diagnostic screening tests.

Athletes in whom a potentially life-threatening cardiovascular abnormality is found face the probability of being restricted from participating in certain types of athletic activity. Participation guidelines for athletes with cardiovascular disease are detailed in the recommendations of the 26th Bethesda Conference [36]. Future goals should continue to focus on the prevention of SCD. The development of a cost-effective screening process that incorporates the use of echocardiography, although having its own set of inherent limitations, may prove to be the most viable option.

References


