Received: 11 May 2017 | Accepted: 20 May 2017

DOI: 10.1111/chd.12494

SPECIAL SECTION ON CORONARY ANOMALIES

WILEY Congenital Heart Disease

Sudden cardiac death in the young: Epidemiology and overview

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Abstract

Sudden cardiac death (SCD), particularly in the young athlete, is a rare though devastating event for families, institutions, and communities at large. It can also affect the nonathlete and occur at rest, although most commonly associated with exercise activities and/or sports participation. Common causes of SCD include cardiomyopathies, particularly hypertrophic cardiomyopathy in the United States, congenital coronary artery anomalies, channelopathies, among others. This report will explore an overview of the prevalence and causes of SCD in the young.

KEYWORDS

arrhythmia, cardiomyopathy, channelopathy, coronary artery anomalies, sudden cardiac death

Sudden cardiac death (SCD) in the young is especially tragic.[1] Often SCD is the first symptom of the underlying cardiac disease. While the problem of SCD in the young is often highlighted by SCD in athletes, in fact, SCD extends to nonathletes as well. In the athlete, the risk of SCD is between 0.5 and 1 athletes for each 100 000 athletes per year.[2] However, the risk for all young individuals may be much higher. In a database from the Resuscitation Outcomes Consortium the incidence of cardiac arrests in individuals under age 21 was 8 per 100 000 years.[3]

In NCAA athletes, SCD accounts for 16% of the deaths compare to accidents which account for 51%, suicides for 9%, homicide for 6%, and cancer for 7%.[4] Causes of SCD in this population include autopsy negative SCD (the most common and found in 31%) followed by anomalous coronary arteries in 14%. Hypertrophic cardiomyopathy (HCM), dilated nonischemic cardiomyopathy and myocarditis each accounted for 8% of the SCDs. In Maron's series of 1866 deaths in young competitive athletes 416 (22%) were due to traumatic injury, 65 (3%) secondary to commotio cordis and 1049 (56%) due to cardiovascular causes.[5] HCM was found in 251 (24%), coronary artery anomalies in 119 (11%), possible HCM in 57 (5%), myocarditis in 41 (4%), and arrhythmogenic right ventricular dysplasia (ARVC) in 30 (3%). In military recruits SCD incidence is much higher (13 per 100 000 years),[6] and anomalous coronary arteries are most commonly found (33%) followed by myocarditis (20%), coronary artery disease (CAD) (16%), and HCM (13%). Males appear to be more likely to undergo SCD than females.[7] In addition, nonwhites are overrepresented in SCD registries compared to Whites.[5]

Hypertrophic cardiomyopathy has a prevalence of 1 and 500 individuals.[8] It is most commonly nonobstructive at rest, and therefore individuals will have no cardiac murmurs. It does not manifest phenotypically until the middle to late teens and the genetic abnormalities involve the contractile cardiac proteins. Young individuals are more likely to die suddenly; with age, heart failure deaths and stroke become more common. Risk factors for SCD include syncope, family medical history of SCD due to HCM, massive wall thickness, nonsustained ventricular tachycardia (VT) and a hypotensive response to exercise.[9]

Anomalous coronary arteries are the most common cause of SCD in many series. It is important to note that death occurs during exertion and that approximately one half of individuals have symptoms, most commonly exertional syncope, in the weeks prior to death.

Commotio cordis, SCD due to blunt chest trauma, is now is reported up to 20 times a year.[10] Commotio cordis occurs in young males with baseball, hockey, softball, and lacrosse the most common sports. Impacts occur directly over the cardiac silhouette. An animal model has demonstrated ventricular fibrillation with chest impact; important variables are timing to a narrow window on the upslope of the T-wave, velocity of ball, impact site, hardness of impact object, and individual susceptibility (Figure 1).[11]

Arrhythmogenic right ventricular dysplasia is the most frequent cause of SCD in Italian athletes.[12] It is also an inherited condition characterized by morphologic fatty replacement and thinning of the right ventricular wall. Subsequent reentry arrhythmias arise from this abnormal substrate and typically arise during exercise. Importantly, it has been shown in both an animal model and more recently in the humans that regular intense exercise increases the risk of arrhythmias and the development of heart failure.

The *long QT syndrome* involves abnormalities in cardiac channel which are involved in repolarization. These individuals have

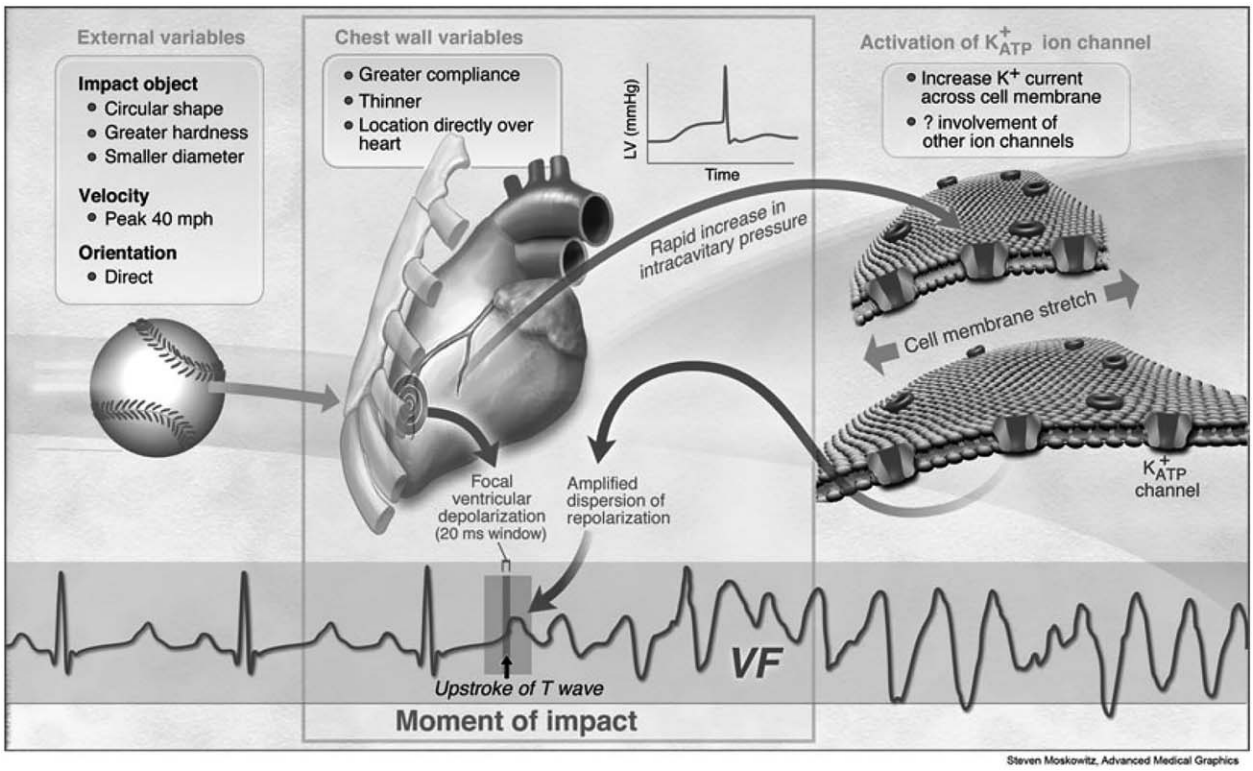

FIGURE 1 Commotio cordis; initiation of ventricular fibrillation with chest impact. From *Journal of Cardiovascular Electrophysiology*, with permission.[1]

arrhythmias typically precipitated by emotional or physical stress. The long QT 1, 2, and 3 syndromes account for 75% of the long QT individuals. Importantly these 3 genotypes can be diagnosed with genetic testing. There is increasing evidence that knowledge of the specific L QT subtype is useful for management. Diagnosis may be difficult because of the overlap in the QTCs of normal individuals compared to the others with long QT syndrome.

Coronary artery disease may also occur in individuals under age 35. Typically, one-half dying suddenly with coronary disease had evidence of an old infarct, and three-fourth have evidence of an acute infarct

Myocarditis both acutely increases the risk of SCD and can cause myocardial scarring which increases the risk of SCD lifelong.

Catecholaminergic polymorphic VT is a rare disease that presents in childhood and young adults. Patients present with exercise-induced polymorphic VT and bidirectional VT. Arrhythmias generally occur at a similar exertional threshold. The prognosis is poor.

Early repolarization has been associated with sudden cardiac arrest. In survivors of SCD with no other underlying cardiac abnormality early repolarization is seen in up to one-third. However, early repolarization is also quite common in normal individuals, and especially common in young athletes. Importantly, the early reports that early repolarization in inferior leads was more deadly have not been confirmed. Thus, outside the setting of sudden cardiac arrest, the finding of early repolarization should not lead to further evaluation or restriction from sport.

Malignant bileaflet mitral valve prolapse syndrome has been described to consists of bileaflet mitral valve prolapse, abnormal T waves and nonsustained VT. Single leaflet mitral valve prolapse is likely not to increase the risk of SCD.

Spontaneous coronary artery dissection is particularly observed in young females. Individuals present with acute myocardial infarction secondary to coronary artery dissection and intimal bleeding. These individuals also have marked tortuosity and smooth narrowing of the coronary arteries.

Myocardial bridging is thought not to increase the risk of SCD, but does not decrease the risk of arthrosclerosis due to tortuous flow and turbulent flow.

In conclusion, SCD in the young is generally due to underlying cardiac disease. These diseases may be structural or due to channelopathies. However, precise diagnosis is often lacking both precardiac and postcardiac arrest.

CONFLICT OF INTEREST

None.

DISCLOSURES

None.

AUTHOR CONTRIBUTIONS

The author contributed in the concept/design, drafting, critical revision and approval of the article.

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How to cite this article: Link MS. Sudden cardiac death in the young: Epidemiology and overview. *Congenital Heart Disease*. 2017;12:597–599. https://doi.org/10.1111/chd.12494