Texas Heart Institute Journal

Ali Massumi Cardiac Arrhythmia Symposium

Sudden Cardiac Death in Athletes

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Sudden cardiac death (SCD) in athletes is a rare, tragic event that often captures public interest, especially among individuals who value physical health and wellness. We review the most common causes of SCD in athletes and present expert recommendations for managing their treatment and exercise.

Although exercise and improved physical fitness have been correlated with significantly lower risks for cardiovascular disease–related death, limited emerging data reveal a possible U-shaped curve of the relationship between leisure-time physical activity and death. This suggests that long-term extreme and strenuous athletics may induce pathologic cardiac remodeling that nullifies the cardiovascular benefits of exercise. Moreover, participating in competitive sports has been linked to increased genetic penetrance and arrhythmic risk in those with underlying heart disease, thus acting as an acute trigger for increased risk of SCD. However, it is clear from observational studies that sudden death is exceedingly rare in athletes (5–10 per 1 million annually) and rare in the absence of heart disease.

Hypertrophic Cardiomyopathy

Hypertrophic cardiomyopathy (HCM) is the most common cause of SCD in young adults <25 years old, although the risk of sudden death persists through midlife and beyond.[1] In addition, the differential diagnosis of cardiac hypertrophy can include athlete's heart, infiltrative storage disorders, hypertensive heart disease, and so on.[2] Given the challenges of studying the SCD population and the lack of formally controlled studies, current exercise guidelines from the American Heart Association and the American College of Cardiology are based on expert opinion and case studies.[2,3] Previous guidelines recommended that patients with HCM in addition to left ventricular hypertrophy (LVH) should strictly limit athletic participation to low-intensity (class IA) sports such as golf and bowling.[4] However, in more recent guidelines, this restriction has been relaxed, with greater emphasis on shared decision-making.[2] According to the current consensus, the asymptomatic, genotype-positive HCM patient without LVH can reasonably participate in competitive (class IIA) athletics, and that implantable cardioverter-defibrillator (ICD) criteria for athletes with HCM should not differ from those for nonathletes (Class III recommendation).[2]

Anomalous Coronary Arteries

Anomalous coronary arteries are the second most common abnormality associated with SCD in young athletes. They are epitomized by the anomalous origin of the left coronary artery from the right anterior sinus of Valsalva. Death occurs during exertion, when transient aortic root dilation compresses the anomalous artery, leading to cardiac ischemia and ventricular tachyarrhythmias. The condition is difficult, if not impossible, to screen for; however, nearly 50% of athletes have some symptoms of exertional syncope in the weeks before SCD.[3]

Arrhythmogenic Right Ventricular Cardiomyopathy

Arrhythmogenic right ventricular cardiomyopathy (ARVC) is another leading cause of death in young athletes. Characterized by morphologic fibrofatty replacement and thinning of the RV wall,[4] ARVC is an inherited disease of the desmosomal proteins. It

★ ***CME Credit***

Citation:
Tsang DC, Link MS. Sudden cardiac death in athletes. Tex Heart Inst J 2021;48(4):e207513. doi: 10.14503/THIJ-20-7513

Key words:
Arrhythmias, cardiac; arrhythmogenic right ventricular cardiomyopathy; athletes; cardiomyopathies; cardiomyopathy, hypertrophic; cardiovascular abnormalities; commotio cordis; death, sudden, cardiac; sports

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is associated with an increased risk of heart failure and reentrant arrhythmias arising from the abnormal substrate, with syncope the strongest predictor of SCD.[5] Exercise appears to be an independent risk factor for expediting expression of the disease phenotype and triggering fatal tachyarrhythmias; consequently, athletes with a definite, borderline, or possible diagnosis of ARVC should not participate in competitive sports, with the possible exception of low-intensity (class IA) sports (Class III recommendation).[4,5]

Long QT Syndrome

Long QT syndrome (LQTS) is a complex and multifactorial disorder that predisposes individuals to life-threatening ventricular arrhythmias, usually precipitated by emotional or physical stress. More than 11 genetic abnormalities involving potassium and sodium cardiac channels that give rise to LQTS have been identified, and they play important roles in cardiac repolarization.[4] Fortunately, LQTS can be treated effectively with β-blockers, permanent pacemakers, and ICDs. Symptomatic athletes with suspected or diagnosed cardiac channelopathy should be fully informed about their condition and refrain from all competitive sports until a comprehensive evaluation has been completed and treatment has started; patients who are asymptomatic after 3 months of treatment may resume competitive sports (Class I recommendation).[3,4] For athletes with either symptomatic LQTS or electrocardiographically manifested LQTS (QTc >470 ms in men and >480 ms in women), participation in competitive sports (except swimming) may be considered after 3 months of treatment and after precautionary measures are implemented (Class IIb recommendation).[3,4]

Coronary Artery Disease

Coronary artery disease is the most common cause of death in athletes >35 years old and is responsible for a substantial percentage of SCD in young athletes. Autopsy data reveal evidence of old infarcts in approximately one-fourth and acute infarcts in approximately three-fourths of these SCD cases. Therefore, athletes with known coronary artery disease should undergo left ventricular function evaluation (Class I recommendation) and maximal exercise testing, to gauge their exercise tolerance and to test for inducible ischemia and exercise-induced electrical instability while receiving medical therapy including β-blockers (Class I recom-

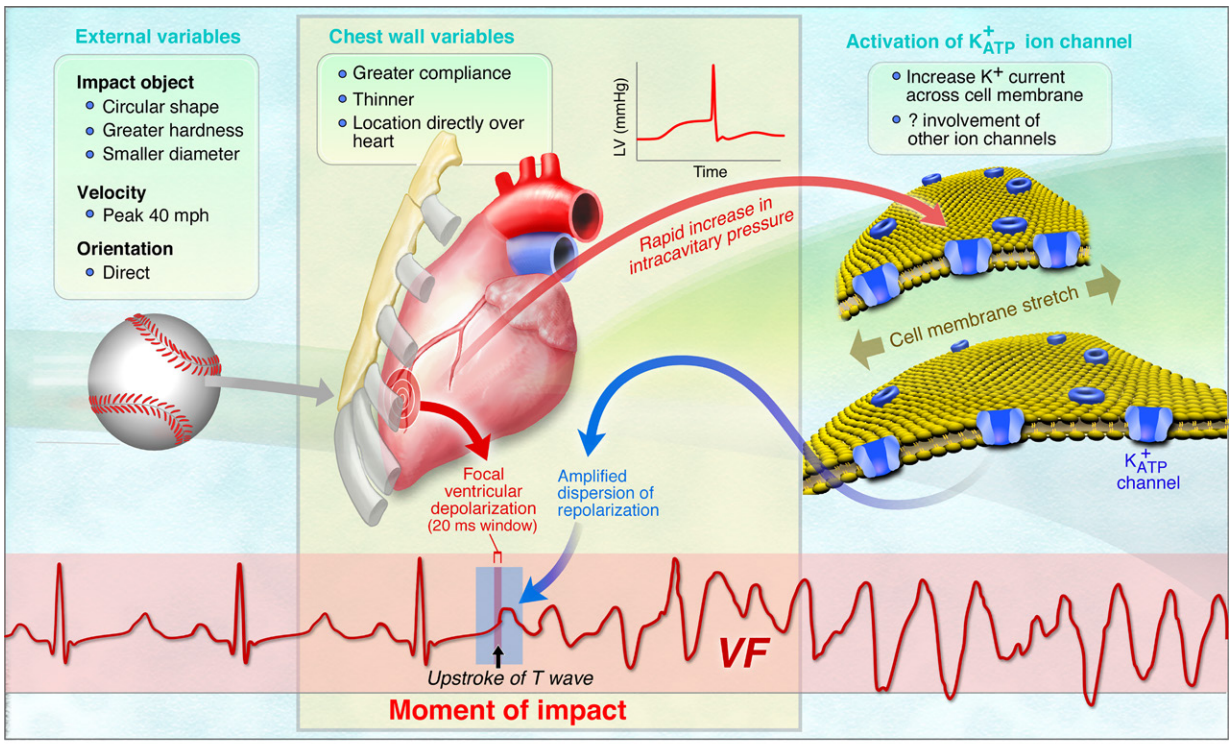

Fig. 1 *Illustration shows variables and proposed mechanism of commotio cordis. Several variables of commotio cordis have been described in animal models and include characteristics of the impact object such as shape, hardness, diameter, and velocity. In addition, cell membrane stretching likely leads to ion channel activation, which increases the dispersion of repolarization and provides the substrate for ventricular fibrillation.*

K^+_{ATP} = ATP-sensitive potassium; LV = left ventricle

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mendation).[3] After the evaluations are complete, patients need to decide whether the health and psychological benefits of exercise outweigh the risks of SCD. According to the current consensus, athletes can reasonably participate in all competitive activities if their resting left ventricular ejection fraction is >50% and they have no inducible ischemia or electrical instability (Class IIb recommendation).[3]

Commotio Cordis

Commotio cordis, defined as sudden cardiac arrhythmia caused by a blunt, nonpenetrating impact to the chest wall, may account for a substantial percentage of sudden deaths in the young. Before the turn of the 21st century and improvements in resuscitation rates, commotio cordis was the second leading cause of SCD in athletes and was frequently associated with baseball and fistfights.[2] Various factors affect survivability, including the shape of the impact object, as well as its hardness, diameter, velocity, and vector orientation; likewise, the athlete's chest wall compliance, overlying habitus, and site of impact affect outcome (Fig. 1).[6] In theory, commotio cordis leads to a rapid increase in intracavitary pressure and focal ventricular depolarization, resulting in amplified dispersion of repolarization and life-threatening ventricular fibrillation. Therefore, improved resuscitation methods, including wide availability of automated external defibrillators, and mitigation strategies, such as chest wall protection and use of safety baseballs, may decrease the incidence of SCD due to commotio cordis.

Indications for Implantable Cardioverter-Defibrillators

Several factors warrant consideration in athletes who have cardiomyopathy and ICDs. According to a European Society of Cardiology position statement, the indications for ICD implantation in competitive athletes with cardiomyopathy should be no different from those for the general population, and being an athlete should not be a primary or unique indication for ICD implantation.[5] Before returning to sports, athletes need to know that an ICD does not alter the arrhythmogenic substrate or prevent malignant arrhythmias. Above all, the decision to participate in competitive sports should be individualized, and it should be made only after carefully considering the type of underlying cardiomyopathy and fully disclosing the ICD-related risks including shocks, lead failure, and trauma.

Conclusion

Exercise is generally good for individuals, including most with cardiomyopathies, and is still recommended for health promotion and cardiovascular disease prevention. Sudden death in athletes is rare and almost always involves underlying heart disease. However, current screening techniques are suboptimal. Commotio cordis may account for a substantial percentage of sudden deaths in the young. The decision to continue participating in sports should be shared between athletes and their physicians, and efforts to reduce the incidence of SCD should include resuscitation.

Published: 15 October 2021

Section editor: Mohammad Saeed, MD, FACC

Meeting presentation: Presented at The Ali Massumi Cardiac Arrhythmia Symposium; Houston, 15 February 2020.

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