Syncope in Young Athletes

David P. Chan, MD, Director of Electrophysiology, Columbus Children's Hospital Heart Center

Brad Smith Medical Photography

Syncope is a transient loss of consciousness and postural tone due to inadequate cerebral perfusion. Up to 15 percent of children have had at least one episode of syncope. The most common cause is vagally mediated. When it occurs in young athletes, it is particularly dramatic. In part, this is due to the media's attention when athletes have died on the playing field. The most common sports associated with sudden death in young athletes are basketball and track. Fortunately, most young athletes with syncope usually do not have a malignant pathology and the incidence of sudden death is rare among healthy athletes. It is estimated that it occurs in 1 in 100,000 to 300,000 athletes. Because potential life threatening cardiac conditions may present with syncope as the initial symptom, malignant cardiac pathology should be considered. The challenge is to distinguish these from the more common benign etiology. This article will discuss the warning signs and screening guidelines that may predict increased risk for sudden death. With advent of new technology and resuscitative practices, improved survival can also be achieved.

Vagally Mediated

Vasodepressor syncope is the most common cause of syncope in children. These otherwise healthy children develop syncope in the upright position or during specific situations that may trigger onset of symptoms. The more common symptom includes pain when over-heated. For the athlete, symptoms often occur after cessation of physical activity while just standing. There is usually a prodrome that may include dizziness, nausea, headaches, blurry vision, seeing spots, feeling hot and/or abdominal pain. Loss of consciousness rarely lasts more than one to two minutes. There are usually little to no symptoms after regaining consciousness.

One of the commonly accepted pathophysiology mechansims involves activation of the Bezold-Jarisch reflex either with being upright or due to exposure to a noxious stimulus. The reflex results in paradoxic bradycardia and/or vasodilation, which ultimately compromises the perfusion of the central nervous system, leading to syncope. When the patient loses postural tone and becomes supine, venous return improves, perfusion normalizes, and the patient regains consciousness. If the patient is 'helped' to an upright position too soon, the symptoms can quickly recur.

Cardiac Syncope

There are three general categories of cardiac causes for syncope. In almost all cases, the syncopal episode occurs during the act of physical exertion. In contrast to vasodepressor syncope patients, these patients rarely will have warning of impending loss of consciousness. The duration of the syncopal episode can also be variable. These are important distinctions that can be critical in guiding the need for more extensive evaluations.

The most common cause for cardiac syncope is arrhythmia related. Interestingly, SVT, the most common tachyarrhythmia in children, rarely will cause an otherwise healthy child to pass out. The exception is in patients with Wolff-Parkinson-White syndrome who may develop syncope if there is concurrent atrial fibrillation. Rapid stimulation of the ventricles from this form of atrial tachyarrhythmia can induce ventricular fibrillation and sudden death. Although this has been described in adults, it rarely occurs in children.

Ventricular tachycardia and fibrillation are rare in the young athlete. Survivors of congenital heart surgeries that pass out should be evaluated for life-threatening arrhythmias. In otherwise healthy children, possible causes for ventricular arrhythmias include long QT syndrome, Brugada syndrome, arrhythmogenic right ventricular dysplasia and acute myocarditis. With the exception of myocarditis, causes are often genetic in origin. Unique to long QT syndrome, a family history of previous drownings, death immediately after being startled and/or congenital deafness may be present.

In addition to these abnormalities, children with normal hearts may develop a lethal arrhythmia if there is blunt trauma to the chest. Commotio-cordis is more common among males with peak age of occurrence between 13 and 18 years old. In one reported series, only 16 percent survived the event. Although chest protectors that are commonly available may help, in this series, 28 percent of the victims were wearing one of these devices.

Bradyarrhythmias, i.e. sinus node dysfunction or complete heart block, typically are found in survivors of congenital heart surgery. In otherwise healthy children, hypothyroidism, autoimmune diseases, and Lyme disease should be ruled out. Stokes-Adams attacks can occur in bradycardia-related syncope. These patients, after loosing consciousness, can develop tonic-clonic seizure movements that are short in duration but does not develop post-ictal symptoms.

Structural heart diseases make up the third category of cardiac causes for syncope. Obstructive heart lesions that can cause syncope include aortic stenosis (AS), hypertrophic obstructive cardiomyopathy (HOCM), mitral valve stenosis, left atrial myxoma, pulmonary venous obstruction and pulmonary hypertension. With exercise, cardiac output does not increase appropriately due to the fixed obstruction. In addition, patients with AS and HOCM can develop ischemia during exertion. In patients with HOCM, there may be major coronary arteries that are bridging vessels, i.e., surrounded by myocardium. During exercise, increase in contractility can lead to complete occlusion of the vessel. The resulting compromise in myocardial perfusion can lead to ventricular arrhythmias, syncope, and sudden death.

Additional patients that may have ischemia with exercise include patients with aneurysmal coronary arteries as a result of past diagnosis of Kawasaki's disease. A rare congenital defect that can lead to syncope is anomalous course of the left coronary artery. In this lesion, a major vessel courses between the aorta and the pulmonary trunk. With exercise, the increased pressure and flow through the great vessels can act as a vise to compromise coronary perfusion. If ischemia occurs, ventricular fibrillation may follow.

Marfan's syndrome patients may also have sudden collapse due to rupture of the aorta. These patients with marfanoid features will present with complaints of stabbing chest pain that radiate to the back. This is often the harbinger of impending rupture of the aorta and therefore should be referred for immediate medical care.

Neurologic Syncope

Most seizures are not related to postural position or with exertion. They can occur anytime and usually there is an aura prior to the episode. Many have incontinence. Most importantly, the loss of consciousness and tonic clonic movements persists for a longer period of time even when the patient becomes supine. Post-ictal symptoms are usually present.

Heat Stroke

Heat stroke should be considered in the athlete that has passed out. Typically it occurs during times of high temperature and humidity. The patient will show signs of severe dehydration and elevation in body temperature.

Evaluating the Patient with Syncope

The initial evaluation of a young athlete who has lost consciousness should be oriented to determining if the child is hemodynamically stable. The vitals signs, including heart rate/rhythm, blood pressure, oxygen saturation, and temperature, will guide the initial need for emergent resuscitation. The presence of ventricular fibrillation should prompt immediate defibrillation with 2 to 4 joules/kg. Once the child is stable, thorough investigation for the cause can be performed.

History

The history component is the most important part of the evaluation. Distinguishing characteristics of each type of syncope can guide the work up in the most appropriate and cost effective manner. The position of the patient associated with symptoms, presence of tonic clonic movements, incontinence, physical appearance, duration of loss of consciousness, and symptoms after regaining consciousness should be noted. A thorough family history eliciting any history of sudden unexplained deaths or near deaths could indicate a familial tendency for malignant arrhythmias.

There are multiple signs, symptoms, and historical medical information that may be very helpful in screening young athletes that may have increased risk for sudden death. Specific guidelines for a thorough screening questionnaire are available from published sources. The questions are focused in revealing clues to individuals that may warrant referral to a specialist for a more thorough evaluation. Among the important items are symptoms of syncope, shortness of breath with exercise, and familial unexplained sudden death, including SIDS. Individuals who are responsible for clearing young athletes for participation should be vigilant in performing a thorough screening process.

Physical Examination

It is unusual to find any physical abnormalities in patients with vasodepressor syncope. Occasionally, orthostatic blood pressure changes can be elicited. Children with structural heart disease typically have findings consistent with their specific pathology. Findings of neurologic deficits should raise a high index of suspicion for seizures as the cause for the syncopal episode. The examination should include complete vital signs. Heart rate that is either inappropriately slow or fast may portend the presence of arrhythmias. Listening for murmurs should be performed while the patient is both sitting and supine. In addition, a murmur that is accentuated when rising from a squatting position could indicate a dynamic left ventricular outflow obstruction as seen in HOCM. Upper extremity hypertension and/or decreased lower extremity pulses may indicate the presence of a coarctation. Examination should encompass all vital organs. Details of the rest of the physical exam are beyond the scope of this article.

It is clear therefore, the medical professional that performs the screening physical examination should be skilled in detecting important abnormalities to rule out structural heart disease. Although some states require physicians to administer this screening evaluation, this is not uniformly the case. This is obviously a point of discussion for what constitutes an adequate examination

Diagnostic testing

An electrocardiogram should be performed in every patient that presents with syncope. Specific attention should be given to rule out heart block, pre-excitation and long QT intervals. Any value above 0.44 seconds may indicate the diagnosis of long QT syndrome. Establishing a definitive diagnosis of long QT syndrome remains difficult. Genetic testing for the genetic abnormalities associated with long QT syndrome is evolving and hopefully will help in identifying at risk individuals. Patients with ARVD, Brugada syndrome, or cardiac structural disease all may have unique EKG findings. Unfortunately, an EKG is not 100 percent specific or sensitive for these diagnoses.

Although countries such as Italy and Japan perform universal screening EKG's in all children, this is not the case in the United States. The reasons include the economic burden on society, the potential for false positives and the relatively low sensitivity to detect structural abnormalities.

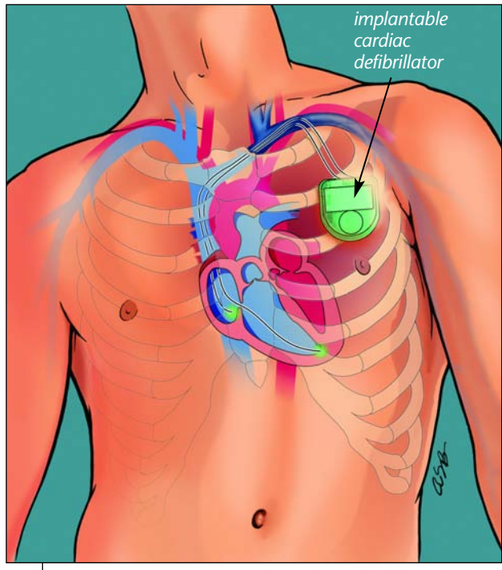
Other cardiac diagnostic tests, including exercise test, tilt table test, echocardiograms, Holter monitors, trans-telephonic event monitors, and electrophysiology testing, should be reserved for patients who have histories or physical findings that indicate a specific diagnosis that warrant these tests.

Therapy

Chronic treatment of patients with vasodepressor syncope is based on our understanding of the Bezold-Jarisch reflex. Patients are counseled to maintain adequate hydration. The patient should avoid all caffeinated beverages due to the diuretic effect of caffeine. We have reported more than 90 percent of patients with marked improvement or amelioration of their symptoms with adequate hydration. Pharmacologic intervention should be considered if fluid therapy has failed to improve symptoms. Common medications prescribed include Florinef and beta blockers. In addition, recent reports have suggested Serotonin uptake may also play a role in vagally mediated syncope. As such, there are some patients that may benefit from Serotonin uptake receptor inhibitors (SSRI). It should be cautioned that one of the side effects of SSRI's includes increase suicidal tendencies, especially in the young.

Specific therapies for the various cardiac etiologies are obviously unique to each disease. These may include medical therapy such as beta blockers for long QT syndrome, interventional therapy for valvular obstruction, pacemakers for bradyarrhythmias, or implantable cardiac defibrillators (*Figure 1*) for survivors of ventricular fibrillation related syncope. The athlete who has suspected malignant pathology should be restricted from participation in organized sports until a thorough evaluation can be completed.

Figure 1



Anthony Baker Medical Illustration

The athlete who collapses from cardiac syncope invariably has developed a malignant arrhythmia such as ventricular fibrillation or polymorphic ventricular tachycardia. The only effective therapy to restore adequate perfusion and protect vital organs involve early intervention. These include CPR to provide temporary circulatory support, activation of the emergent medical services (calling 911), and most importantly, rapid access to an automatic external defibrillator (AED-seen on page 8). The need for urgency lies in the sobering statistic that less than 5 percent of sudden death victims survive when they collapse remote from a medical facility. The success rate of defibrillation is about 70 to 80 percent in the first minute. This decreases by about 10 percent for every minute that passes. This illustrates the importance of rapid defibrillation for the best chance of survival and preservation of vital organs. As such, facilities that host events with large crowds and have activities that increase the risk of sudden death should consider having public access to AED's. The general guideline is to have these devices in close proximity to a phone so calling 911 and retrieving the device can be done efficiently. Furthermore, the American Heart Association recommends access to the device and the telephone within 3 minutes of walking time.

AED's are effective and easy to use. An interesting report demonstrated nearly the same level of accurate setup of an AED device between 6th graders and emergency medical technicians. The accuracy of the device to distinguish malignant arrhythmias and the clear verbal commands given by the machine all contribute to the relatively high effectiveness of AED's. Good Samaritan laws that are nearly universal in this country will protect the individual(s) that use the device from potential liability. Ironically, there are documented cases where negligence has been established when the devices were NOT available.

Prognosis

Most young athletes who pass out have vasodepressor syncope and usually can be managed successfully with fluid therapy alone. Over time, the severity and frequency of symptoms decreases or resolves in most patients. All patients with vasodepressor syncope can lead normal lives without physical restrictions.

In contrast, if the episode was clearly proven to be cardiac in origin or if there is significant congenital heart history, these individuals are restricted from organized competitive sports. Detail evaluation and potential corrective surgery are needed prior to lifting this restriction. Unfortunately, there are individuals who will never be cleared for full participation in sports.

David P. Chan, MD, FAAC, FAAP, director of Electrophysiology, director of Cardiology Fellowship at Columbus Children's Hospital. Medical Degree and Pediatric Residency from Wayne State University. Cardiology Fellowship at Mayo Clinic, The Ohio State University, and Rainbow Children's Hospital, Cleveland, Ohio. Board Certification in Pediatrics and Pediatric Cardiology.