Sudden Cardiac Death: A Pediatrician’s Role

Benjamin H. Hammond, MD,* Kenneth G. Zahka, MD,* Peter F. Aziz, MD*

*Department of Pediatric Cardiology, Cleveland Clinic Children’s, Pediatric Institute, Cleveland Clinic Foundation, Cleveland, OH

Education Gaps

1. There is a broad differential diagnosis to be considered in cases of sudden cardiac arrest, sudden cardiac death.
2. Timely diagnosis can circumvent progression to cardiac arrest in at-risk individuals.
3. All children should be screened and testing should be reserved for those with increased risk.

Objectives

After completing this article, readers should be able to:

1. Assess risk of sudden cardiac arrest/sudden cardiac death (SCA/SCD) with a screening history and physical examination.
2. Identify the mechanisms of distinct etiologies of SCA/SCD.
3. Recognize findings consistent with risk of SCA/SCD on a 12-lead electrocardiogram.
4. Address the concerns and questions of individuals and families after SCA/SCD.

Sudden cardiac death (SCD) or sudden cardiac arrest (SCA) may occur in populations known to be at high risk or in individuals previously unrecognized to harbor underlying disease. This review focuses on the pediatrician’s role in identifying the latter group.

DEFINITIONS

SCA is a “severe malfunction or cessation of the electrical and mechanical activity of the heart, resulting in almost instantaneous loss of consciousness and collapse” that precedes SCD. (1) SCD is defined as rapid, unexpected death from cardiac causes that occurs within 1 hour of symptoms. (2) SCD that is known to be secondary to a primary arrhythmia is termed sudden arrhythmic death syndrome (SADS).

EPIDEMIOLOGY

SCA is a common cause of death in adults, with the annual incidence estimated to be approximately 100 per 100,000, with variation seen with age, sex, and race. (3) These deaths are predominantly associated with coronary artery disease. In individuals younger than 18 years, SCA is much less common, with an estimated

AUTHOR DISCLOSURE Drs Hammond, Zahka, and Aziz have disclosed no financial relationships relevant to this article. This commentary does not contain a discussion of an unapproved/investigative use of a commercial product/device.

ABBREVIATIONS

ACC American College of Cardiology
AED automated external defibrillator
ARVC arrhythmogenic right ventricular cardiomyopathy
CPVT catecholaminergic polymorphic ventricular tachycardia
ECG electrocardiogram
HCM hypertrophic cardiomyopathy
ICD implantable cardioverter-defibrillator
LQTS long QT syndrome
LVNC left ventricular noncompaction cardiomyopathy
LVOT left ventricular outflow tract
RV right ventricle
SADS sudden arrhythmic death syndrome
SCA sudden cardiac arrest
SCD sudden cardiac death
TdP torsade de pointes
VA ventricular arrhythmia
VT ventricular tachycardia
annual incidence of out-of-hospital cardiac arrest of 8.3 per 100,000 person-years. (4) Boys are more likely to be affected than girls, with a 2:1 incidence reported from Denmark. (5) In terms of race, incidence varies by diagnosis, as demonstrated by a recent autopsy study showing hypertrophic cardiomyopathy (HCM) to be more common in African American athletes and arrhythmogenic right ventricular cardiomyopathy (ARVC) to be more common in white athletes. (6) The etiologies of SCD have been explored by anatomical and molecular autopsy, with the most common positive finding being HCM. Autopsies performed on athletes with SCD report sizable groups of autopsy-negative sudden death. (6)(7) Given the potential for false-negatives on molecular autopsy, it is reasonable to consider inherited arrhythmias as potential diagnoses in these groups.

RECOGNIZING PATIENTS AT RISK FOR SCA/SCD

There has been an increased awareness of the risk of SCA/SCD in competitive athletes. Although this risk is not exclusive to athletes, competitive athletes have been shown to be at increased risk for SCA/SCD, which has led to the implementation of preparticipation screening. (8)(9) The pediatrician has the opportunity to identify risk factors for SCA/SCD as part of regular medical care of all individuals as well as during an athlete’s preparticipation screening. The tools to identify those at risk for SCA/SCD include the patient’s family and personal history and clinical examination. Those who have previously been diagnosed to be at risk for SCA/SCD should be followed regularly by a pediatric cardiologist. Asking each patient whether they have been diagnosed previously with a cardiac condition when establishing care can identify those previously discovered to be at risk. The American Heart Association has developed a 14-element screening recommendation for competitive athletes before participation in sports (modified and elaborated in Table 1). Use of electrocardiography (ECG) as a universal screening tool is controversial and is not currently recommended in the United States. (9) We recommend that these elements of screening be applied to all patients and not only for sports participation clearance.

The Challenge of Syncope

There is little doubt that syncope/presyncope is a common presenting problem to emergency departments, urgent care clinics, and general pediatric offices. This accounts for up to 3% of all pediatric emergency department visits, with 15% to 25% of children and adolescents experiencing at least 1 episode of syncope before adulthood. Most of these events are vasovagal, and although they may result in injury, they are not a precursor for SCA/SCD. (13) Identification of an etiology for syncope that could result in SCA/SCD is essential.

TABLE 1. Recommended Elements of the History and Physical Examination (9)(10)

<table>
<thead>
<tr>
<th>PERSONAL HISTORY</th>
<th>FAMILY HISTORY</th>
<th>PHYSICAL EXAMINATION</th>
</tr>
</thead>
<tbody>
<tr>
<td>Chest pain occurring with exertion?</td>
<td>Congenital heart disease?</td>
<td>Cardiac auscultation (supine, sitting, and standing/squatting)?</td>
</tr>
<tr>
<td>Syncope/dizziness/lightheadedness with exertion?</td>
<td>Congenital deafness?</td>
<td>Femoral or pedal pulses with simultaneous radial pulse comparison</td>
</tr>
<tr>
<td>Early fatigue/shortness of breath?</td>
<td>Arrhythmias?</td>
<td>Features of Marfan syndrome</td>
</tr>
<tr>
<td>Palpitations?</td>
<td>Long QT syndrome?</td>
<td>Resting blood pressure</td>
</tr>
<tr>
<td>History of hypertension?</td>
<td>ICD implantation?</td>
<td></td>
</tr>
<tr>
<td>History of cardiac testing?</td>
<td>Sudden cardiac death before age 50 y?</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Drowning?</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Unexplained single-car accidents?</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Syncope or seizures?</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Cardiomyopathies (&quot;abnormal heartmuscle&quot;)?</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Marfan syndrome?</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Other syndromes or known genetic mutations?</td>
<td></td>
</tr>
</tbody>
</table>

ICD=implantable cardioverter-defibrillator.

*Chest pain is very unlikely to be cardiac in etiology; consider myocardial ischemia if there is an association with peak physical exertion.

*A first-degree relative of a patient with congenital heart disease is 3 to 80 times more likely to have congenital heart disease compared with the general population according to a Danish cohort study. (11)

*Systolic murmurs in hypertrophic cardiomyopathy are louder when standing from a squatting position.

*Genetic mutations leading to arrhythmias and other causes of sudden cardiac death have been identified in up to 30% of first-degree relatives. (12)
Syncope that occurs with exertion should be considered a warning sign for SCA/SCD for every health-care provider. For example, a patient who was running on the cross-country team and fell unconscious in the middle of the run would be considered to have exertional syncope. A cross-country runner who stops after a long run, sits down, then stands up and loses consciousness would typically not be considered to have exertional syncope. The most likely etiology in the latter scenario would be vasovagal syncope. Vasovagal syncope will generally involve warning signs such as auras, dizziness, and lightheadedness; whereas exertional syncope will be sudden and unanticipated. Syncope in the water (swimming, diving, etc) presenting as drowning or near-drowning should also be considered exertional syncope with the potential for SCA/SCD until proved otherwise. Individuals with exertional syncope should avoid the triggers for their syncope and avoid unsafe situations where syncope could cause injury (ie, heights, driving, swimming) until they have been fully evaluated by a pediatric cardiologist.

Family History Is Key to Effective Screening
Experience has shown that we will not get a complete family history simply by asking, “Is there any family history of heart disease?” Table 1 provides specific points to address in the family cardiac history. Because most inherited arrhythmias and structural heart etiologies of SCA/SCD are autosomal dominant, all first-degree family members of the affected individual should be evaluated by, depending on age, a pediatric or adult cardiologist with expertise in the underlying disease. In this setting, we discourage screening by ECG or echocardiography testing alone.

ETIOLOGIES OF SCD
The 2012 AAP policy statement (10) on SCA describes the associated cardiac diagnoses grouped primarily as structural/functional and electrical. We have used these major categories to frame our discussion. The structural/functional group includes cardiomyopathies (HCM, dilated cardiomyopathy, left ventricular noncompaction cardiomyopathy [LVNC], and ARVC), myocarditis, coronary anomalies and atherosclerotic disease, aortopathies, congenital heart diseases, and pulmonary hypertension. The electrical group includes inherited arrhythmias (long QT syndrome [LQTS], short QT syndrome, Brugada syndrome, and catecholaminergic polymorphic ventricular tachycardia [CPVT]), Wolff-Parkinson-White syndrome, and the traumatic electrical phenomenon of commotio cordis. Table 2 gives an overview of these diagnoses with their ECG findings and major therapies.

Structural/Functional Etiologies of SCD
Hypertrophic Cardiomyopathy. HCM is the most common cause of SCD in young people, reported to be present in 1 in 500 of the general population. (14) A review of the US National Registry of Sudden Death in Athletes (1980-2011) found that 36% of the 8,422 confirmed cardiac deaths were due to HCM.

(6) It is a disease involving a thickened left ventricle predominantly of the interventricular septum. The mitral valve can cause dynamic subaortic obstruction by anterior motion of the valve leaflets during systole coming into contact with the septum (systolic anterior motion). Individuals can develop left ventricular outflow obstruction leading to unexplained syncope with or without exertion, and eventually have symptoms of heart failure. The degree of outflow obstruction, however, is not an established risk factor for SCA. Mutations in the genes encoding cardiac sarcomeres can lead to significant myocardi remodeling and septal hypertrophy with resultant myocardial ischemia. Scar formation and disorganized myocytes make these patients prone to ventricular arrhythmias (VAs) with subsequent arrest. Medical therapy with β-blockers and calcium channel blockers is recommended in patients with symptomatic (angina, dyspnea) HCM. (15) An implantable cardioverter-defibrillator (ICD) may be indicated for patients determined to have significant risk factors. These risk factors include previous cardiac arrest, documented ventricular tachycardia (VT), a recent history of syncope, a family history of SCD associated with HCM, left ventricular wall thickness of at least 30 mm, and abnormal blood pressure response during exercise. Cardiac magnetic resonance imaging can elucidate degree of scar formation to assist in risk stratification. (16) Current sports participation guidelines for these individuals recommend avoidance of all competitive sports except those defined as low intensity because significant sympathetic activity may place them at greater risk for VT although these same guidelines offer the opportunity to liberalize sports participation in selected low-risk individuals. (17) A personal history of shortness of breath, chest pain, or syncope with exertion can raise suspicion for this diagnosis, especially when combined with a family history of SCD, cardiac surgery in early adulthood, or ICD implantation in multiple generations.

Dilated Cardiomyopathy and LVNC. Two other forms of cardiomyopathy associated with SCA/SCD are dilated cardiomyopathy and LVNC. These pediatric cardiomyopathies have a reported incidence less than 1:100,000. (18) Dilated cardiomyopathy, involving progressive dilation of the ventricles with subsequently decreased systolic function, has a poor prognosis, with 40% of children undergoing cardiac transplant or dying within 5 years of diagnosis. There is a 5-year SCA/SCD incidence rate of 3% secondary to VAs. (19)
<table>
<thead>
<tr>
<th>SCA/SCD ETIOLOGY (PREVALENCE IN POPULATION)</th>
<th>PRINCIPAL MECHANISM OF SCA/SCD</th>
<th>ECG FINDINGS</th>
<th>TREATMENT</th>
</tr>
</thead>
<tbody>
<tr>
<td>Structural/Functional Etiologies</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>HCM (1:500)</td>
<td>VAs</td>
<td>LVH with T-wave inversions in V4–V6</td>
<td>β-Blocker Myectomy, ICD for high-risk patients</td>
</tr>
<tr>
<td>DCM and LVNC (&lt; 1:100,000)</td>
<td>VAs</td>
<td>Sinus tachycardia, LVH</td>
<td>Heart failure management ICD, heart transplant</td>
</tr>
<tr>
<td>ARVC (1:2,000–5,000)</td>
<td>Fibrofatty scar formation of RV leading to VAs</td>
<td>T-wave inversions and epsilon waves in the right precordial leads</td>
<td>β-Blockers, antiarrhythmics Ablation Heart transplant</td>
</tr>
<tr>
<td>Myocarditis (3:1,000)</td>
<td>VAs associated with myocardial dysfunction/inflammation (acute), scar (chronic)</td>
<td>Low voltages, sinus tachycardia, ST and T-wave changes, variable AV block</td>
<td>Supportive care Possible use of corticosteroids, IVIG ECMO, VAD, heart transplant</td>
</tr>
<tr>
<td>Coronary anomalies (1–6.5:1,000)</td>
<td>Myocardial ischemia leading to VAs</td>
<td>Myocardial ischemia with exercise, abnormal Q waves, ST changes</td>
<td>Surgical vs catheter-based intervention</td>
</tr>
<tr>
<td>Aortopathies (6.5:100,000 for Marfan syndrome)</td>
<td>Acute hemodynamic compromise</td>
<td>LVH</td>
<td>Angiotensin receptor blockers, β-Blockers Surgery</td>
</tr>
<tr>
<td>Congenital heart disease (1:100)</td>
<td>Multifactorial: heart block, ventricular dysfunction → VAs</td>
<td>Possible AV or bundle branch block, widened QRS</td>
<td>Treat underlying anatomy May require arrhythmia surgery, ICD</td>
</tr>
<tr>
<td>PH (1:16,000)</td>
<td>Spontaneous VAs, dissection/rupture of pulmonary artery, PH crisis</td>
<td>RVH</td>
<td>Treat underlying anatomy and physiology (anti-PH medications)</td>
</tr>
<tr>
<td>Electrical Etiologies</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>LQTS (1:2,500)</td>
<td>Torsade de pointes</td>
<td>Prolonged QTc</td>
<td>β-Blockers ICD, consider LCSD</td>
</tr>
<tr>
<td>SQTS (unknown)</td>
<td>VAs</td>
<td>QTc &lt; 330 ms, tall peaked T waves</td>
<td>QTc prolonging medications ICD implantation</td>
</tr>
<tr>
<td>Brugada syndrome (1:1000)</td>
<td>Torsade de pointes</td>
<td>Coved-type ST elevations V1,V2</td>
<td>Ablation ICD implantation for high-risk patients</td>
</tr>
<tr>
<td>CPVT (1:10,000)</td>
<td>Bidirectional VT</td>
<td>Normal at baseline, exercise-induced VT</td>
<td>β-Blockers, antiarrhythmics +/- ICD (can ↑ risk of refractory VF) LCSD</td>
</tr>
<tr>
<td>Wolff-Parkinson-White syndrome (4–6:10,000)</td>
<td>Rapid anterograde conduction of atrial fibrillation</td>
<td>Preexcitation (short PR interval) delta waves</td>
<td>β-Blockers Ablation</td>
</tr>
<tr>
<td>Commotio cordis (unknown)</td>
<td>Nonpenetrating precordial trauma → VAs</td>
<td>Normal</td>
<td>Resuscitation (high mortality)</td>
</tr>
</tbody>
</table>

ARVC = arrhythmogenic right ventricular cardiomyopathy, AV = atrioventricular, CPVT = catecholaminergic polymorphic ventricular tachycardia, DCM = dilated cardiomyopathy, ECG = electrocardiogram, ECMO = extracorporeal membrane oxygenation, HCM = hypertrophic cardiomyopathy, ICD = implantable cardiac defibrillator, MIG = intravenous immunoglobulin, LCSD = left cardiac sympathetic denervation, LQTS = long QT syndrome, QTc = corrected QT interval, LV = left ventricle, LVH = left ventricular hypertrophy, LVNC = left ventricular noncompaction, PH = pulmonary hypertension, RV = right ventricle, RVH = right ventricular hypertrophy, SCA = sudden cardiac arrest, SCD = sudden cardiac death, SQTS = short QT syndrome, VA = ventricular arrhythmia, VAD = ventricular assist device, VF = ventricular fibrillation, VT = ventricular tachycardia.
LVNC is a disorder of the left ventricular myocardium wherein compaction is incomplete, resulting in prominent trabeculations with deep recesses and 2 distinct layers of compacted and noncompacted myocardium. Clinical features in both diagnoses range from severe ventricular dysfunction with end-stage heart failure to normal function without clinical features of heart failure. There is similarly a range of treatment options usually involving β-blockers, angiotensin-converting enzyme inhibitors, anticoagulants, and aldosterone antagonists, followed by mechanical support and transplant as indicated. Patients with LVNC have an increased risk of lethal VAs, with an associated SCA/SCD rate of 6%. Individuals can present with chest pain and syncope that occur with exertion or with aborted cardiac arrest. Several more can be diagnosed by screening of family members.

**Arrhythmogenic Right Ventricular Cardiomyopathy.**
ARVC has a prevalence in the general population of 1 in 5,000, with a prevalence of 1 in 2,000 individuals in Italy and Germany. It is an autosomal dominant disease caused by a genetic defect of the cardiac desmosomes leading to myocyte detachment and cell death predominantly involving the right ventricular (RV) myocardium. There is remodeling of the cellular connections and replacement of the RV myocardium with “fibrofatty” scar tissue, which can be a substrate for VAs leading to SCD. The RV will become dilated and dysfunctional, with regional wall-motion abnormalities. Presenting symptoms may be palpitations or exertional syncope. ECG can demonstrate T-wave inversions in the precordial leads (V1–V4), with decreased QRS voltages in the limb leads. Widening of the QRS complexes in the right precordial leads reflects an enlarged RV. Characteristic epsilon waves (small, peaked wave between the S and T waves) in V1 and V2 can be seen in patients with an advanced form of the disease (Fig 1A). Sympathetic activity has been associated with an increased risk of VAs in these patients such that these patients are recommended against participation in competitive sports. Heart failure can develop with dilation and dysfunction of the RV. Medical therapy has had modest success, although in patients with significant disease, medical therapy is refractory. Catheter ablation is therapeutic but not curative due to the progressive nature of this disease. Patients with recurrent VAs are ultimately listed for heart transplant.

**Myocarditis.** Autopsies of young athletes with SCD demonstrated that 7% of the cases were related to myocarditis. It involves inflammation of the myocardium, which is usually secondary to a viral infection. Other potential etiologies include bacterial, fungal, or parasitic infections; hypersensitivity reactions; autoimmune disease; toxins; and Kawasaki disease. In the acute phase of the illness, myocardial edema, poor function, and acidosis can lead to VAs and SCA/SCD. As healing occurs, fibrosis develops (chronic myocarditis), which can be a substrate for VAs. SCD secondary to VAs may occur during the acute or chronic phase of the illness, even years after the initial diagnosis and treatment. It can be challenging to diagnose patients before the disease more fully manifests itself with rapid deterioration of ventricular function. Acute myocarditis typically presents with sinus tachycardia in conjunction with decreased cardiac function, causing symptoms of heart failure. Physical findings may include extra heart sounds due to abnormal filling in the setting of heart failure. In a recent multicenter study, 87% of patients with myocarditis survived with medical management without the need for transplant. Extra-corporeal membrane oxygenation or ventricular assist devices may be required with consideration of heart transplant in refractory cases. Given the difficulty in identifying these patients in the early acute phase of the illness, we

---

**Figure 1.** A. Electrocardiogram in a teenager with multiple syncopal events during exercise shows T-wave inversions in leads V1 through V4 and epsilon waves (arrows) consistent with arrhythmogenic right ventricular cardiomyopathy. B. Electrocardiogram in a patient with Brugada syndrome with typical coved-type ST elevations in leads V1 and V2. Reprinted with permission, Cleveland Clinic Center for Medical Art & Photography ©2018.
recommend that athletes not participate in sports during intercurrent illness.

**Congenital Coronary Anomalies.** Congenital coronary anomalies were the second-most common cardiac etiology of sudden death in the US National Registry, composing 19% of those autopsied. (6) Typical normal coronary anatomy is a right and left coronary artery coming off their respective cusps in the region just above the aortic valve and traversing along the epicardium with branches diving to endocardium to provide perfusion of the cardiac muscle. Abnormal origin and courses of the coronary arteries can lead to exertional coronary ischemia. Anomalous coronary arteries may arise from the wrong aortic sinus, may course between the aorta and the pulmonary artery (interarterial course), or may course within the wall of the aorta (intramural course), which can potentially result in coronary insufficiency with exercise. This ischemia and resultant scar formation can predispose these patients to fatal VAs. The degree of coronary obstruction can be accentuated with exercise, and patients are, therefore, counseled to limit their level of exertion if they are having symptoms of chest pain during exercise. Symptomatic patients should be considered for surgery evaluation. Anomalous coronary arteries do not typically cause symptoms at rest. They may, however, cause exertional chest pain, syncope, dizziness, and palpitations that must be differentiated from more common etiologies. The key to diagnosis is either high-quality focused echocardiography or computed tomographic angiography. Unfortunately, family history and physical examination do not generally provide clues to these diagnoses.

**Atherosclerotic Coronary Artery Disease.** A rare cause of SCD in young patients is myocardial infarction secondary to premature atherosclerotic coronary artery disease. There is significant risk in patients with familial dyslipidemias. The AAP Guidelines for Cardiovascular Health and Risk Reduction in Children from 2011 recommend lipid profile screening as early as 2 to 8 years of age in patients who have a family history suggestive of early coronary artery disease or other individual risk factors. Universal lipid profile screening is recommended between ages 9 and 11 years, and then again between ages 17 and 21 years. (24) The standard practice of asking about a family history of early coronary artery disease (guidelines use a history of myocardial infarction, angina, stroke, coronary artery bypass graft, stent, or angioplasty before age 55 years in males and before age 65 years in females) can lead to early identification and preventive treatment for these patients. (24) Physical examination findings of xanthomas may aide in identifying these individuals as well.

**Syndromic and Nonsyndromic Aortopathy.** Progressive aortic dilation can be a silent process until presenting emergently or fatally with aortic dissection or rupture. This represents a small group of individuals (approximately 3%) in autopsy studies. (6) The individuals who are most at risk can be easily identified, having physical features of Marfan syndrome or Loeys-Dietz syndrome. In Marfan syndrome, a defective fibrillin-1 gene leads to a weakened extracellular matrix in the connective tissue of the aorta with the potential for dissection and rupture. The Ghent criteria are used to diagnose Marfan syndrome and they rely on systemic findings and family history. The pediatrician should ask whether there is any family history of aortic aneurysms, dissections, blindness, eye surgery, or diagnosed Marfan syndrome. Individual history should ascertain problems with eyesight (ectopic lentic) and history of spontaneous pneumothorax. Physical examination should evaluate for pectus deformity, scoliosis, reduced elbow extension, the wrist sign, the thumb sign, hindfoot deformity, pes planus, typical facial features, and reduced upper segment/lower segment and increased arm span/height. (25) These physical examination features compose much of the “systemic score” of the 2010 Revised Ghent Nosology for Marfan Syndrome, which can be calculated in the pediatrician’s office using the systemic calculator provided by the Marfan Foundation (https://www.marfan.org/dx/score). Identification and referral of these individuals is essential for regular monitoring of aortic dimensions with cardiac imaging to determine need and timing of surgical intervention (aortic graft). Recent trials suggest that treatment with angiotensin receptor blocker therapy and β-blockers can slow aortic root dilation, suggesting a potential benefit from early recognition. (26) The only clue to nonsyndromic familial aortic aneurysm may be a family history of aortic dissection or rupture.

**Congenital Heart Disease.** In 1998, Silka et al (27) reported that the risk of SCD was 25 to 100 times greater in patients with congenital heart disease than in the rest of the population. The substrate for lethal VAs can be associated with the patient’s underlying anatomy, such as can occur in patients with tetralogy of Fallot, or with the surgical scar from their repair. Individuals with tetralogy of Fallot are particularly at risk for SCA/SCD, which is believed to be associated with RV dysfunction predisposing to VAs. (28) A widened QRS interval is a factor in risk stratifying a patient with suspected or known VAs for ICD implantation. Long-term follow-up with a pediatric cardiologist is required for most repaired congenital heart defects, and some may require regular ambulatory ECG monitoring for ventricular ectopy. Even in this population,
chest pain and syncope are more likely to be noncardiac, but when there is doubt, consultation with a pediatric cardiologist is encouraged.

**Primary or Secondary Pulmonary Hypertension.** Proposed etiologies of SCA/SCD include acute pulmonary hypertensive crisis leading to low cardiac output, compression of the coronary arteries by a dilated pulmonary artery, pulmonary artery dissection, massive hemoptysis, and rupture of the pulmonary artery. (29) An acute pulmonary hypertensive crisis may occur when the pulmonary vascular resistance acutely increases (may be in response to hypercarbia, acidosis), leading to right-sided heart failure, low cardiac output, and myocardial ischemia with potential for arrest. These patients are notorious for having poor outcomes after cardiac arrest. They are discouraged from participation in competitive sports. A prominent second heart sound can arouse suspicion, and an ECG with findings of RV hypertrophy should lead to an evaluation by a pediatric cardiologist.

**Electrical Etiologies of SCA/SCD**

Individuals at risk for SADS typically present with exertional syncope. Every individual presenting with exertional syncope should have an ECG. As we discuss etiologies of SADS diagnoses, we will review the ECG findings specific to patients considered to be high risk, which can guide referral to a pediatric electrophysiologist.

**Long QT Syndrome.** LQTS is a congenital rhythm disorder with an estimated prevalence of 1:2,500 that is associated with multiple genetic mutations that affect ion channel function. It is more common in white populations than in other races. (30) Prolongation of the QT interval can be secondary to channelopathies affecting repolarization. Specifically, the IKs potassium channel, the IKr potassium channel, and the INa sodium channel are predominantly affected secondary to genetic mutations. Prolongation of the repolarization phase of the cardiac myocytes can lead to a torsade de pointes (TdP) when a premature ventricular contraction occurs during the repolarization phase of the cardiac cycle. The autosomal recessive form of LQTS can be associated with congenital deafness. The most common types are identified as types 1, 2, and 3 (LQT1, LQT2, and LQT3). Characteristic LQTS triggers include exercise and increased sympathetic tone in LQT1, auditory and emotional triggers in LQT2, and sleep in LQT3. β-Blocker therapy, which acts to reduce sympathetic tone, can benefit patients with LQT1 and LQT2. Recent recommendations for sports participation have evolved as studies have demonstrated safety in sports participation for previously asymptomatic individuals on β-blocker therapy. (31) Individuals with a history of SCD, TdP on ECG monitoring, or a more high-risk type of LQTS are more likely to get an ICD device. Most patients with LQTS will not require ICD implantation. All

![Figure 2. Electrocardiogram in a patient whose father died of sudden cardiac death that shows QT prolongation. A clinician can count the small boxes (40 milliseconds each) between the lines drawn above and calculate the intervals. The Bazett formula (QT/RR) can then be used to calculate the corrected QT interval (QTc). Machine reads often miscalculate this interval, and each clinician should, therefore, be comfortable with manual QTc measurement. Reprinted with permission, Cleveland Clinic Center for Medical Art & Photography ©2018.](image-url)
individuals with known or suspected LQTS should be counseled to avoid multiple drugs that have been associated with QT prolongation (http://www.crediblemeds.org). Accurate measurement of the QT interval involves correcting for heart rate known as the Bazett formula: corrected QT interval (QTc) = QT interval/√RR interval (Fig 2). This is a valuable skill for every clinician to develop given the frequent inaccuracy of machine-generated reads. The Schwartz criteria (32) are used clinically to determine the probability of LQTS (Table 3). Genetic testing and screening ECGs are performed for first-degree family members after a diagnosis has been made.

**Short QT Syndrome.** A rare cause of SCD is short QT syndrome (SQTS) diagnosed by ECG as a QTc of 330 milliseconds or less with tall, peaked T waves. Short atrial and ventricular refractory periods have been shown to be associated with gain-of-function mutations of the potassium channels. SCD may occur secondary to VT or fibrillation. Antiarrhythmic medications that can prolong the QT interval can be therapeutic in these individuals. Many patients require ICD implantation.

**Brugada Syndrome.** Brugada syndrome has a prevalence of approximately 0.15% in adults in Asia (specifically Japan) and a 0.02% prevalence in the West. (33) It is diagnosed by a 12-lead ECG demonstrating coved-type ST elevations (Fig 1B) in the right precordial leads (V1–V3). If there is uncertainty regarding the ST segments in the precordial leads, repositioning of leads V1 and V2 to the second intercostal space may improve capture of the characteristic ST morphology. This finding has been associated with sudden death from polymorphic VT (TdP). A mutation of the SCN5A gene (Ina sodium channel) occurs in 20% to 30% of individuals with Brugada syndrome. (34) This is the same gene that is affected in LQT3, but there is a gain of function in LQT3 compared with a loss of function in Brugada syndrome. Abnormal repolarization provides the substrate for TdP when a premature ventricular contraction comes during this phase of the cardiac cycle. These individuals can be asymptomatic and diagnosed by incidental finding on ECG. Those who present with symptoms (exertional syncope, SCA) or a spontaneous Brugada pattern on ECG are generally recommended to undergo ICD implantation. Similar to LQTS, these individuals should be counseled to avoid certain drugs (http://www.brugada drugs.org).

**CPVT.** Individuals undergoing screening for LQTS with a history of exertional syncope, but without QTc prolongation, may be found to have CPVT. There is an estimated prevalence of 1 in 10,000. (35) Depending on the history, the pediatric cardiologist may perform a stress exercise test, which could induce VAs with CPVT. These arrhythmias are associated with calcium channel mutations (most commonly RYR2), which can lead to increased calcium release. This predominantly occurs during diastole and can lead to VAs when incited by premature contractions. β-Blockers have been shown to reduce VAs in these patients. Flecainide and left cardiac sympathetic denervation are additional treatment options for those with refractory events despite adequate β-blockade. (36) ICD implantation should be approached with caution because ICD discharges can be proarrhythmic due to the release of catecholamines, leading to a clustering of recurrent episodes of VT or ventricular fibrillation in a short period (“electrical storm”). (36)

**Wolff-Parkinson-White Syndrome.** Wolff-Parkinson-White syndrome is characterized by delta waves seen on ECG. A delta wave is a slurred upstroke of a QRS complex with a shortened PR interval, representing ventricular preexcitation by way of anterograde conduction through an accessory
pathway. Although narrow complex tachycardia can lead to hemodynamic instability, the major concern in these individuals is atrial fibrillation (Fig 3) leading to ventricular fibrillation due to rapid anterograde conduction through the accessory pathway. Patients can present with a narrow complex tachycardia with delta waves seen on ECG after conversion to sinus rhythm. Symptoms can be palpitations with abrupt onset of tachycardia with or without syncope. The role of the ECG in this diagnosis is to identify individuals with preexcitation who are, therefore, at risk for SCA/SCD. These individuals should stop participation in competitive sports until they are evaluated by a pediatric electrophysiologist. Ablation of the accessory pathway is the definitive treatment in these individuals.

**Commotio Cordis.** Commotio cordis is Latin in origin, meaning “agitation of the heart.” This term has been used to describe the result of nonpenetrating trauma to the precordium resulting in VT or ventricular fibrillation. (37) There is nothing known of who, if anyone, is more at risk for commotio cordis. Prevention strategies are, therefore, used to avoid the type of trauma associated (ie, a baseball catcher or hockey goalie wearing chest protection). A recent review suggested that induced VT or ventricular fibrillation is determined by the location of the trauma (directly over the heart) and by the timing of the blow (occurring during early repolarization). (37) These patients are difficult to resuscitate, and mortality has been reported to be approximately 75%. Commotio cordis is more likely to occur in sports with projectiles, such as baseball, soccer, lacrosse, and hockey, although it can occur in any circumstance wherein there is blunt precordial trauma. It is important to counsel athletes and coaches regarding the dangers of direct blows to the chest and to use protective equipment when necessary.

**ACUTE MANAGEMENT OF SCA**

The period of effective defibrillation is believed to occur during the first few minutes after cardiac arrest. Regular training in the use of automated external defibrillators (AEDs) and cardiopulmonary resuscitation has been recommended for pediatricians and other medical staff. Pediatricians have been recommended to advocate for training in cardiopulmonary resuscitation and AED use in the community, in addition to effective placement of AEDs in the community. (38) In particular, families, schools, and

---

**Figure 3.** A. Electrocardiogram (ECG) in an adult with Wolff-Parkinson-White syndrome diagnosed by the presence of delta waves. B. Illustration of conduction down the AV node (AVN) and the accessory pathway (AP). The first illustration shows conduction down both the AVN and the AP, leading to preexcitation of the ventricle (delta wave). The second illustration shows loss of delta waves with prolongation of the PR interval when conduction is down the AVN and up the AP. The third illustration demonstrates atrial fibrillation with rapid anterograde conduction down the AP. C. ECG in the same adult patient with wide complex, irregular tachycardia consistent with rapid anterograde conduction down the AP. D. ECG in the same adult patient with a normal PR interval and narrow QRS morphology after ablation of the AP. Reprinted with permission, Cleveland Clinic Center for Medical Art & Photography ©2018.
coaches of individuals who have been identified as high risk for SCA/SCD should be prepared to act in the event of SCA.

TARGETED SCREENING

Any individual suspected of having a high risk of SCA/SCD should be referred to a pediatric cardiologist as soon as possible. If the individual has been symptomatic, suspension of the activities that led to those symptoms is recommended until a thorough cardiology evaluation. Athletes and nonathletes alike with concern on ECG for QTc prolongation (LQTS), preexcitation (Wolff-Parkinson-White syndrome), coved-type ST elevations in V1, V2 (Brugada syndrome), T-wave inversions in the lateral precordial leads (HCM), or epsilon waves in leads V1, V2 (ARVC) should not return to play until they are evaluated by a pediatric cardiologist. We reemphasize the importance of screening all individuals with the recommended individual and family history questions discussed previously herein and a thorough physical examination. In the absence of the risk factors discussed, there is a group of individuals who need to be seen by a pediatric cardiologist for targeted screening. These are the first-degree relatives of SCA/SCD victims, so-called cascade screening. (39) The term cascade refers to the branching out of genetic testing that starts with first-degree family members of an SCD victim and subsequently extends to the first-degree family members of those who test positive, continuing in this pattern until all screened individuals have been identified. Genetic counselors and pediatric cardiologists can guide this screening process, which may include genetic testing, ECG, echocardiography, or exercise stress testing.

PSYCHOSOCIAL EFFECTS OF SCD

In the aftermath of SCD, family members are traumatized and often seek clarity from their pediatrician. A recent review by Ingles and James (40) gave recommendations for follow-up with these patient families in 3 defined stages: initial contact (ensuring family support), referral to a cardiac genetics clinic, and ongoing care (management of SCD-related anxiety). There are several online resources that may be helpful to families as well as to patients who survive SCA. Involvement in these groups and contribution to awareness groups or fundraisers can benefit grieving family members. Clinicians should have a low threshold for referral to a clinical psychologist when SCD-related anxiety is significant. Although genetic screening may initially increase a family member’s anxiety in the short term, return to baseline functioning in the long-term is more likely with this knowledge. (40) In addition, clinicians should recognize and, as appropriate, attempt to alleviate feelings of guilt in family members of the deceased. Guilt may be felt for symptoms that went unrecognized, for encouragement of sports participation in an at-risk child, or even for being a carrier of the genetic mutation that led to a child’s fatal disease. Taking time to educate and reassure can help families heal.

Summary

- It is critically important to recognize exertional syncope and to immediately refer these patients to a pediatric cardiologist.
- Use of the American Heart Association–recommended preparticipation history and physical examination should be included in a standard history and physical examination for all patients in this age group because nonathletes are also at risk for sudden cardiac death (SCD). (8)(9)
- All first-degree family members of victims of sudden cardiac arrest (SCA)/SCD should be screened for SCA/SCD by a pediatric cardiologist.
- Families of SCA/SCD victims can benefit from education by a well-informed pediatrician regarding the multiple etiologies of SCA/SCD and the difficulty in diagnosing these conditions before they occur.

To view teaching slides that accompany this article, visit http://pedsinreview.aappublications.org/content/40/9/456.supplemental.

Sudden Cardiac Death: A Pediatrician’s Role

Benjamin H. Hammonds, MD, Kenneth G. Zaias, MD, Peter F. Azzi, MD
Department of Pediatric Cardiology, Case and Clinic Children’s, Taft Medical Institute.
Cleveland Clinic Foundation, Cleveland, Ohio

References for this article are at http://pedsinreview.aappublications.org/content/40/9/436.
1. A 16-year-old African American male athlete experienced a sudden cardiac death (SCD) during a basketball game. The patient was a competitive basketball player who was a member of his high school basketball team. He was otherwise healthy with no medical conditions and was taking no medications. Family history is significant for an uncle who had an SCD at age 47 years. Among the following potential causes of SCD, which one is the most likely cause in this patient?
   A. Arrhythmogenic right ventricular cardiomyopathy.
   B. Coronary artery disease.
   C. Hypertrophic cardiomyopathy.
   D. Long QT syndrome (LQTS).
   E. Supraventricular tachycardia.

2. A 10-year-old boy is brought to the clinic by his parents for a follow-up after an emergency department visit. The patient was seen in the emergency department a week ago with a 4-day history of fever, myalgias, cough, and congestion. He was diagnosed as having viral infection and was discharged home on supportive care. Today, the parents report that his symptoms have not improved. Earlier today he started having increased wet cough, vomiting, and decreased energy. He has been less active and was noted to have shortness of breath and wheezing. On physical examination today he is afebrile and in mild respiratory distress. Examination of the lungs shows diffuse coarse rhonchi and wheezes. There are subcostal and intercostal retractions. Heart examination shows sinus tachycardia, normal S1 and S2, loud S3, and an S4 gallop and no murmurs. His liver is palpated 2 cm below the right costal margin. Which of the following is the most likely diagnosis that explains the clinical presentation in this patient?
   A. Acute bronchiolitis.
   B. Acute myocarditis.
   C. Acute hepatitis.
   D. Aspiration pneumonia.
   E. Reactive airway disease.

3. A 5-year-old boy is brought to the clinic for a school physical and health supervision visit. The patient was born with tetralogy of Fallot and is status post repair. He has normal growth and development and is taking no medications. The parents are concerned about him starting school and participating in physical education. They also wonder if they should allow him to play any sports in the future. Which of the following is the most appropriate recommendation to provide the parents at this time?
   A. No restrictions on sports participation because the child has normal anatomy after repair.
   B. No restrictions on sports participation if the patient has normal electrocardiographic and echocardiographic findings.
   C. Restrict all sports participation indefinitely.
   D. Restrict participation in contact sports only.
   E. Routine cardiology follow-up with Holter monitoring for ventricular ectopy and for sports clearance decision.

2019 Pediatrics in Review now is approved for a total of 30 Maintenance of Certification (MOC) Part 2 credits by the American Board of Pediatrics through the AAP MOC Portfolio Program. Complete the first 10 issues or a total of 30 quizzes of journal CME credits, achieve a 60% passing score on each, and start claiming MOC credits as early as October 2019. To learn how to claim MOC points, go to: http://www.aappublications.org/content/moc-credit.
4. A 12-year-old girl was evaluated by a pediatric cardiologist after an episode of syncope during exercise. The episode occurred at the gym while she was watching TV while exercising on the treadmill. She was diagnosed as having LQTS. The parents are concerned and ask your opinion about potential triggers for the LQTS. In addition to exercise, which of the following could potentially be one of the triggers of LQTS?

A. β-Blockers.
B. Bright lights (visual stimulation).
C. Increased sympathetic tone.
D. Sleep deprivation.
E. Swimming in cold water.

5. You volunteer to provide the preparticipation clearance evaluation of the athletes at your local middle school. You use the American Academy of Pediatrics preparticipation screening questionnaire that is sent by the school to be completed by the parents before the sports physical examination. The questionnaire contains individual and family history questions along with history of past sports injuries. You review each completed questionnaire before you perform the physical examination. Which one of the following requires targeted screening by a pediatric cardiologist before the sports participation clearance decision?

A. All athletes before participation in competitive team sports.
B. All athletes before participation in high-intensity sports.
C. All athletes before their 12th birthday.
D. Athletes who are first-degree relatives of sudden cardiac arrest/SCD victims.
E. Athletes with a history of vasovagal syncope.
Sudden Cardiac Death: A Pediatrician's Role
Benjamin H. Hammond, Kenneth G. Zahka and Peter F. Aziz
Pediatrics in Review 2019;40;456
DOI: 10.1542/pir.2018-0241
Sudden Cardiac Death: A Pediatrician's Role
Benjamin H. Hammond, Kenneth G. Zahka and Peter F. Aziz
Pediatrics in Review 2019;40;456
DOI: 10.1542/pir.2018-0241

The online version of this article, along with updated information and services, is located on the World Wide Web at:
http://pedsinreview.aappublications.org/content/40/9/456