

Sudden Cardiac Death in the Young

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Sudden cardiac death (SCD) in children and adolescents is an uncommon event, yet it is devastating to families who lose a child so suddenly and unexpectedly. Sudden cardiac death in adults is primarily caused by coronary artery disease; however, SCD in the young is due to different etiologies. This article presents the following etiologies associated with SCD in the young: structural and functional abnormalities, primary electrical diseases also known as channelopathies, and a lesser-known cause, that is, *commotio cordis*. Prevention and treatment strategies to be discussed include noninvasive testing for risk stratification, screening for competitive athletes, genetic testing, implantable cardioverter defibrillators, automated external defibrillators, and patient advocacy. Nursing implications will include guidance for patient and family education, recommendations for sports participation, and forging partnerships with communities. **Key words:** *adolescent, child, defibrillator, sports participation, sudden cardiac death*

ALTHOUGH it is a statistically rare event in the young population, sudden cardiac death (SCD) is a devastating occurrence in the lives of families who suddenly, unexpectedly, and permanently lose a child or an adolescent. *Sudden cardiac death* is defined as a natural death due to cardiac causes indicated by an abrupt loss of consciousness within 1 hour of the onset of symptoms; preexisting heart disease may have been known to be present, but the time and mode of death are unexpected.¹ The overall incidence of SCD in the general population is approximately 1/1000 individuals and it affects 300 000 to 400 000 lives per year in the United States.² Findings based on genetic predispositions indicate that (1) a history of SCD in 1 parent equals an 80% increased risk of SCD in his or her offspring,³ (2) a history of SCD in both parents equals a 9-fold increase in risk of SCD in their offspring,³ and

(3) first-degree relatives of SCD victims have a 50% greater risk of SCD than controls.⁴ The following discussion will include an overview of the incidence of SCD in children, adolescents, and young adults, the most common etiologies associated with SCD in the young, strategies for prevention and treatment, and nursing implications.

INCIDENCE

Sudden cardiac death accounts for approximately 5% of all deaths in children⁵⁻⁷ and there are approximately 500 episodes annually of unexpected SCD in children and adolescents in the United States alone. The reported incidences range from 0.8 to 6.2 deaths per 100 000 per year in children, adolescents, and young adults^{5,8-10} and the majority have unrecognized heart disease. Nearly half of all sudden deaths occurring in primarily healthy children exhibit no abnormal findings at autopsy. Ventricular fibrillation, the incidence that was thought to be rare in children, coupled with ventricular tachycardia, is estimated to occur in 12% of children as the initial rhythm during cardiac arrest.¹¹

With respect to young athletes, that is, those involved in high school or collegiate

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sports, the estimated risk of SCD due to cardiovascular causes ranges from 1:100 000 to 1:300 000 athletes per year.¹²⁻¹⁵ In a recent study,¹⁶ the majority of sudden cardiac arrests that were exercise-related in the young occurred (1) at a rate of 69 cases per year over a 7-year period, (2) mostly in high schools, and (3) predominately in males (5:1 ratio). This study included not only competitive athletes but recreational athletes as well. Approximately 5 million athletes compete at the high school level.⁷

ETIOLOGY

The primary etiology of SCD is preexisting coronary artery disease and its sequelae, including acute myocardial infarction, previous myocardial infarction, and heart failure; this accounts for 80% of all SCD victims.¹⁷ Hypertrophic and dilated nonischemic cardiomyopathies account for the second largest etiologies of SCD, with the remaining 5% to 10% due to congenital heart disease (CHD) and ion channel anomalies, that is, primary electrical disturbances that are genetically determined.¹⁷ Since SCD in the pediatric population rarely results from coronary artery disease, with the exception of those born with anomalous coronary arteries, the etiologies associated with SCD in the young fall into the remaining 20%. The following discussion will focus on the etiologies known to be responsible for SCD in the pediatric population and will include structural/functional anomalies, primary electrical abnormalities (also known as channelopathies), and an increasingly noted condition known as *commotio cordis*.

Structural or functional abnormalities

Cardiomyopathies

In the pediatric population, cardiomyopathies are strongly linked to SCD. Although cardiomyopathy is not a common illness in childhood, it is the most common cause of cardiac muscle dysfunction in children. Systolic and diastolic heart failure, along with

electrical degeneration into lethal arrhythmias, contribute to cardiomyopathy as the most common cause of cardiovascular death in children.¹⁸ There are several types of cardiomyopathies: hypertrophic cardiomyopathy (HCM), dilated cardiomyopathy (DCM), restrictive cardiomyopathy, and arrhythmogenic right ventricular dysplasia (ARVD), commonly referred to as arrhythmogenic right ventricular cardiomyopathy (ARVC).¹⁹ The following types of cardiomyopathies will be discussed here: (1) HCM, (2) DCM, and (3) ARVD/C.

Hypertrophic cardiomyopathy

Hypertrophic cardiomyopathy is most often diagnosed during infancy or adolescence. It is the second most common form of cardiac muscle dysfunction in children; however, it is the most common cause of SCD in young, healthy, and athletic individuals and is one of the most common inherited disorders.²⁰⁻²³ HCM comprises approximately 35% to 40% of cardiomyopathies in children.²⁴ The most common symptom is syncope, presumed to be arrhythmogenic. The prevalence of HCM associated with SCD ranges from less than 1% to 3% to 6%.²⁰⁻²³ In cases of sudden death in children, approximately 30% of them were found to have HCM. *Hypertrophic cardiomyopathy* is defined as an autosomal dominant genetic heart disease.²⁵ It has characteristic morphology and is defined as a hypertrophied, nondilated left ventricle in the absence of another systemic or cardiac disease that is capable of producing significant wall thickening.²⁶ The left ventricle increases in size and thickness, particularly along the interventricular septum. In effect, this becomes a dynamic obstruction of the left ventricular outflow tract. The thick muscle wall relaxes abnormally creating an atypical filling pressure that leads to an increased left ventricular end diastolic pressure. In HCM, sudden death is most often associated with left ventricular outflow tract obstruction, myocardial ischemia, ventricular arrhythmias, or a combination thereof.²⁷ In the setting of HCM,

sudden death may also be a result of bradyarrhythmias secondary to sinus node or atrioventricular node dysfunction.²⁴

Dilated cardiomyopathy

Dilated cardiomyopathy is defined as a cardiac disorder characterized by a dilated left ventricle and systolic dysfunction that generally results in congestive heart failure.²⁸ Dilated cardiomyopathy is the most prevalent form of cardiomyopathy in children. Recent studies show that pure DCM occurs in 0.56 per 100 000 pediatric patients. The incidence is higher in males than in females, in African Americans than in whites, and in infants than in older children. The most common causes are idiopathic (unknown), myocarditis (viral and bacterial), neuromuscular disorders, and genetic DCM.²⁹ Dilated cardiomyopathy is more likely to present in the first year of life than at older ages in children. However, when DCM presents in older-aged children, the prognosis is significantly poorer.²⁹ The survival rate of DCM in the first year of life is approximately 75%, whereas in children at 5 years of age, it is 65%.³⁰

In DCM, there is an increase in ventricular chamber size; the ventricular wall thickness is significantly reduced. There is increased myocardial stretch as cardiac enlargement occurs. With atrial dilatation and sluggish flow, the contractile forces in the atrium are reduced, thereby increasing the risk of thrombi. Secondary to the weak myocardial muscle, there is abnormal ventricular filling and contraction that leads to increased ventricular end diastolic pressure. The progression of this course culminates in enlargement of the left ventricle, valve regurgitation, atrial enlargement, and fulminate heart failure with an increased risk of thrombi in the ventricle.¹⁸ This predisposes the patient to significant atrial and ventricular arrhythmias that may potentially be lethal. A recent study by Pahl and colleagues³¹ reveals that sudden death in children with DCM usually occurs within 2 years following diagnosis. Children who either had congestive heart failure or were taking antiar-

rhythmic medications at the time they were diagnosed were noted to be at higher risk for sudden death.³¹

Arrhythmogenic right ventricular dysplasia/cardiomyopathy

Arrhythmogenic right ventricular dysplasia, also known as arrhythmogenic right ventricular cardiomyopathy, is a disorder of the right ventricle (rarely is the left ventricle involved) in which the muscle is replaced by fatty tissue. Because the conduction system is intramuscular, the fatty or scar tissue may disrupt the normal electrical sequence, thus placing the patient at high risk for arrhythmias and sudden death. It is a genetically determined disease³² and the degree of impairment in ARVD/C can range from no functional impairment to severe functional impairment. The incidence of ARVD/C in the general population is difficult to assess; however, it accounts for 29% of sudden deaths in young people in northern Italy.³³

Arrhythmogenic right ventricular dysplasia/cardiomyopathy is difficult to assess in children as they do not tend to meet diagnostic criteria until the age of 8 years, with most of the sudden deaths occurring at or beyond 10 years of age.³² The exact incidence of ARVD/C in the pediatric population is unclear, but it has been estimated at 0.02% to 0.1%.³⁴ The child with ARVD/C may appear healthy and subsequently experience a syncope or sudden death event, although sudden death as the presenting symptom is rare.³² In newly diagnosed patients, right heart failure is uncommon.³⁵

Structural defects

Structural defects are those that occur when normal development of the heart and its vasculature fail to occur, resulting in congenital cardiac abnormalities. Many of these defects can be detected in the fetus and newborn, but some are not discovered until later in life as an incidental finding during a routine examination, symptoms increase as a result of the progression of the disease, or a sudden

cardiac arrest/death event. The primary structural defects associated with SCD in the young to be discussed here are congenital coronary artery anomalies (CCAA) and CHD.

Congenital coronary artery anomalies

Congenital coronary artery anomalies occur when embryonic development of the origin of the coronary arteries fails to occur in the normal manner. Normally, the right coronary artery arises from the right cusp of the aorta and the left main coronary artery arises from the left cusp of the aorta. When this does not occur, the coronary artery is said to be anomalous in origin. Coronary artery anomalies are the second most common etiology of SCD in young athletes,¹³ with an incidence based on autopsy findings of 0.17% to 0.3% and based on coronary artery angiographic findings of 0.6% to 1.55%.³⁶ The prevalence of isolated CCAA is thought to be more than 1% in the general population.³⁷

According to Virmani and colleagues,³⁸ the most lethal coronary artery anomaly is the left coronary artery arising from the right sinus of Valsalva that often results in fatal arrhythmias, often with exercise. The right coronary artery arising from the left sinus of Valsalva may also be lethal in adolescents and young adults, but unlike the anomalous left, it is more often an incidental finding at autopsy.³⁸ Ng and Maginot³⁶ concur, but believe that the addition of an intra-arterial course between the aorta and the pulmonary artery increases the risk of SCD in these 2 types of CCAAs. The percentage of individuals who have an intra-arterial course is estimated to be only 0.92%.³⁹ The mode of death for patients with CCAA is most likely coronary ischemia from compression during exercise and only one-third of individuals who die from this condition are symptomatic.⁴⁰

Congenital heart disease

Sudden cardiac death associated with CHD occurs from a variety of mechanisms. During infancy, SCD occurs because of the failure of fetal circulation to transfer to neonatal circu-

lation, cardiac arrhythmias, and/or operative, perioperative, or postoperative mortality.⁴¹ Patients with repaired CHD face a risk of SCD 25 to 100 times that of the general population.⁴² In the older child with CHD, the mechanisms are more likely to be coronary ischemias, cardiac arrhythmias, sepsis, thrombotic events, and/or crises secondary to pulmonary hypertension.⁴¹ Oechslin and colleagues⁴³ reported a 26% incidence of SCD in adults with CHD, followed by heart failure in 21% and perioperative death in 18%. Mechanisms are thought to include those previously mentioned, with the following causes less likely: stroke, aortic dissection, or rupture of vessels.⁴³ Congenital heart defects that pose a high risk of sudden death, repaired or unrepaired, include pulmonary vascular obstructive disease, hypoplastic left heart syndrome, aortic valve abnormality, Ebstein's anomaly, aortic dissection, aortic stenosis (supravalvular), transposition of the great arteries (both dextro and levo types), and tetralogy of Fallot.⁴¹

Primary electrical abnormalities

Congenital cardiac arrhythmias generally occur in individuals with anatomically normal hearts. These disorders are also categorized as primary electrical abnormalities for this reason, but more recently, with an increasing number being identified by genetic markers, these are being classified as channelopathies. This latter nomenclature is derived from the various cardiac ion channels affected by the specific disorder, for example, potassium or sodium channels. The following disorders will be discussed here: congenital long QT syndrome (LQTS), Brugada syndrome, and catecholenergic polymorphic ventricular tachycardia (CPVT).

Congenital long QT syndrome

The *congenital form of LQTS* has been defined as a genetically determined abnormality of ventricular repolarization that results in cardiac instability that can lead to potentially

life-threatening arrhythmias and/or sudden death. This potentially lethal cardiac arrhythmia is responsible for 3000 to 4000 sudden deaths in children and young adults each year in the United States.⁴⁴ The diagnosis of LQTS relies on a group of clinical and electrocardiogram (ECG) manifestations. The clinical presentations of children and adolescents with LQTS vary and can include the following: syncope (associated with noise, stress, and/or exercise), presyncope, palpitations, seizures, ventricular arrhythmias, cardiac arrest, and sudden death. Associated findings may include congenital atrioventricular block, congenital deafness (rare), syndactyly (rare), and a family history of LQTS or sudden death.

Congenital LQTS is characterized by a prolonged QT interval on the surface ECG that predisposes an individual to life-threatening ventricular arrhythmias. Syncope associated with LQTS is generally due to a specific type of ventricular tachycardia known as *torsades de pointes*. LQTS was originally characterized in 1957 by Jervell and Lange-Nielsen⁴⁵ as a syndrome involving congenital deafness, prolonged QT/*torsades de pointes*, syncope/sudden death, and autosomal recessive transmission. In 1963, Romano et al⁴⁶ and in 1964, Ward⁴⁷ described the characteristics of prolonged QT/*torsades de pointes*, syncope/sudden death, and an autosomal dominant inheritance pattern. In 1997, researchers determined that the *KVLQT1* gene was accountable for the dominant form of Romano Ward syndrome and the recessive form of Jervell and Lange-Nielsen syndrome.⁴⁸ Currently, hundreds of mutations in approximately 10 genes have been identified as being linked to LQTS.^{49,50}

The clinical features associated with LQTS vary and are closely related to the disease-specific gene. These clinical features include the QT morphology characteristics, the response of the QT interval to exercise, arrhythmia triggers, and response to various therapies.⁵¹ The first syncopal event in individuals with this disorder tends to occur in childhood with a mean age of 8 years.⁵² Without treatment, approximately 30% will have

an initial episode of syncope or cardiac arrest before 10 years of age. This incidence increases to 20% by 20 years of age.⁵²

Brugada syndrome

Brugada syndrome is a syndrome of altered repolarization, and like LQTS, it is characterized as a cardiac channelopathy. In the late 1980s, 6 cases were described in which the patients experienced ventricular fibrillation and had peculiar patterns noted on their ECGs, specifically in the ST segment.⁵³ The researchers also noted slight abnormalities in the structure of the right ventricle. In 1992, the Brugada brothers⁵⁴ described an additional 8 patients who experienced sudden cardiac arrest and who had the same ECG characteristics as those described by Martini and colleagues.⁵³ The combination of the ECG characteristics of right bundle branch block and ST-segment elevation coupled with sudden death syndrome was named Brugada syndrome.⁵⁴ By 1999, more than 200 cases of Brugada syndrome were reported and more than 75% of the patients with the disorder were males. The mean age of diagnosis is 40 years; however, it can range from 22 months to 77 years.⁵⁵ Symptomatic patients who present with SCD are generally found to have ventricular fibrillation or syncope due to polymorphic ventricular tachycardia.^{54,56,57}

This syndrome is being recognized more frequently in the pediatric population as a cause of SCD but with some distinct differences. In their study of 30 children younger than 16 years with Brugada syndrome, Probst and colleagues⁵⁸ identified 11 individuals through familial screening and 11 who presented with a syncopal event. The children had a mean age of 8 ± 4 years. All but 1 of the symptomatic patients exhibited the typical ECG pattern of ST-segment elevation in the right precordial leads (V_1 through V_3); there was no male predominance in the number of symptomatic patients. Thirteen percent experienced atrial fibrillation/flutter; yet, the most interesting finding was that nearly half of the syncopal events were related to febrile

illnesses, a finding that significantly impacts how children with Brugada syndrome should be managed.⁵⁸

Catecholaminergic Polymorphic Ventricular Tachycardia

Catecholaminergic polymorphic ventricular tachycardia (CPVT) is an inherited syncope/sudden death syndrome resulting from abnormal handling of calcium by the sarcoplasmic reticulum. First described in 1978⁵⁹ and again in 1995,⁶⁰ Catecholaminergic polymorphic ventricular tachycardia resulted in syncope in children and adolescents with a structurally normal heart and a normal QT interval. Its distinction from other forms of syncope was a form of ventricular tachycardia that was stress-related and consistently reproducible. This type of ventricular tachycardia was also found to be associated with sudden death. The first syncopal event in patients with CPVT tends to occur in childhood, with a mean age of 7.8 years.⁶⁰ Without treatment, approximately 30% of patients will experience syncope or cardiac arrest before 10 years of age, which increases to 20% at 20 years of age.⁶¹ Two genetic mutations have been found to be responsible for approximately half of the cases of CPVT.⁶²

Other

Commotio cordis

Commotio cordis is defined as a life-threatening cardiac arrhythmia caused by a direct, nonpenetrating, often low-impact blow to the chest.⁶³ It most commonly occurs on the athletic field and/or during recreational sports.⁶⁴ Cardiac arrest, primarily due to ventricular fibrillation, is instantaneous, and according to Maron,⁶⁴ the most common sports projectiles are baseballs (41%), softballs (11%), hockey pucks (7.8%), and lacrosse balls (3.9%). *Commotio cordis* is the second leading cause of death in young athletes^{64,65} and is most commonly seen in children and adolescents during common sporting activities.^{65,66} Most of the victims

of *commotio cordis* are young males, with a mean age of 14 years.^{64,67} The prevalence in children is thought to be due to the compliant chest walls in these individuals that allow for greater transmission of the impact energy to the myocardium.⁶⁸ The most common sports activities associated with *commotio cordis* are baseball/softball (58%) and hockey (16%).⁶⁸ According to the US Commotio Cordis Registry,^{64,67} the survival rate in more than 180 cases was only 15%. This is likely due to lack of early recognition and failure to institute aggressive resuscitation efforts in a timely manner, that is, within 3 minutes of receiving the blow to the chest.

STRATEGIES FOR PREVENTION AND TREATMENT

Prevention of SCD generally falls into 1 of 2 categories: primary or secondary. Primary prevention occurs in those who have not experienced a sudden cardiac arrest event, but who are at high risk for such an event. Secondary prevention occurs when an individual actually survives a sudden death event and thus has a greater chance of having another sudden death event. Secondary prevention in the context of an aborted SCD is a fairly easy treatment decision to make and, more often than not, includes placement of an implantable cardioverter defibrillator (ICD). On the other hand, primary prevention is generally reserved for (1) asymptomatic patients; (2) patients who have less-severe symptoms associated with potentially lethal conditions, for example, syncope associated with HCM or Brugada syndrome; or (3) family members of patients who experience SCD but who may show no overt signs of disease.⁶⁹ Clinicians caring for these patients and their families face difficult decisions in determining the mode of treatment with respect to primary prevention measures. Any strategy chosen should have benefits that outweigh the risks associated with the chosen therapy(ies). Stratification of risks can aid in these decisions and are an integral component of care for these individuals and their families. A myriad of diagnostic tools

are available to assist pediatric cardiovascular health care providers in stratifying the risk of SCD in select patients.⁷⁰

Noninvasive testing for risk stratification

Sudden cardiac death risk stratification measures are divided into those that are considered noninvasive and those that are considered invasive. The entire gamut of diagnostic tools used by pediatric cardiology clinicians is beyond the scope of this discussion, so invasive measures, that is, percutaneous electrophysiologic testing and diagnostic/interventional cardiac catheterization, will not be discussed here. The following noninvasive measures have been chosen as they most pertain to the etiologies associated with SCD and will include (1) history and physical examination, including the family pedigree; (2) the ECG, including 24-hour ambulatory monitoring; (3) exercise stress testing; and (4) echocardiography. Because of the increasing importance of more specialized measures to reduce the risk of SCD, the following topics will be discussed separately: (1) screening for competitive athletes, (2) genetic testing, (3) ICDs, (4) automated external defibrillators (AEDs), and (5) patient advocacy.

The history and physical examination of any child, adolescent, or young adult with a documented or suspected arrhythmia is pivotal in determining subsequent testing. The physical examination itself may not provide pertinent information, but the patient and family history is invaluable.⁷⁰ Campbell⁷¹ recommends the periodic, but the routine use of a SCD risk assessment form for patients undergoing well-child visits. (Table 1). This form can also be used during a cardiology consultation to stratify the risk of SCD and/or as a tool for preparticipation sports screening.⁷¹ In light of the recently discovered genetic predisposition of many of the etiologies for SCD, Vincent⁷² recommends that family care of these individuals includes the development of a pedigree in addition to routine family screening.

The ECG, including 24-hour ambulatory ECGs, can provide critical information on

some of the substrates that are responsible for SCD in the young. The ECG may be helping in diagnosing patients with LQTS, Brugada syndrome, ARVD/ARVC, cardiomyopathy, and arrhythmias associated with CHD. Specific electrocardiographic findings may include prolongation of the corrected QT interval, T-wave alternans associated with LQTS, ST-segment elevation and/or right bundle branch block associated with Brugada syndrome, epsilon waves and T-wave inversion associated with ARVD/ARVC, chamber enlargement and/or hypertrophy associated with cardiomyopathy, and/or ventricular arrhythmias associated with LQTS, Brugada, ARVD/ARVC, and CHD.⁶⁹ In addition, QRS duration has been used to predict patients with CHD at risk for SCD.⁶⁹ Should patients with CCAA experience myocardial ischemia during the ECG, critical information will be provided about this defect. Ambulatory monitors should not be used to diagnose a prolonged QT interval but can be used to assess arrhythmia frequency, T-wave morphology, and bradycardia-induced ECG changes. They can also be used to detect nonsustained episodes of ventricular arrhythmias in patients with HCM, CHD, and ARVD/ARVC. It is important to note that not all patients with LQTS have electrocardiographic evidence of the disease at the time of test performance.

Exercise stress testing can also yield valuable information to be used to stratify the risk of SCD. Owing to the fact that exercise can trigger arrhythmias and possible SCD in specific patients, the exercise stress testing is essential, but not limited to, the following diseases: LQTS, ARVD/ARVC, CPVT, HCM, and CCAA.⁶⁹ Factors that may be elucidated by the use of exercise stress testing in individuals for the purposes of risk stratification include failure of the QT interval to prolong during exercise in patients with LQTS, abnormal blood pressure response in patients with HCM, ischemia in patients with CCAA, and ventricular arrhythmias associated with LQTS, Brugada syndrome, CPVT, and postoperative CHD.

Echocardiography is seldom used to diagnose arrhythmias in the pediatric population,

Table 1. Sudden Cardiac Death Risk Assessment Form^a

Ask these questions (or have parents complete for your review) at periodic times during well-child visits (neonatal, preschool, before/during middle school, and before/during high school): <i>Patient history questions—tell me about any of these in your child: Yes/No</i> Has your child fainted or passed out during or after exercise, emotion, or startle? Has your child ever had extreme shortness of breath during exercise? Has your child had extreme fatigue associated with exercise (different from other children)? Has your child ever had discomfort, pain, or pressure in his chest during exercise? Has a doctor ever ordered a test for your child's heart? Has your child ever been diagnosed with an unexplained seizure disorder or exercise-induced asthma not well controlled with medication? <i>Family history questions—tell me about any of these in your family:</i> Are there any family members who had a sudden, unexpected, unexplained death before age 50 (including SIDS, car accident, drowning, or near drowning)? Are there any family members who died suddenly of "heart problems" before age 50? Are there any family members who have had unexplained fainting or seizures? Are there any relatives with certain conditions, such as - Enlarged heart—hypertrophic cardiomyopathy - Dilated cardiomyopathy - Heart rhythm problems—long QT syndrome - Short QT syndrome - Brugada syndrome - Catecholaminergic ventricular tachycardia - Arrhythmogenic right ventricular cardiomyopathy - Marfan syndrome (aortic rupture) - Heart attack (age 50 or younger) - Pacemaker or implanted defibrillator - Deaf at birth (congenital deafness) <i>Please explain more about any "yes" answers here:</i> <i>Parent signature:</i> <i>Physician signature:</i> <i>Date:</i>

^aReproduced with permission from Campbell.⁷¹

but it does have value in determining the degree of structural or functional abnormalities in patients who have disease etiologies that predispose them to SCD. For example, the diagnosis of HCM depends solely on the echocardiographic findings,⁷³ which can include asymmetric hypertrophy of the interventricular septum (with the presence or absence of systolic anterior motion of the mitral valve) and hypercontractile systolic function.⁷³ Echocardiographic findings associated with DCM include reduced ventricular wall thickness, left ventricular enlargement, abnormal ventricular filling and contraction,

increased ventricular end diastolic pressure, valve regurgitation, and atrial dilatation.¹⁸

Screening for competitive athletes

The first reports of sudden death in athletes appeared in the 1980s. The prevalence was unknown because there were such few reports. In addition, the cause of the SCD was largely unknown. Today, the incidence of sudden death in young, competitive athletes is 0.61 per 100 000 per year.⁶⁷ There is much debate in the cardiovascular and lay community on how to prevent SCD in the athlete. The definition of a competitive athlete is one

who participates in an organized team or individual sport that requires systematic training and regular competition against others and places a high premium on athletic excellence and achievement. The primary objective of screening is to reduce the adverse cardiovascular events associated with organized sports and enhance the safety of the athletic participation.⁷⁴

The current American Heart Association guidelines purpose to screen for personal and family history of cardiovascular issues. There are 12 questions: 8 are personal history and 4 are family history. If 1 question results in a positive answer, the athlete should be referred to a cardiologist for further evaluation. High school and middle school athletes must have parental verification of the answers to the questions before a referral is made.⁷⁴ The benefit to screening is that there are signs and symptoms leading to SCD that could potentially be found on a screening tool. These symptoms include chest pain, syncope, palpitations, fatigue, and dizziness. Conversely, the detriment is that the screening and subsequent testing may reveal a false-positive diagnosis. Therefore, the content of the screening tool must be thorough and asked in the correct fashion, using open-ended questions that require a definitive answer, that is, yes or no, so that the family takes responsibility for the reported history resulting in a more complete and accurate history.⁷¹

Genetic testing

Over the last 5 years, genetic testing has become increasingly available to the medical community. Currently, the disorders that are amenable to testing are HCM, LQTS, Brugada syndrome, CPVT, HCM, and ARVC. While these tests are accessible, the cost to families may be somewhat prohibitive (\$5000). Often, companies that perform genetic testing work with insurance companies to reduce the cost to the patient.

Many genotypes have been discovered for LQTS. However, the isolation of particular genes for disorders such as CPVT and ARVC is not as advanced. Therefore, testing nega-

tive for one of these disorders does not mean that the patient does not have the disease. It may only mean that another subtype of that gene has not yet been identified. The ACC/AHA/ESC guidelines⁷⁵ recommend family specific testing upon the identification of a gene-positive family member. PGxHealth, which manufactures FAMILIONTM, a series of genetic tests for inherited cardiac syndromes, is a testing company that has available genetic tests for the following diseases: LQTS, Brugada syndrome, CPVT, ARVC, HCM, and DCM. The company utilizes testing that analyzes specific ion channel genes to detect the mutations that cause cardiomyopathies and conduction system disorders. These tests also aid in the diagnoses of disorders such as LQTS, Brugada syndrome, and CPVT. This testing can help to classify genetic causes of a disorder and, upon further testing, can identify whether family members have the same mutation. This information helps to guide the care provider in selecting modalities of treatment and medication choices.

Implantable cardioverter defibrillators

Children, adolescents, and adults younger than 21 years of age comprise less than 1% of the total ICD population. In the past, ICD implantation in the pediatric population in the past was reserved for secondary prevention. However, because of the rapid identification of genetic markers for conditions that pose a risk of SCD, ICDs are being used with increased frequency for primary prevention as well. These devices have improved overall survival, that is, 95%, in a recent series of 76 patients⁷⁶ with a survival rate of 97% at 2 years and 89% at 5 years. Alternatively, ICD placement in the pediatric population is associated with significant morbidity. Complications include lead failure,^{76,77} ICD "storms" with excessive shocks,⁷⁶ infection,⁷⁶ and inappropriate shocks.⁷⁶⁻⁷⁸

Automated external defibrillators

Automated external defibrillators were introduced by the American Heart Association

in 1992 for first responders of adults in cardiac arrest in an effort to improve their survival. Statistics have shown that the quicker an adult receives defibrillation from a witnessed cardiac arrest in the presence of ventricular fibrillation, the higher the survival rate.⁷⁹ Automated external defibrillators have been successfully used in several public settings, including airport terminals,⁸⁰ airplanes,⁸¹ and casinos.⁸² These devices have been deemed safe and effective for use in children aged 1 to 8 years and are being used with increasing frequency in the pediatric population.⁷⁹

Automated external defibrillators have not always been successful in the resuscitation efforts of young athletes compared with other groups of children and adolescents who experience SCD.^{83,84} Berger⁸⁵ speculates that this may be due to the higher proportion of young athletes with HCM who have thicker hearts and/or high catecholenergetic levels during competition. This has not been substantiated in the literature at this time. Alternatively, AEDs have been successful in school settings. In 1999, Project ADAM (Automated Defibrillators in Adam's Memory) was established in southeast Wisconsin after the deaths of several high school athletes.⁸⁵ The goals of the project were to (1) educate the public about SCD and preparticipation sports screening, (2) advocate for cardiopulmonary resuscitation training prior to graduation, and (3) place AEDs in the schools in the state of Wisconsin. Since the initiation of the program, 7 lives (2 adolescents and 5 adults) have been saved. A 15-year-old female adolescent who was successfully resuscitated was found to have a CCAA that was surgically repaired and a male adolescent who was successfully resuscitated was found to have LTQS. Currently, 25% of the schools in the state of Wisconsin now have AED programs and Project ADAM is forming affiliate programs in Orlando, Florida, Atlanta, Georgia, and Philadelphia.

Patient advocacy

There are several advocacy groups that have a vested interest in SCD in children,

adolescents, and adults (Table 2).⁸⁶⁻⁸⁹ These groups are uniquely positioned to serve patients, their family members, physicians, and other health care professionals.⁹⁰ These groups have been responsible for (1) providing information on current research being conducted in the field, information about genetic testing, and importantly a list of medications that could potentially harm individuals with LQTS,⁸⁶ (2) lobbying Congress to establish the month of October as sudden cardiac arrest awareness month, the first of which was observed in October 2008,⁸⁷ (3) collaborating with researchers to provide data on exercise-related sudden cardiac arrest in the young,^{16,88} and (4) providing a quarterly literature review that contains currently available research and clinically based publications categorized by disease etiology.⁸⁹

Nursing implications

Survivors of SCD are in a precarious emotional state. They are often afraid of recurrence of SCD as well as the implementation of various treatment modalities, such as an ICD. Nurses have several obligations when it comes to caring for these individuals and their families. First and foremost, the patient and the family must be educated about the specific disease etiology that puts them at risk for SCD.⁷⁰ Fear of the unknown is a common cause of anxiety in anyone with a chronic health condition but it is heightened when SCD may be lurking in their future. Understanding the reasons why they are thought to be at risk for SCD, and whether they have experienced such an event or not, will assist them in accepting the treatment options that are undertaken by their health care providers to potentially decrease that risk. Second, nurses must understand that restriction from sports participation and other activity limitations is a significant issue that affects the health of children, adolescents in particular, directly because it is a "socially sanctioned arena" where energy can be positively channeled.⁹¹ Nurses should work diligently to help these individuals understand the reasons

Table 2. Patient Advocacy Groups

Name	Mission/purpose	Web address
Cardiac Arrhythmias Research and Education Foundation ⁸⁶	To formulate, promote, and lead initiatives to prevent sudden cardiac death due to acquired and heritable heart rhythm disorders by advocating increased support for comprehensive scientific research and clinical trials. educating patients, the public, and health care professionals to increase awareness. advancing strategies to identify, protect, and support at-risk individuals and their families.	http://www.longqt.org/
Heart Rhythm Society ⁸⁷	The Heart Rhythm Society is the international leader in science, education, and advocacy for cardiac arrhythmia professionals and patients, and the primary information resource on heart rhythm disorders. Its mission is to improve the care of patients by promoting research, education, and optimal health care policies and standards.	http://www.hrsonline.org/
Parent Heart Watch ⁸⁸	Parent Heart Watch is a national network of parents, families, and partners dedicated to reducing sudden cardiac arrest in youth. Its mission is to inform, educate, advocate, and implement nationwide programs that help achieve the mission and vision objectives.	http://www.parentheartwatch.org/
Sudden Arrhythmia Death Syndromes Foundation ⁸⁹	To save the lives and support the families of children and young adults who are genetically predisposed to sudden death due to heart rhythm abnormalities.	http://www.sads.org/

for the restrictions and assist them in finding alternative venues that can coexist with their specific disease etiologies. Third, nurses should partner with patient advocacy groups and local communities to educate the general public about SCD in the young and encourage them to support the placement of AEDs and emergency response systems in as many schools as possible. These community outreach programs are slowly, but surely having an impact on the survival of patients experiencing SCD in communities.⁹²

In summary, although SCD in the young is not entirely preventable, steps have been

taken in reducing the risk and soliciting the public to become more involved in recognizing and helping to decrease the number of youth who succumb to SCD. Nurses can have a major impact on the lives of these individuals and their families by providing as much correct information as possible regarding the diseases and treatment options, providing much-needed psychosocial support to these patients and their families, and advocating for increased public awareness regarding SCD in this patient population in an effort to increase the number of survivors.

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