American Academy of Pediatrics
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POLICY STATEMENT

Pediatric Sudden Cardiac Arrest

abstract

FREE

Pediatric sudden cardiac arrest (SCA), which can cause sudden cardiac death if not treated within minutes, has a profound effect on everyone: children, parents, family members, communities, and health care providers. Preventing the tragedy of pediatric SCA, defined as the abrupt and unexpected loss of heart function, remains a concern to all. The goal of this statement is to increase the knowledge of pediatricians (including primary care providers and specialists) of the incidence of pediatric SCA, the spectrum of causes of pediatric SCA, disease-specific presentations, the role of patient and family screening, the rapidly evolving role of genetic testing, and finally, important aspects of secondary SCA prevention. This statement is not intended to address sudden infant death syndrome or sudden unexplained death syndrome, nor will specific treatment of individual cardiac conditions be discussed. This statement has been endorsed by the American College of Cardiology, the American Heart Association, and the Heart Rhythm Society. *Pediatrics* 2012;129:e1094–e1102

SECTION ON CARDIOLOGY AND CARDIAC SURGERY

KEY WORDS

syncope, cardiovascular disease, long QT, cardiomyopathy, athlete, heart disease

ABBREVIATIONS

AAP—American Academy of Pediatrics
AED—automated external defibrillator
AHA—American Heart Association
CPR—cardiopulmonary resuscitation
CPVT—catecholaminergic polymorphic ventricular tachycardia
ECG—electrocardiography
EMS—emergency medical services
HCM—hypertrophic cardiomyopathy
LQTS—long QT syndrome
PPE—preparticipation evaluation
SCA—sudden cardiac arrest
SIDS—sudden infant death syndrome
VF—ventricular fibrillation

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INCIDENCE OF PEDIATRIC SUDDEN CARDIAC ARREST

In the United States, there is no centralized or mandatory registry for pediatric sudden cardiac arrest (SCA). Available data generally are collected through media reports, from lay SCA advocacy groups, or from peer-reviewed publications, often from major referral medical centers. Episodes of resuscitated cardiac arrest (aborted cardiac death) are even more difficult to document retrospectively. The Centers for Disease Control and Prevention has estimated that every year in the United States, approximately 2000 patients younger than 25 years will die of SCA.[1] Other older reports estimate the frequency of SCA in children and adolescents to be between 0.8 and 6.2 per 100 000 per year.[2–6] Two studies suggest that the frequency of SCA in adolescents and young adults actually may be increasing.[7,8] Although SCA occurs even at young ages and at rest, the likelihood of child and young adult SCA for those with underlying cardiovascular disease is increased by athletic participation.[9] Nonetheless, 2 studies from Maron et al[10,11] estimate fewer than 100 cases of SCA in young US competitive athletes each year. An Italian study reported a baseline incidence of SCA in young competitive athletes at 1:25 000 before implementing a national screening program.[12] Corrado et al identified a 2.5 times relative risk for SCA attributable to sports activity in adolescent and young adult athletes versus an age-matched nonathletic population,[13] related to underlying cardiac disorders.

Reporting and referral biases affect our knowledge of SCA incidence. The difficulty in determining cause of death in patients with primary cardiac electrical disorders (so-called "autopsy negative") must be acknowledged. Many of these now recognized electrical disorders have been described only recently, confounding older literature that details the cause of pediatric SCA identified at autopsy.

CARDIAC DISORDERS PREDISPOSING YOUTH TO SCA

Underlying cardiac disorders associated with pediatric and young adult SCA are listed in Table 1. In general, causes can be considered (1) structural or functional (expected to be identified with echocardiography or at autopsy); (2) primary electrical (most commonly associated with structurally and functionally normal hearts); or (3) other, including use of illicit drugs and stimulants (eg, cocaine, ephedra) or prescription medications (eg, erythromycin, ketoconazole, carbamazepine). The reader is directed to reference texts and previous publications for more detail about each of these individual conditions.[14,15]

TABLE 1 Cardiac Disorders Predisposing to Pediatric and Young Adult SCA

Structural/functional
1. Hypertrophic cardiomyopathy[a]
2. Coronary artery anomalies
3. Aortic rupture/Marfan syndrome[a]
4. Dilated cardiomyopathy or restrictive cardiomyopathy[a]
5. Myocarditis
6. Left ventricular outflow tract obstruction
7. Mitral valve prolapse
8. Coronary artery atherosclerotic disease
9. Arrhythmogenic right ventricular cardiomyopathy[a]
10. Postoperative congenital heart disease
Electrical
11. LQTS[a]
12. Wolff-Parkinson-White syndrome
13. Brugada syndrome[a]
14. Catecholaminergic polymorphic ventricular tachycardia[a]
15. Short QT syndrome[a]
16. Complete heart block
Other
17. Drugs and stimulants; some prescription medications
18. Primary pulmonary hypertension[a]
19. Commotio cordis

[a] Familial/genetic.

GENETICS OF PEDIATRIC SCA

The identification of disease-causing genetic mutations is progressing rapidly in all areas of medicine. Evaluation of large cohorts of ostensibly healthy individuals has begun to catalog the common polymorphisms and the background rate of rare genetic variants of uncertain significance within the general population. For cardiac disease, the science of genotypic evaluation has not yet advanced to the point at which genotype alone (isolated from clinical phenotypic description) can routinely and accurately risk stratify for clinical outcome. Many cardiac disorders, including hypertrophic cardiomyopathy (HCM) and the cardiac ion channelopathies, are known to be genetic.[16,17] Several studies have documented the efficacy of genetic testing of first-degree relatives of persons who have died of SCA. A 2003 study[18] reported cardiac symptoms in 27% of surviving relatives, with a 22% incidence of unexpected premature sudden death in addition to the proband in any relative and a 6% incidence of sudden death in a first-degree relative. After evaluating 49 cases of young autopsy-negative SCA, Tester and Ackerman[19] reported 17 cases with genetic/molecular evidence for long QT syndrome (LQTS) or catecholaminergic polymorphic ventricular tachycardia (CPVT) disease–causing mutations; 9 (53%) of these cases had a family history of SCA or syncope documented by the medical examiner. A personal history of syncope, seizure, or previous cardiac arrest was detailed for 7 individuals whose deaths were attributable to SCA. In a 2005 report, genetic testing established a likely cause of death in 17 of 43 autopsy-negative persons (40%). Genetic testing of family members revealed an additional 151 presymptomatic and undiagnosed disease carriers (average of 8.9 per family).[20]

Recognizing the genetic nature of many of the disorders listed in Table 1, the role of a detailed, comprehensive family history (and considering consultation with an expert in cardiac genetics) is readily apparent. The primary goal is prospective identification of any family member, even if asymptomatic, who is genotypically or phenotypically affected by a disease entity predisposing a person to SCA. A 2008 publication discusses the role of family history for evaluating cardiomyopathy and ion channelopathies predisposing people to SCA.[21] A 3-generation pedigree as a family history tool is highly effective for clinical evaluation; a family history template suggested by the US Surgeon General's Family History Initiative is available free at www.hhs.gov/familyhistory.

WARNING SIGNS AND SYMPTOMS

Although SCA may be the sentinel event, symptoms in patients with structural-functional or primary electrical disorders may, in fact, be relatively common before SCA. Often, these warning signs or symptoms may be misinterpreted or disregarded by both family members and medical personnel. These points were emphasized in a 1996 publication[22] that summarized 9 previous studies. Preceding symptoms of dizziness, chest pain, syncope, palpitations, or dyspnea and a family history of premature, unexpected sudden death were noted in 25% to 61% of the study population. Deaths were exertion-related in 8% to 33% of cases. A study of 162 young persons (15–34 years of age)[23] undergoing autopsy evaluation after SCA found 92

cases with a history of syncope/presyncope, chest pain, palpitations, or dyspnea; 26 of these subjects had a family history of SCA. In a study of natural death in people 5 to 35 years of age,[24] the most common cause of sudden death was presumed arrhythmia in those with no or minimal heart disease (29%). Eleven percent of cases were exercise-associated. A history of SCA was reported in 4.5% of first-degree relatives of the descendents. Importantly, symptoms may be nonspecific and confusing in athletes, who may overexert until physical exhaustion.

In most cases, the immediate cause of SCA is a lethal ventricular tachyarrhythmia (ventricular fibrillation [VF] or pulseless ventricular tachycardia) causing cardiac collapse. Some of these arrhythmias (eg, torsades de pointes, the characteristic tachyarrhythmia associated with LQTS) may be short lived and self-terminating, causing episodes of syncope/presyncope or episodes of seizure-like activity.[19,22–25] These neurologic signs and symptoms may direct referral to a neurologist, inadvertently misdirecting the patient away from cardiac evaluation and, thus, delaying correct diagnosis and treatment. These tachyarrhythmia-associated SCA events must be distinguished from the well-recognized but poorly understood entity called sudden unexpected death in epilepsy.[26,27] In the latter, this primary neurologic event may cause a cardiac death, mediated through abnormalities of cardiovascular autonomic function.[28] Chest pain is almost never present in patients with primary electrical disorders but is more likely in patients with cardiomyopathies,[29,30] congenital coronary artery abnormalities,[31] or aortic disease (eg, dissection or rupture associated with Marfan syndrome[32]). Other nontypical cardiac presentations also may misdirect patients to other consulting medical subspecialties. Symptoms suggestive of exercise-induced bronchospasm may be present in patients with HCM and dilated or restrictive cardiomyopathy. Cardiomyopathy-associated wheezing is attributable to decreased left ventricular compliance, mitral insufficiency, or pulmonary venous hypertension with pulmonary edema. Failure of empirical exercise-induced bronchospasm medication or normal pulmonary function testing should prompt cardiovascular evaluation. Drowning or near-drowning has been associated with LQTS and CPVT.[33,34] Approximately 5% to 10% of sudden infant death syndrome (SIDS) cases may stem from channelopathic mutations in genes associated with LQTS, Brugada syndrome, and CPVT.[35–38] Congenital deafness has been noted in some types of LQTS.[39] Patients with congenital deafness should be evaluated for LQTS if the deafness is not otherwise associated with another recognized syndrome or anomaly. Febrile seizures may be a presenting sign of children affected with Brugada syndrome.[40]

SCREENING TECHNIQUES

The role of any screening effort is to identify individuals at risk; unaffected or low-risk individuals should be cleared, and conversely, those affected should be appropriately restricted counseled, and treated. Not all SCAs can be foreseen, even in the best of circumstances. No screening protocol has yet proven to be effective in this role or validated as highly effective.

Sports Preparticipation Evaluation and Cardiovascular Risk Assessment

As noted by aforementioned studies, it is estimated that as many as half of pediatric SCA cases exhibited a personal/familial sudden death warning sign or symptom (such as previous exercise-triggered faint or family history of premature unexplained sudden death). Thus, there is an opportunity to identify individuals at risk for pediatric SCA without technology-based screening programs, such as the 12-lead electrocardiography (ECG) and echocardiography; however, despite the aforementioned data supporting the fact that preceding warning signs and symptoms may be present in many patients and families at risk for SCA, most published studies have not substantiated the efficacy of current athletic preparticipation evaluation (PPE) processes. Only 3% of 158 athletes with SCA were suspected of having cardiovascular disease using a PPE screen, leading the authors of a 1996 study to conclude that "pre-participation screening appeared to be of limited value for identification of underlying cardiovascular abnormalities."[41] The 1996 study was retrospective, and the details of the PPE questionnaire used and the adequacy of PPE were not reported. This report also predated description of some of the disease entities now known to cause pediatric SCA. More recently, an investigation from the United Kingdom concluded that family history and personal symptom questionnaire alone were inadequate for identification of at-risk patients and families.[42] The 2008 UK study used a comprehensive PPE format and trained examiners, with little reported benefit, which reveals the potential failure of a single PPE at 1 point in time.

In contrast to a single PPE initiated only before athletic participation, a more thorough cardiovascular risk-assessment process, applied throughout childhood and adolescence (the continuum of well-child care), can be provided for any patient, of any age, by any care provider (Table 2). Patient and family histories can and do change over time, necessitating an update of information for the care provider. Families should be encouraged to provide complete and

TABLE 2 Pediatric Sudden Cardiac Death Risk Assessment Form

	Yes	No
Patient history questions: Tell me about any of these in your child…		
Has your child fainted or passed out during or after exercise, emotion, or startle?		
Has your child ever had extreme shortness of breath and/or discomfort, pain, or pressure in his or her chest during exercise?		
Has your child had extreme fatigue associated with exercise (different from other children)?		
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?		
Family history questions: Tell me about any of these in your family…		
Are there any family members who had a sudden, unexpected, unexplained death before age 50 (including SIDS, car crash, drowning, others) 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: HCM		
Dilated cardiomyopathy		
Heart rhythm problems: LQTS		
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)		
Please explain more about any "yes" answers here:		
Parent signature:		
Physician signature:		
Date:		

Ask these questions (or have parents complete for your review) at periodic times during well-child visits (neonatal, preschool, before or during middle school, and before or during high school).

accurate responses concerning history. PPE screening of athletes exclusively omits >25 million US schoolchildren who do not participate in sports. Postponing cardiovascular risk assessment until a more formal high school or collegiate athletic PPE screen also will delay detection of patients and families at risk for SCA before high school. For any PPE or cardiovascular risk assessment to succeed, medical providers must be aware of sudden death warning signs and symptoms and respond appropriately with comprehensive cardiovascular evaluation, referral, treatment, and activity restrictions as appropriate. The use of competent and qualified examiners is still a necessity, but recent data reveal that 35% of states allow nonphysicians with little cardiovascular training to perform the screening evaluations.[43]

The American Heart Association (AHA) has documented a 12-element recommendation for preparticipation screening of competitive athletes.[44] Another PPE, sponsored and endorsed by the American Academy of Pediatrics (AAP) and 5 other agencies, is now widely used throughout the United States for childhood and adolescent athletic PPE.[45] This document acknowledges the wide variation in physician competencies and documentation for PPE examinations. Many states have endorsed the use of this PPE to eliminate unnecessary variability and to more effectively screen for cardiovascular risk. The updated fourth edition of this PPE form became available in April 2010.

None of the aforementioned PPEs or cardiovascular risk-assessment tools have been validated but serve as standard templates for more comprehensive screening. Likewise, no true sensitivity or specificity data exist for prediction of risk of SCA by PPE. Among the many warning signs and symptoms, the following 4 appear to represent more ominous positive responses (based on expert opinion):

1. Have you ever fainted, passed out, or had a seizure suddenly and without warning, especially during exercise or in response to auditory triggers such as doorbells, alarm clocks, and ringing telephones?
2. Have you ever had exercise-induced chest pain or shortness of breath?
3. Are you related to anyone with sudden, unexplained, and unexpected death before the age of 50?
4. Are you related to anyone who has been diagnosed with a sudden death–predisposing heart condition such as HCM, LQTS, Brugada syndrome, and so forth? (See Table 1.)

Once a cardiovascular disorder listed on Table 1 is suspected or diagnosed, referral to and management by pediatric/adult cardiologists or heart rhythm specialists experienced with the particular sudden death–predisposing heart condition is crucial.

Another important time, resource, and cost-benefit issue centers around obtaining the detailed and accurate cardiovascular risk assessment or PPE forms in the primary care office setting. This time-consuming process is currently poorly reimbursed and difficult to prioritize and validate in a busy practice.

ECG Screening

Although some data suggest that SCA screening may be enhanced with the addition of ECG, broad-scale ECG screening has not been tested or implemented in the United States. Mandatory screening of Japanese schoolchildren since 1973[46,47] has demonstrated a greater sensitivity of ECG versus history and physical examination. Competitive Italian athletes undergo required PPE and ECG, with ECG reportedly demonstrating 77% greater power to detect HCM

than history and physical examination alone.[2] Italy also has reported a newborn ECG screening program to identify infants at risk for SIDS secondary to abnormal cardiac repolarization.[48] For Olympic athletes, the International Olympic Medical Committee issued a screening protocol including ECG in 2004.[49] A 2005 European Society of Cardiology consensus statement on cardiovascular preparticipation screening of young competitive athletes recommends 12-lead ECG in addition to focused history and physical examination.[50] Some US studies have suggested that ECG screening may be cost-effective on the basis of estimated cost per year of lives saved.[51,52]

The 2007 AHA scientific statement/screening guidelines[44] (coauthored by S.B. and M.J.A.) did not recommend standard ECG assessment, however, citing false-positive and false-negative results, cost-effectiveness, feasibility, and medicolegal concerns. Wide-scale ECG screening would require a major infrastructure enhancement not currently available in the United States. Recent reassessment of ECG "normal" values has helped to decrease false-positive findings.[53] Competitive athletes are known to have unusual but occasionally benign ECG findings, consistent with "athlete's heart," that must be differentiated from ECG findings attributable to pathologic conditions.[54]

The role of routine ECG screening in the United States to prevent SCA is not settled and will require more data and debate. Readers are referred to recently published debates of the subject for further details.[55,56]

Molecular Autopsy

The genetic nature of many cardiac ion channelopathies predisposing youth to pediatric SCA is being defined rapidly.[17] When children die suddenly, there may be no previous evaluation or diagnosis. Conventional autopsies often fail to identify a condition responsible for sudden death. These autopsy-negative cardiac conditions have previously defied complete definition. As already described, complete evaluation of a child who died of SCA through detailed clinical and targeted genetic testing of immediate family members may identify specifically the cause of SCA and direct appropriate care and genetic counseling to surviving family members. The cardiac channel postmortem genetic analysis (also known as "molecular autopsy")[57] remains a research test but soon may evolve into a standard clinical practice. Unfortunately, current standards of care for autopsy do not yet ensure that a postmortem sample suitable for DNA analysis is retained. Further, despite the evidence that approximately 25% to 35% of autopsy-negative sudden unexplained death is channelopathic, health insurance companies currently do not accept responsibility for molecular autopsy of the deceased in the United States. The cost would befall the medical examiner and, ultimately, the community; however, far more expensive testing of all first-degree surviving family members currently is used clinically and reimbursed. An important next step will be the development of guidelines at a public health level for postmortem genetic testing.

PRIMARY PREVENTION OF SCA

Primary prevention of SCA depends on patient diagnosis, specific etiology, and etiology-specific treatment. Treatment options include but are not limited to medical therapy, device therapy (eg, pacemakers, internal cardioverter defibrillators), activity-restriction guidelines, avoidance of certain classes of medications, and family emergency preparedness. The details of primary prevention, given that they are etiology specific and prescribed by a consulting cardiologist, are beyond the scope of this policy statement.

SECONDARY PREVENTION OF SCA

When SCA primary prevention strategies (ie, patient identification, treatment, activity restriction, and counseling) have failed, SCA still may occur, and secondary prevention (resuscitative) efforts are required. The AHA has proposed a "chain-of-survival"[58] beginning with early symptom recognition and 911 emergency medical services (EMS) contact, followed by effective bystander cardiopulmonary resuscitation (CPR), early defibrillation, and finally, advanced hospital care. The published outcomes for out-of-hospital pediatric cardiac arrest are dismal; survival to hospital discharge occurs in approximately <10% of children, and many have severe neurologic sequelae.[59–63] Poor outcomes may be related to prolonged periods of no cardiac output, in part because many out-of-hospital arrests are unwitnessed, and only approximately 30% of children received bystander CPR[61] (note also that bystander CPR more than doubles patient survival rates[64]).

Bystanders report that they do not perform CPR because of panic or fear of failure[65] and unwillingness to perform mouth-to-mouth rescue breathing. Recent studies suggest that "compression-only" CPR may be more effective than standard CPR with ventilation,[66,67] by using faster (approximately 100 per minute) and deeper compressions, in adults for witnessed nonasphyxial arrest (arrest not secondary to, for example, drowning, hanging, or carbon monoxide poisoning). To date, there are no pediatric studies with respect to compression-only CPR. Because pediatric patients are more likely to experience respiratory arrests, compression only may not be suitable. Two studies report VF as the initial rhythm in 19% to 24% of out-of-hospital pediatric cardiac arrests, excluding deaths attributable to SIDS.[68,69] VF and ventricular tachycardia generally have been considered more favorable initial SCA rhythms than

either asystole or pulseless electrical activity, with a higher rate of survival to hospital discharge, when prompt defibrillation (termination of VF) and return of an organized perfusing rhythm is achieved. As part of the chain of survival, public access defibrillation using automated external defibrillators (AEDs) has a prominent role.[70] Data from witnessed VF arrest in adults show that appropriate use of AEDs can lead to long-term survival rates >70%.[71,72] AEDs have now been recommended for children younger than 8 years,[73,74] with still insufficient scientific evidence to warrant official recommendation for or against AED use in children aged 1 year or younger. A 2007 AAP policy statement addressed current pathophysiology of VF and recommendations for AED use in children; readers are referred to this publication for further detail.[75]

SCHOOL AED PROGRAMS

The average school-aged child spends 28% of the day and 14% of his or her total annual hours in school.[76] In addition, adults (parents, grandparents, teachers, staff, and visitors) crowd our schools. As an area of higher traffic, schools have become sites for implementation of AED programs. In 1 report, 67% of schools activate EMS for an emergency involving a student, and 37% activate EMS for an emergency involving an adult.[76] A 2007 report detailed a 16-year history of SCA in Seattle city and King county schools, providing a framework for reasonable and rational school-based emergency programs.[77]

A growing number of states have mandated school AED programs. The cost-effectiveness of school AED programs has been reported by Berger et al.[78] Key components for a comprehensive school-preparedness program include education and all-staff awareness, knowledge and application of effective bystander CPR techniques, implementation of a lay-rescuer AED program, and written emergency action plans,[79] with all steps reinforced with effective communication throughout the school campus and periodic practice drills. Current principles guiding this recommendation for schools, primary clinicians, and school physicians have been detailed in the AAP policy statement "Medical Emergencies Occurring at School."[80] At this time, there are no published data to support the efficacy of home AEDs.[81]

RECOMMENDATIONS FOR PEDIATRIC CARE PROVIDERS

Evidence-based recommendations frequently are designated as class I, II, or III, indicating the supporting level of evidence. For pediatric SCA, the level of evidence does not permit a meaningful use of this terminology. This statement has been endorsed by the American College of Cardiology, the American Heart Association, and the Heart Rhythm Society.

All steps in the primary and secondary SCA-prevention strategies should be optimized if pediatric SCA is to be prevented.

RECOMMENDATIONS

Important steps for consideration include:

1. Recognize the warning signs and symptoms of SCA, including those that may "misdirect" initial evaluation to noncardiac specialties and, thus, delay correct diagnosis.
2. Understand the role of comprehensive and accurate family history and pedigree for preventing SCA stemming from inherited cardiac genetic disorders.
3. Use standardized PPE forms and processes to minimize unnecessary variation.
4. Ensure that identified patients and/or families with known or suspected cardiac disorders are referred to a pediatric cardiac center for further comprehensive evaluation and management. Appropriate secondary testing may include ECG, echocardiography, exercise testing, or genetic testing, as indicated.
5. Advocate for autopsy evaluation by a medical examiner familiar with rarely encountered heritable cardiac diseases causing SCA when pediatric SCA occurs. Procurement of and retention of DNA-bearing tissue for subsequent molecular autopsy should be encouraged for autopsy-negative cases.
6. Support education programs for effective bystander CPR and appropriate AED use.
7. Support development of effective school emergency response programs.
8. Consider participation in school emergency response programs as a medical director.
9. Support efforts to mandate a central registry for pediatric SCA as a reportable event.
10. Support recommendation for evidence-based evaluation of national screening processes and programs.

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