



Sudden Death in the Young: Information for the Primary Care Provider

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There are multiple conditions that can make children prone to having a sudden cardiac arrest (SCA) or sudden cardiac death (SCD). Efforts have been made by multiple organizations to screen children for cardiac conditions, but the emphasis has been on screening before athletic competition. This article is an update of the previous American Academy of Pediatrics policy statement of 2012 that addresses prevention of SCA and SCD. This update includes a comprehensive review of conditions that should prompt more attention and cardiology evaluation. The role of the primary care provider is of paramount importance in the evaluation of children, particularly as they enter middle school or junior high. There is discussion about whether screening should find any cardiac condition or just those that are associated with SCA and SCD. This update reviews the 4 main screening questions that are recommended, not just for athletes, but for all children. There is also discussion about how to handle post-SCA and SCD situations as well as discussion about genetic testing. It is the goal of this policy statement update to provide the primary care provider more assistance in how to screen for life-threatening conditions, regardless of athletic status.

INTRODUCTION

There is a growing movement to identify pediatric and young adult athletes who may be at risk for sudden cardiac arrest (SCA) or sudden cardiac death (SCD) during sports participation.¹⁻⁵ The sudden death of a young athlete is always tragic for the family and community. However, the sudden death of a young nonathlete is no less tragic. In today's society, the nonathlete is much less visible because of the great attention paid to athletics in the lay press, which creates the perception that only athletes have an increased risk of sudden cardiac events. SCA and SCD in young people have been addressed in several articles that have focused efforts toward disease recognition as well as

abstract

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prevention.⁶⁻¹¹ There have been numerous studies addressing preparticipation screening, including whether electrocardiography (ECG) or other noninvasive tests should be added.^{2-4,12,13} Currently, several groups are studying the question of the use of preparticipation ECG; to date, none have published conclusive data on its overall effectiveness, practicality of implementation, or cost.¹⁴

One of the most important people in both primary and secondary prevention models is the primary care provider (PCP), who manages children from infancy into late teenage years or even young adulthood and has a long-standing relationship with the child, family, and community at large. PCPs are involved with school preparticipation screening and are often the first called when a cardiac symptom or cardiac arrest occurs.

The purpose of this article is to provide PCPs with a strategy for screening, evaluation, and management of risk of SCA and SCD in the young with practical and updated information.

As in the 2012 policy statement, “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.”⁸

STRATEGY FOR SUDDEN DEATH PREVENTION

This policy statement proposes that the same screening detail that is used for athletes should also be applied to the nonathlete.

Figure 1 demonstrates an encounter of a pediatric patient with the PCP for a routine visit or for new, concerning symptoms.

The PCP encounter should ultimately separate patients into 2 basic groups, those with identifiable or suspicious risk factors for SCA or SCD, to be discussed in a later section, and those without risk factors. For those with risk factors, referral to a pediatric cardiologist or electrophysiologist is the next appropriate step to initiate a comprehensive cardiovascular evaluation appropriate for the presenting risk factors. There are patients who, despite the best screening efforts, could still experience a SCA; therefore, a secondary prevention plan is important.

Multiple studies have led to current resuscitation methods, such as the American Heart Association (AHA)’s Basic Life Support, Pediatric Advanced Life Support, and Advanced Cardiac Life Support, featuring the “chain of survival,” with revisions made every few years.¹⁵ Although out-of-hospital cardiac arrest survival statistics remain dismal, there has been improvement in survival, most likely a result of an increase in lay rescuer cardiopulmonary resuscitation (CPR) education, an increase in recognition of cardiac arrest, and an increase in willingness to intervene by lay rescuers who have learned to perform high-quality CPR and automated external defibrillator (AED) use, assisted by an increase in public access to AEDs.¹⁶ The number of lay people with life support training generally remains low.¹⁷ This highlights the important role for the PCP to be a community advocate for more Basic Life Support training.

THE PCP’S ROLE IN PRIMARY PREVENTION

The difficult task of identifying those at risk for cardiac events often begins with the PCP, including physicians, physician assistants, and nurse practitioners,

via routine physical examination or when addressing specific symptoms. Although there is no one-size-fits-all screening method to identify those at risk, it is helpful for the PCP to have an understanding of the common conditions that put young patients at risk for SCA and SCD.

Cardiomyopathies

A primary cardiomyopathy is usually associated with an anatomically normal heart with abnormal myocardial cellular structure or function that can affect both systolic and/or diastolic function. The World Health Organization and International Society and Federation of Cardiology Task Force recognize 5 basic forms of cardiomyopathy¹⁸:

1. Dilated cardiomyopathy: enlarged, dilated left and/or right ventricle with or without decreased systolic function.
2. Hypertrophic cardiomyopathy (HCM): abnormally thickened ventricular myocardium without cause (eg, hypertension, coarctation, aortic stenosis, etc). HCM is reported as the most common cause of SCA and SCD in young athletes.¹⁹
3. Restrictive cardiomyopathy: normal to thickened ventricular walls and normal ventricular size with impaired diastolic function and often with dilated atria.
4. Arrhythmogenic cardiomyopathy (includes arrhythmogenic right ventricular cardiomyopathy [ACM]): enlarged, dilated right ventricle with or without decreased systolic function often associated with frequent arrhythmias (can be seen in the left ventricle as well).
5. Unclassified cardiomyopathies: this includes left ventricular noncompaction (the left ventricular myocardium is abnormal with hypertrabeculation and crypt

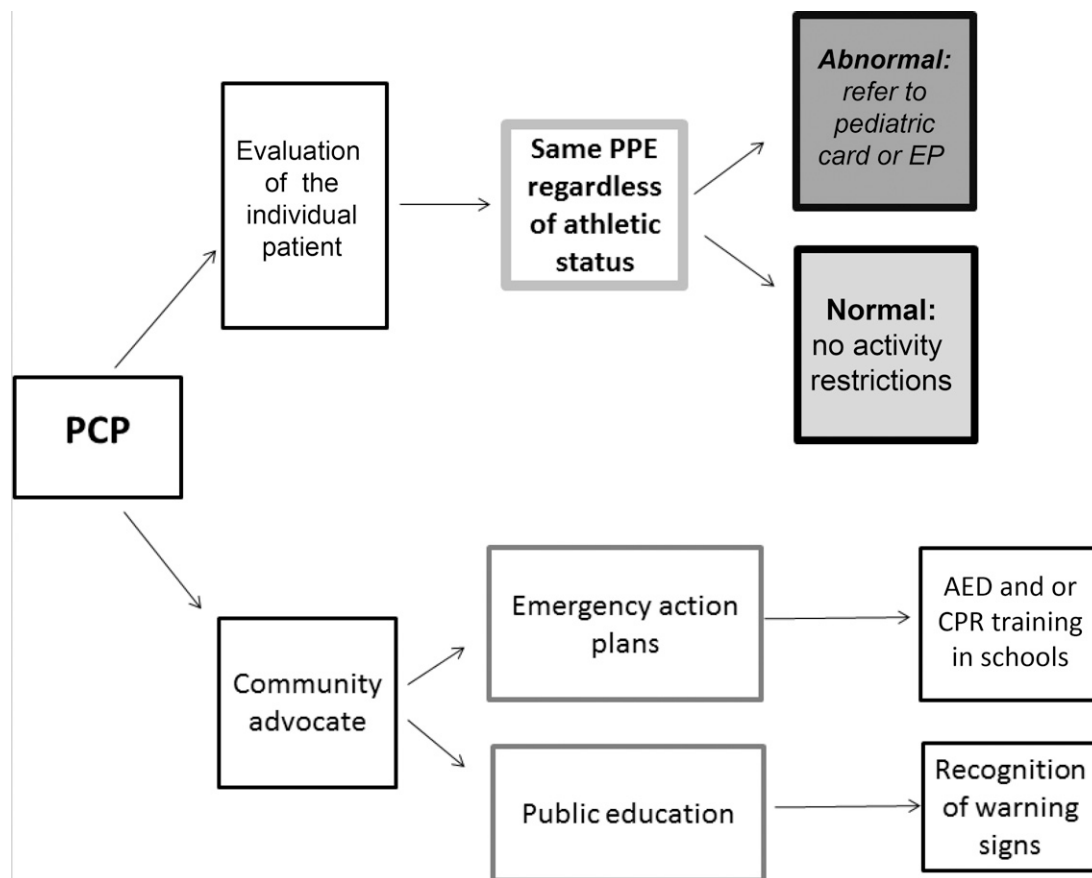


FIGURE 1 Flowchart revealing ideal role of the PCP in prevention of SCD in the young. EP, electrophysiologist.

formation of the left ventricular wall with thickened, normal, or thinned and dilated myocardium with or without impaired systolic function).

The clinical features of each type of cardiomyopathy are displayed in Table 1. Morphologic differences are significant between each type of cardiomyopathy, and, therefore, imaging (echocardiography, MRI, etc) is advised. The potential for life-threatening arrhythmias is a unifying factor for all these cardiomyopathies.

Channelopathies

Channelopathies are generally identified in patients who otherwise have normal cardiac anatomy and function. The defect involves the ion

channels in the cardiac cell membrane or in intracellular proteins that interact with ion transport and may result in identifiable abnormalities on the ECG. Imaging is not helpful in diagnosing a channelopathy except to exclude cardiomyopathy as an etiology for a cardiac event. The primary channelopathies include the following:

1. Long QT syndrome (LQTS): prolongation of the corrected QT interval (QTc) with abnormalities in T-wave morphology, some of which are associated with specific genotypes. Sudden infant death syndrome (SIDS) may be attributable to LQTS in approximately 10% of cases.²⁰
2. Short QT syndrome: extremely rare condition with abnormal

shortening of the QTc with prominent and peaked T waves.

3. Brugada syndrome (BrS): associated with a coved and elevated ST elevation in ECG leads V1 and V2. Specific Brugada T-wave morphologies may indicate an elevated risk of cardiac arrest.
4. Catecholaminergic polymorphic ventricular tachycardia (CPVT): resting ECGs in these patients are almost always normal. CPVT is mostly identified with exercise testing that results in increased ventricular ectopy and even polymorphic ventricular tachycardia (VT).
5. Idiopathic ventricular fibrillation (IVF): patients presenting with ventricular fibrillation (VF) in whom known cardiac, respiratory, metabolic, and toxicological

TABLE 1 Listing of the 5 Classification Groups and Clinical Features of Cardiomyopathies According to the World Health Organization

Cardiomyopathy Class	Age at Presentation	Presentation	Family History	Genetic Inheritance	ECG	Echocardiogram Findings	MRI Findings	Making the Diagnosis	Risk for SCA or SCD
DCM	Any age, including fetal	Can be incidental finding; heart failure; poor cardiac output; failure to thrive; arrhythmias; LMNA mutation-associated DCM can also present with conduction disease. ⁶⁷	Inherited DCM can exist in ~50% of those presenting before 18 y of age. ⁶⁷	For most familial DCM, inheritance is AD, but X-linked inheritance is also common. ⁶⁷ LMNA-positive patients have autosomal dominant inheritance. Pediatric patients have a higher incidence of mitochondrial or metabolic-based DCM that are autosomal recessive.	Can be normal; AV block; BBB; atrial or ventricular ectopy; low-voltage QRS.	Dilated LV with mildly to severely reduced function; mitral regurgitation.	Consistent with echocardiogram findings.	Clinical symptoms and/or echocardiogram findings of LV dilation with reduced function; genetic testing is helpful when the family history suggests familial DCM.	Variable depending on genetic mutation (higher with LMNA mutation), degree of myocardial involvement, and presence of arrhythmias; lower incidence of SCD than other cardiomyopathy forms. ⁶⁸ Higher risk with poorer LV function.
HCM	Any age, including fetal	Incidental; heart murmur; anginal chest pain; cardiac arrest.	Often others in family have known HCM or will subsequently be found to have HCM.	For all sarcomeric forms of HCM, inheritance is AD; Danon syndrome (storage disease phenocopy) is X linked.	Can be normal, usually has LVH and/or RVH, sometimes with very large R or S waves; LAD: ventricular arrhythmias.	LV wall thickness > 14 mm or 2 SDs for wt; reduced LV cavity size; longer-than-normal anterior mitral valve leaflet; abnormal papillary muscles; resting and/or dynamic LVOT obstruction; MR.	Consistent with echocardiogram findings but can reveal mitral valve morphology more precisely and can be used for detecting late gadolinium enhancement that is associated with ventricular arrhythmias. ⁶⁹⁻⁷²	Echocardiogram and/or MRI findings consistent with HCM; genetics are helpful to identify sarcomeric from other forms including storage disease; can help with prognosis and can help with identification of family members.	In adults, risk is based on established risk factors, including previous SCA, LV thickness ≥30 mm, abnormal BP response to exercise; family history of SCA; LGE > 15%; VT on Holter. In children, risk factors include those presenting in infancy or with inborn errors of metabolism. ⁷³ Restrictive physiology increases risk of death or transplant; increased LV thickness and LGE correlates with VT. ⁷¹
RCM	Any age	Heart failure; poor cardiac output; fatigue.	Less than half have family history, although there can be some crossover with family members with HCM.	Shares some similar gene mutations as HCM; inheritance is AD.	Can have findings similar to HCM but may be normal as well.	Normal ventricular systolic function with dilated atrial; some hypertrophy may be present.	Evidence of diastolic dysfunction.	Echo criteria indicating abnormal diastolic function along with symptoms of heart failure; some HCM patients have RCM features. Genetic testing can be supportive. Several HCM gene mutations can be seen with RCM.	Significant with generally poor prognosis; two-year survival of <50%. ⁷⁴

TABLE 1 Continued

Cardiomyopathy Class	Age at Presentation	Presentation	Family History	Genetic Inheritance	ECG	Echocardiogram Findings	MRI Findings	Making the Diagnosis	Risk for SCA or SCD
ACM	Preadolescence ⁷⁵	Incidental; syncope, palpitations, SCA.	Possible to have other family members with the same diagnosis.	Nearly all are AD.	Inverted T waves in the right precordial leads beyond V1; ε waves; LBBB PVCs or VT; Prolonged S waves in V1-V3.	Often normal but may reveal dilated RV, abnormal aneurysmal areas of RV, MRI often reveals more detail.	Fibrotic replacement of ventricular myocardium; aneurysmal areas of free wall; possible LV involvement.	Needs to meet specific diagnostic criteria per ACM task force ⁷⁶ ; MRI alone will not satisfy task force criteria; genetic testing is helpful when positive.	Significant, even in children; disease is progressive and VT from multiple foci can develop.
LV Noncompaction (LVNC)	Any age including fetal	Incidental; murmur on examination; heart failure; SCA.	Present in 30% of first-degree relatives of index cases. ⁷⁷	For identified gene mutations inheritance is autosomal dominant; yield on genetic testing is currently low.	Usually abnormal with LVH, key finding is the T-wave abnormalities; QT prolongation, or preexcitation ⁷⁷ ; arrhythmias more common with decreased LV function.	Often normal but may reveal dilated RV, abnormal aneurysmal areas of RV, MRI often reveals more detail.	Consistent with echo findings, the noncompaction-to-compaction ratio of the LV myocardium should be $\geq 2.3:1$; LV function can be assessed. ⁷⁹	Fulfillment of echo and/or MRI criteria; genetic testing when results are positive is helpful but negative-result genetic testing does not diminish diagnosis from task force criteria.	In part dependent on LV function but SD has been reported with normal LV function. ^{80,81}
Normal variant: Athlete's heart	Teenage and up; has been reported in preadolescent age as well. ⁸²	During screening evaluation or for evaluation of potential cardiac symptoms.	None.	None.	Some benign ECG changes can be seen. Criteria have been modified for use in the athlete. Changes can include sinus brady, first-degree AV block, early repolarization, isolated voltage criteria for LVH, LAE, RAE, LAD, RAD. ⁸³	LV cavity dilation, normal diastolic properties, wall thickness up to 14 in male individuals (with rare exception in African Americans); female individuals with athlete's heart never have wall thickness >11 mm; rowing and cycling athletes tend to have thicker LV dimensions. ⁸⁴	Similar to echo dimensions with LV dilation and limits on LV wall thickness; will not see late gadolinium enhancement.	ECG and echo criteria have been established to help differentiate HCM from athlete's heart. In borderline cases, a period of 3-6 mo of deconditioning may be needed. Those with athlete's heart will have significant return toward normal dimensions whereas HCM patients will not. ⁸⁵	For true athlete's heart that has excluded HCM, no increased risk above the general population.

AD, autosomal dominant; AV, atrioventricular; BBB, bundle branch block; BP, blood pressure; DCM, dilated cardiomyopathy; LAD, left axis deviation; LAE, left atrial enlargement; LBBB, left bundle branch block; LGE, late gadolinium enhancement; LMNA, lamin A/C gene; LVOT, left ventricular outflow tract; LV, left ventricle; LVH, left ventricular hypertrophy; MR, mitral regurgitation; MRI, magnetic resonance imaging; PVC, premature ventricular contraction; RAD, right axis deviation; RAE, right atrial enlargement; RV, right ventricle; RVH, right ventricular hypertrophy.

etiologies have been excluded through clinical evaluation. The term IVF is used when the cardiac arrest remains unexplained despite this investigation.

Table 2 demonstrates the features of the most common channelopathies to help discern the characteristics of each. When LQTS or BrS have been diagnosed or suspected, it is important that any new medications, such as antibiotics, antifungal agents, or stimulants for attention-deficit/hyperactivity disorder be checked for potential contraindications in these disorders (for LQTS, use <https://Crediblemeds.org>, and for BrS, use www.brugadadrugs.org/avoid/). The PCP should be aware that, for patients with BrS, fever can trigger cardiac events.

Congenital Heart Disease

Patients with congenital heart defects, including those that have been surgically repaired or palliated, are at risk for arrhythmias. Risk factors in this population are often a result of scarring from surgery, ongoing hemodynamic abnormalities, residual lesions, or decreased ventricular function.^{21,22} The most common association of SCA and congenital heart disease is VT. However, atrial arrhythmias can also cause SCA or SCD if the tachycardia rate is fast enough and rapid atrioventricular conduction occurs.

Wolff-Parkinson-White Syndrome

Wolff-Parkinson-White syndrome (ventricular preexcitation) on the ECG indicates there is at least 1 accessory pathway that conducts antegrade from atrium to ventricle. These pathways are most commonly noted for causing supraventricular tachycardia. Rarely, atrial fibrillation in the presence of Wolff-Parkinson-White syndrome can result in VF as a result of rapid conduction of atrial

impulses down the accessory pathway to the ventricles. Criteria based on adult studies define pathways as high risk depending on how rapidly the pathway can conduct.²³⁻²⁶

The previous theory that patients with intermittent preexcitation on ECG would be at low risk for SCA or SCD does not seem to hold true for symptomatic pediatric patients.²⁷ Consultation with a pediatric electrophysiologist should be considered in all cases of Wolff-Parkinson-White pattern on an ECG, regardless of the presence or absence of symptoms, to aid in risk stratification and potentially consider a curative ablation procedure.

Commotio Cordis

Commotio cordis is the term applied to a sudden impact to the chest that causes VF and results in SCA or SCD without evidence of cardiac damage.²⁸ Commotio cordis is, perhaps, one of the most concerning of all sudden death conditions because it occurs in children with completely normal hearts from both a structural and molecular or ion channel standpoint. The impact is most often from a blunt object such as a ball, fist, elbow, or helmet.

Baseball is the sport with the highest frequency of commotio cordis events. For primary prevention, there is some evidence that some chest protectors may reduce the incidence of commotio cordis.²⁹ If no cardiac disease is identified in survivors of commotio cordis after a full cardiac evaluation, they can return to sports participation.³⁰ Prompt recognition of commotio cordis with initiation of CPR and defibrillation is important for survival, although some commotio cordis victims do not survive despite prompt initiation of resuscitation.

Anomalous Coronary Arteries

In multiple studies of the causes of sudden death, an anomalous coronary artery is second only to HCM.¹⁹ Outside the neonatal period, when anomalous left coronary artery from the pulmonary artery is usually identified, detection of an anomalous coronary artery can be difficult because it is rare and often has no symptoms until presenting with SCA, usually in the teenage years. A high index of suspicion is advised for patients with syncope or atypical chest pain (Table 3). Typical pediatric chest pain most commonly represents musculoskeletal pain. Atypical chest pain is pain that raises alarm for an underlying cardiac cause and is not the usual or typical pain.

Echocardiography can often be used to identify the abnormal origins or course of the proximal coronary arteries, but computed tomography scan, MRI, or coronary angiography may be more definitive. ECG in a neonate with anomalous left coronary artery from the pulmonary artery will usually have deep and wide Q waves in leads I and aVL. For other coronary anomalies, the ECG is typically normal at rest. Treatment is surgical unroofing or reimplantation of the anomalous coronary.

Aortopathies

Patients with aortopathies, such as Marfan syndrome, familial thoracic aortic aneurysm and dissection, bicuspid aortic valve with aortic dilation, Loeys-Dietz syndrome, and Ehler-Danlos syndrome are at increased risk of aortic dilation and dissection. Patients frequently have no symptoms, but often there is a family history of aortic dilation or dissection. Aortic rupture or dissection accounts for 2% of sudden deaths in athletes.¹³ There is evidence that isometric exercise, exercise that uses Valsalva maneuver, or sudden increases in blood pressure place an extraordinary wall stress on the aorta

TABLE 2 Listing of Common Channelopathic Conditions and Clinical Features

Channelopathy	Age at Presentation	Presentation	Family History	Genetic Inheritance	EKG or Holter	Echocardiogram Findings	Stress Test Findings	Making the Diagnosis	Risk for SCA or SCD
LQTS	Any, including fetal.	Incidental detection on ECG; syncope; atypical seizures; cardiac arrest.	Variable penetrance; first-degree relatives tend to have shorter QTcs and possibly less risk of events. ⁸⁶	Mostly autosomal dominant; penetrance is variable.	Prolonged QTc >460–470 in male individuals and >470–480 in female individuals with abnormal T waves; T-wave morphology can be dependent on genotype. QTc can be variable on multiple ECGs in the same patient and may have atrial arrhythmias, including atrial fibrillation.	Normal.	Some differences by genotype; long QT 1 shows QTc prolongation with exercise that sustains into recovery, whereas LQT2 shows an abnormal shortening at peak exercise, followed by abnormally long QTc in recovery. ⁸⁷ LQTS patients may not always achieve the predicted maximum heart rate.	A single ECG cannot always rule in or out LQTS. Often, multiple ECGs or stress testing + or – epinephrine challenge are needed. Holter monitoring is not ideal for diagnosis but can be supportive for obvious QTc prolongation and/or the presence of ventricular ectopy. Sinus brady can also be present.	(1) Related to QTc duration, genotype, past history of symptoms, and sex of the individual, with male individuals at greater risk when young and female individuals at greater risk after teenage years. ^{88,89} (2) Risk of positive-genotype patients with normal QTc is significantly lower, but higher than genotype-negative family members. ⁹⁰ Caution is needed in prescribing medicine that could prolong the QTc; (3) SIDS cases attributable to LQTS ~10%. ²⁰ ; (4) Previous syncopeal event is associated with increased risk. ⁹¹ Significant for Gollob score >5. Risk is 2.6-fold greater for QTc <300. ⁹⁵ Reliability of Gollob score in question based
Short QT syndrome	Rare condition; presents at any age including infancy ⁹¹ ; most common in adolescence to 30s. ⁹²	Most common presentation is cardiac arrest or syncope. ⁹² Significant recurrence rate in those already with SCA or syncope.	Present in 44% of familial kindreds. ⁹²	Autosomal dominant; penetrance favors male individuals. ⁹²	Short QTc ≤340 ms with tall peaked T waves or <360 ms with pathogenic mutation, family history of SQTs, family history of SD in age ≤40, survival of VT or VF ⁹² ; may	Normal anatomy; decreased LV function reported in SQTs patients. ⁹⁵	Maximum HR may be less than normal. QTc does not shorten and approaches normal at peak exercise. ⁹¹	Gollob SQTs Diagnostic Criteria Score: high probability for score ≥4. ⁹⁴	Significant for Gollob score >5. Risk is 2.6-fold greater for QTc <300. ⁹⁵ Reliability of Gollob score in question based

TABLE 2 Continued

Channelopathy	Age at Presentation	Presentation	Family History	Genetic Inheritance	ECG or Holter	Echocardiogram Findings	Stress Test Findings	Making the Diagnosis	Risk for SCA or SCD
BrS	Predominance in male individuals; prevalence is high in Southeastern Asia; can be at any age but typically in the 4th or 5th decades. ⁹⁶	Incidental detection on ECG is common. Cardiac arrest at night is common. ⁹⁶ Syncope; can be asymptomatic with incidental discovery. Nocturnal agonal respiration; chest discomfort; fever can induce Brugada ECG changes and cardiac arrest. ^{97,98}	History of SCD in 26% of affected patients with BrS. For asymptomatic patients, a type 1 spontaneous ECG pattern was seen in 19%. ⁹⁹	Mostly autosomal dominant penetrance favors male individuals. There are 23 genes associated with BrS but genetic testing yield remains low. ¹⁰⁰	ST elevation in V1-V2 classified into classes I, II, or III; may have sinus node dysfunction and atrial arrhythmias. ⁹⁶	Normal.	Exercise effects in BrS are uncertain. Stress testing of patients with BrS revealed VT, PVCs, and ST elevation in 57% with 5 revealing BrS ECG pattern. ¹⁰¹	BrS diagnostic criteria based on ECG ST segment morphology. Drug challenge to induce type 1 pattern. Risk is highest in those with spontaneous type I BrS pattern on ECG and syncope (10% of patients with BrS). ^{98,102} Fever increases risk of SCA. ⁹⁸ Antipyretics are indicated and an important treatment in patients with BrS.	on report by Mazzanti. ⁹²
CPVT	Mostly children and adolescents but can be any age.	Often can be delayed because of normal ECG ¹⁰³ ; can present with cardiac arrest as the first symptom in 35% to 38% of patients. ¹⁰⁴	Variable penetrance with 50% of family relatives with the CPVT phenotype. ¹⁰⁵ Prediction difficult; medical treatment of asymptomatic gene-positive patients may be recommended.	Autosomal dominant for RYR2 and KCNJ2 mutations (60% and 2%, respectively, of CPVT) and autosomal recessive for CASQ2 (2%). ^{106,107}	Normal baseline ECG; Holter can reveal ventricular arrhythmias, including bidirectional VT; which may have atrial arrhythmias. ¹⁰⁴	Normal.	Induction of ventricular arrhythmias, including PVCs, ventricular couplets, and/or nonsustained VT in 77%. ¹⁰⁸	Stress testing and Holter monitor will frequently reveal increased ventricular ectopy or even VT with increased sinus rate; the presence of bidirectional VT is consistent with CPVT. Sinus bradycardia can also be present.	Significant: initial presentation is cardiac arrest in 33% to 38% of patients. ¹⁰⁴
IVF	Average age reported as 35-40 y ¹⁰⁹ but can occur in younger patients ¹¹⁰ and even infants. ¹¹¹	Syncope or cardiac arrest; frequently during athletic activity ¹¹² ; may be difficult to differentiate from CPVT.	van der Werf revealed 20 relatives of 83 IVF probands (4.9%) of first-degree relatives of IVF patients died, but only 2 were <50 y old. ¹¹³	Limited data but appears genetic related; RYR2 implicated as autosomal dominant. ¹¹²	Normal.	Normal.	Normal.	Difficult but mostly diagnosis of exclusion after documented VF in absence of other channelopathies in the presence of normal cardiac anatomy and function.	Limited in pediatric age; Stefanelli et al ¹¹⁰ reported 1 in 4 surviving IVF patients with appropriate shocks from an ICD.

HR, heart rate; LQ12, long QT 2; LV, left ventricle; PVC, premature ventricular contraction; SQTs, short QT syndrome.

TABLE 3 List of Symptoms Differentiating Between Typical (Benign) Chest Pain and Atypical Chest Pain

Features of "Typical" Chest Pain	Features of "Atypical" Chest Pain
Sharp	Pressure in sternum or left chest
Focal "it hurts right here!"	May radiate to neck or left arm
Brief	Associated diaphoresis
Changes with position	Associated dyspnea
Right sided (could be on left)	Associated nausea
Changes with breathing	Associated syncope
Tenderness can be elicited with palpation or pressure over the area	Onset with exercise, straining, or stress (unless features of typical chest pain are present)

and should be avoided.³¹ Collision sports are to be avoided.

WHAT IS THE GOAL OF CARDIAC SCREENING?

The goals of any screening program need to be clarified as to whether they are expected to identify any heart condition, regardless of how significant, or only target those defects associated with SCA and SCD. A thorough personal history, family history, and physical examination are the cornerstone for screening for all children and youth. The AHA recommends a 14-point history and physical screening for athletic participation commonly known as the preparticipation evaluation (PPE) (Table 4).

A positive response to any of the 14 points may prompt the need for cardiovascular evaluation at the discretion of the PCP. This screening has a tendency to detect cardiac problems regardless of their significance. ECG screening has been shown to detect some cardiac lesions that pose a risk for SCA and SCD as well as others that do not pose a risk of SCA and SCD or require restriction from athletic participation.^{32,33} There are still limitations because normal resting ECGs would also fail to detect life-threatening cardiac conditions, such as CPVT.

It Is Recommended That the Focus of Screening Should Be on Sudden Death Prevention and Identification of Those at Risk for SCA and SCD

In a 2012 policy statement, the American Academy of Pediatrics

(AAP) recommended 4 questions directed toward SCA and SCD detection for which a positive response suggested an increased risk for SCA and SCD.⁸ Similar to the AHA screening question tool, the 4 questions in the AAP policy statement are based on expert opinion. In contrast to the AHA tool, the AAP tool is intended to be used in all children regardless of athletic participation. Modifications have been made to these 4 questions with wording that can be directly applied to a family questionnaire. PCPs, at their discretion, may find a positive response to be a significant cue to perform a cardiovascular evaluation. The fifth edition of the AAP publication *Preparticipation Physical Evaluation* noted the AAP recommends an annual comprehensive health supervision visit from ages 6 to 21 years by physicians, nurse practitioners, or physician assistants with the clinical training outlined by state law. The goal of integrating the PPE into the health care home may be more easily achieved if the PPE portion of the examination is addressed every 2 to 3 years, rather than annually, to allow a different focus for evolving child and adolescent risk at each visit.³⁴ It is recommended that SCA and SCD screening should be performed for all children (athlete or not) at the same time as the PPE examination or at a minimum of every 3 years or on entry into middle or junior high school and

into high school. Depending on family and PCP concerns, more frequent screening may be appropriate. The modified 4 questions, also based on expert opinion, are as follows:

1. Have you ever fainted, passed out, or had an unexplained seizure suddenly and without warning, especially during exercise or in response to sudden loud noises, such as doorbells, alarm clocks, and ringing telephones?
2. Have you ever had exercise-related chest pain or shortness of breath?
3. Has anyone in your immediate family (parents, grandparents, siblings) or other, more distant relatives (aunts, uncles, cousins) died of heart problems or had an unexpected sudden death before age 50? This would include unexpected drownings, unexplained auto crashes in which the relative was driving, or SIDS.
4. Are you related to anyone with HCM or hypertrophic obstructive cardiomyopathy, Marfan syndrome, ACM, LQTS, short QT syndrome, BrS, or CPVT or anyone younger than 50 years with a pacemaker or implantable defibrillator?

What Should Be Done With the Child Who Has a Positive Finding on a Screening Examination or Whose Parents Sought ECG Screening and Were Found to Have an ECG Abnormality?

A positive response from the 4 questions above or an abnormal

TABLE 4 Adapted From the AHA's Recommended 14-point Screen for Cardiovascular Disease

AHA's 14-Point PPE
Personal history
1. Chest pain, discomfort, tightness, or pressure related to exertion
2. Unexplained syncope or near-syncope not felt to be vasovagal or neurocardiogenic in origin
3. Excessive and unexplained dyspnea or fatigue or palpitations associated with exercise
4. Previous recognition of a heart murmur
5. Elevated systemic blood pressure
6. Previous restriction from participation in sports
7. Previous testing for the heart, ordered by a physician
8. Family history of premature death (sudden and unexpected or otherwise) before 50 y of age attributable to heart disease in ≥ 1 relative
9. Disability from heart disease in close relative < 50 y of age
10. Hypertrophic or dilated cardiomyopathy, LQTS, or other ion channelopathies, Marfan syndrome, or clinically significant arrhythmias; specific knowledge of genetic cardiac conditions in family members
Physical Examination
11. Heart murmur, not felt to be innocent
12. Femoral pulses to exclude aortic coarctation
13. Physical stigmata of Marfan syndrome
14. Brachial artery blood pressure (sitting position), preferably taken in both arms

Adapted from Maron BJ, Friedman RA, Kligfield P, et al; American Heart Association Council on Clinical Cardiology; Advocacy Coordinating Committee; Council on Cardiovascular Disease in the Young; Council on Cardiovascular Surgery and Anesthesia; Council on Epidemiology and Prevention; Council on Functional Genomics and Translational Biology; Council on Quality of Care and Outcomes Research, and American College of Cardiology. Assessment of the 12-lead electrocardiogram as a screening test for detection of cardiovascular disease in healthy general populations of young people (12–25 years of age): a scientific statement from the American Heart Association and the American College of Cardiology. *J Am Coll Cardiol.* 2014;64(14):1479–1514.

ECG should prompt further investigation that may include referral to a pediatric cardiologist or pediatric electrophysiologist. A pediatric electrophysiologist will have better insight for suspected channelopathies or arrhythmia issues and will recommend studies as needed. Ideally, the child with a positive response to these questions should be held out of athletic participation until the cardiovascular evaluation is complete.

THE PRIMARY CARE PHYSICIAN'S ROLE IN SECONDARY PREVENTION

What to Do When a Child Has a Cardiac Arrest

When a child has a cardiac arrest, secondary prevention efforts, including resuscitation, are required. Prompt recognition of cardiac arrest with high-quality CPR and early defibrillation are the major determinants of survival, with survival decreasing by 10% with every minute delay in CPR and AED administration.³⁵ The AAP and AHA support efforts to improve survival by early symptom recognition, the

use of 911 or emergency medical services (EMS), effective bystander CPR, and deployment and use of AEDs in the community. Bystander CPR improves the response time to defibrillation, can more than double the survival to hospital discharge, and leads to improved neurologic outcome.³⁶ Unfortunately, only 20% to 30% of people who experience a cardiac arrest outside of the hospital receive bystander CPR or defibrillation, and the CPR performed by bystanders may be suboptimal.³⁷ Annual rates of CPR training in the United States are low, with only 4% of the US population being trained.¹⁷ CPR and AED training is recommended as a high school graduation requirement. To support this effort, there are school-based CPR training programs available from the World Health Organization and the AHA.^{38,39}

Evaluation of the Victim or Survivor of Cardiac Arrest

A comprehensive evaluation of the survivor of cardiac arrest should be undertaken at the direction of a cardiologist with expertise in conditions associated with SCA and SCD.

Important elements in the evaluation of SCA include the following:

1. careful review of the medical history and event, including pre-event symptoms;
2. a multigenerational family history;
3. ECG;
4. exercise stress test (some primary arrhythmias are only seen with or immediately after physical exertion);
5. echocardiography to identify structural defects and abnormal cardiac function; and
6. additional testing as needed, including cardiac MRI, computed tomography, electrophysiology testing, and/or provocative drug testing.

If a clinical phenotype is suspected or proven, targeted genetic testing may be indicated. In the survivor of SCA, genetic testing should be guided by the results of medical evaluation. Results may be used for individual diagnosis, treatment, and screening of at-risk family members for subclinical disease.⁴⁰ Genetic testing results should be interpreted in consultation with a physician specializing in inherited arrhythmia conditions, a

physician specializing in genetics, and/or a genetic counselor.

What to Do When a Child Cannot Be Resuscitated

SCD occurs in approximately 2000 patients younger than 25 years (excluding SIDS deaths) every year in the United States.⁴¹ Autopsy studies of young individuals who have suffered SCD have shown that a structural cardiac cause (HCM, congenital heart anomalies, and myocarditis) is present in the majority of the patients; however, the cause remains unexplained in a significant proportion (6% to 40%).⁴²⁻⁴⁵ Unexplained SCD is often attributed to cardiac arrhythmia caused by cardiac ion channel dysfunction, which is undetectable in a conventional autopsy. Diagnostic yield in families is 4 times higher when there is a survivor of SCA compared with those in whom there was an SCD.⁴⁶ When SCD occurs, assessment of the cardiac anatomy by a skilled medical examiner at the time of autopsy is important. For individuals who do not survive and have no apparent previously identified cause or diagnosis on conventional autopsy, especially if clinical evidence points toward a diagnosis of LQTS or CPVT, a targeted molecular autopsy is recommended⁴⁷ (Table 5). Genetic testing results should be interpreted in consultation with a physician specializing in inherited arrhythmia

conditions, a physician specializing in genetics, and/or a genetic counselor.

The etiologies of SIDS are varied, with the majority of cases attributable to noncardiac causes. Therefore, victims of SIDS do not necessarily require a rigorous cardiac genetic evaluation unless the circumstances at the time of death or family history are suggestive of an arrhythmic death.

COMMUNICATION AND BEREAVEMENT

After a cardiac arrest, communication between the health care team and the family can have a significant effect on the grief response. The initial reaction is frequently shock, followed by other emotional reactions, including anger, guilt, depression, rage, apathy, and loneliness.⁴⁸ Preparing the family for the process that follows death is important (postmortem examination, referral to the medical examiner or coroner, registering the death).⁴⁹ Asking questions and receiving information about the cause of death is important to families.⁴⁸ Blaming oneself for not saving the life of the family member is common, and therefore, reassurance is key in alleviating the guilt.⁴⁸ Survivors of SCA are at risk for posttraumatic stress disorder.⁵⁰ Providing information on bereavement support groups can be

helpful. The HeartRescue Project's Life After SCA initiative provides resources to help survivors and their loved ones return to living happy, healthy, and fulfilled lives.⁵¹ Parent Heart Watch is a national group of parents whose children have experienced SCA and can provide unique support to bereaved families.

EVALUATION OF REMAINING FAMILY MEMBERS

Many of the cardiovascular diseases that put young individuals at risk for sudden death have a familial inheritance pattern. Screening relatives provides the opportunity to identify at-risk individuals and initiate management.⁴⁰ Importantly, in the absence of a diagnosis in the affected individual, cascade screening of first-degree relatives has improved the diagnostic yield of testing. Detailed cardiovascular evaluation of first-degree SCA and SCD relatives has shown that 22% to 30% of these families had evidence of inherited cardiac disease.^{52,53} Steinberg et al reported that 18% of surviving relatives of unexplained SCA and SCD victims reported one or more cardiac symptoms in a first-degree relative of the proband.⁵² Consistent with these findings, it has been recommended that first-

TABLE 5 Recommendations From the National Association of Medical Examiners for Autopsy Evaluation of Young Sudden Death Victims

Recommendations From Medical Examiner
1. At a minimum, samples should be saved from individuals aged ≤ 40 y who die suddenly and unexpectedly and whose deaths remain unexplained at the completion of the autopsy.
2. Circumstances that should be considered suspicious for a possible genetic etiology include, but are not limited to, the following: a. Drowning, particularly in the case of a sober or experienced swimmer; b. Single motor auto crashes when no mitigating factors are present (eg, toxicology results negative, favorable road conditions); c. An unexplained seizure; d. An unexplained death of an individual with a family history of sudden death or inherited heart disease, such as a cardiomyopathy, thoracic aneurysm, or known genetic cardiac diagnosis; e. All deaths that are sudden and unexplained for which cause of death is not clear at autopsy.
2 For the purpose of potential genetic testing and/or DNA banking, an appropriate sample is 5–10 mL of blood collected at autopsy or as part of an external examination that is preserved with K2 EDTA (usually a purple top tube). ⁴⁸

Adapted from Middleton O, Baxter S, Demo E, et al. National Association of Medical Examiners Position Paper: Retaining Postmortem Samples for Genetic Testing. *Acad Forensic Pathol.* 2013;3(2):191–194.

degree family members of patients with SCA and SCD be informed of the potentially increased risk. An assessment should be offered at a center with experience in the diagnosis and management of inherited cardiac diseases.⁴⁰ The initial battery of tests for first-degree relatives usually includes a visit to a pediatric cardiologist or electrophysiologist, an ECG, an exercise stress test (if old enough to exercise), and an echocardiogram. It is reasonable to order molecular genetic testing from the victim after SCA. If a disease-causing variant is identified in the victim, cascade molecular and clinical screening of family members is indicated.⁴⁰ Cascade screening means evaluation beginning with first-degree relatives of the SCA victim. Depending on the results of those screening tests, other family members may need testing as well.

UNDERSTANDING IMPLANTABLE CARDIOVASCULAR DEVICES

Cardiovascular implantable electronic devices can store substantial amounts of diagnostic data related to arrhythmia documentation.⁵⁴

- Pacemakers are electronic devices that stimulate the heart with electrical impulses to maintain or restore a normal rhythm in people with slow heart rhythms. Pacemakers can be programmed to record abnormal rhythms but cannot provide a shock to restore sinus rhythm during an arrhythmia.
- Implantable cardioverter-defibrillators (ICDs) have pacemaker functions but are also capable of providing therapy for tachyarrhythmias, including VT and VF. Therapy can involve either overdrive pacing or a shock that restores sinus rhythm.

Interrogation of an ICD is important after a delivered shock because the arrhythmia that was treated will be recorded along with the therapy and the post-shock rhythm.

- An implantable loop recorder (ILR) is a small device implanted under the skin that can store ECG recordings of the heart rhythm. An ILR can be programmed to record automatically when the patient's heart rate deviates outside the range that is chosen by the physician. An ILR can also be activated by the patient to record during symptoms. ILRs cannot pace or provide therapy.

AED AND CPR

AEDs can accurately detect VF in children of all ages and differentiate shockable from nonshockable rhythms with a high degree of sensitivity and specificity.⁵⁵ For children from birth to 8 years of age, it is reasonable to use an AED pediatric dose-attenuator system and a pediatric pad to reduce delivered energy if one is available; if not, the rescuer should use a standard AED.⁵⁵ Current AHA guidelines do not recommend compression-only CPR for young children.⁵⁶

CARDIAC EMERGENCY RESPONSE PLANS AND THE PCP AS ADVOCATE

On any given day, as many as 20% of the combined US adult and child population can be found in schools. Therefore, school nurses, athletic trainers, and teachers are often required to provide emergency care during the school day and for extracurricular activities, including sports.⁵⁷ A cardiac emergency response plan (CERP) is needed to facilitate an efficient and structured response to SCA. Essential elements of a CERP include the following:

1. establishing an effective communication system;

2. training of anticipated responders in CPR and AED use;
3. access to an AED for early defibrillation;
4. acquisition of necessary emergency equipment;
5. coordination and integration of on-site responder and AED programs with the local EMS system; and
6. practice and review of the response plan.⁵⁸

This plan should target a collapse-to-EMS call time of <1 minute, provision of first aid and CPR when appropriate, and a collapse-to-first shock time of <3 minutes for SCA if an AED is on-site. It is recommended that at least 10% of staff and 50% of physical education staff should have current CPR and AED certification.⁵⁹ At least 2 successful emergency response drills should be conducted every year.⁵⁹

The PCP and pediatric cardiologist can have a major impact in advocating for schools and school districts not only to have a sufficient number of AEDs but also that the staff is continually well trained, the equipment is maintained, and a CERP is in place. Many states have passed legislation requiring CPR or AED training for students to graduate from high school or as part of the health curriculum.⁶⁰ The task force supports efforts through either legislation or local or statewide high school associations to make CPR and/or AED training a requirement for students to graduate from high school.

ROLE OF THE LICENSED ATHLETIC TRAINER

Licensed athletic trainers (LATs) are school-based health care professionals who collaborate with the health care team. The services LATs provide include prevention, emergency care, and therapeutic

intervention.⁶¹ The LAT needs to be able to determine an athlete's readiness to participate and, if necessary, consult with the supervising team physician and/or treating physician. They also play an important role in identifying unsafe facilities or playing environments as well as developing and implementing an emergency action plan in collaboration with supervising team physicians.⁶¹ LATs can be important advocates for CPR and AED use training and for AED placement in public areas, including schools, athletic fields, and arenas.

RETURN TO ACTIVITY AFTER CARDIAC ARREST

The AHA and others have issued recommendations for aerobic and resistance training in children and adolescents.⁶² These recommendations are based on findings that regular physical activity reduces the risk of long-term adverse health outcomes. There is evidence that childhood levels of cardiovascular risk factors predict early subclinical atherosclerosis and cardiac pathology and adult morbidity and mortality.⁶³ Encouraging patients who have suffered cardiac arrest to have a healthy lifestyle including exercise may be beneficial. Exercise restriction needs to be balanced with the potential for lifelong risk of SCA and SCD and the development of other conditions associated with cardiovascular risk. To facilitate a safe return to exercise, these patients may benefit from a medically supervised cardiac rehabilitation program. Any patients, including athletes, who have suffered a cardiac arrest from VT or VF from a cause that cannot be reversed or well-managed with other means (eg, medication) should undergo a thorough evaluation with strong consideration of ICD placement.⁶⁴

There are specific recommendations for those desiring athletic participation with ICDs under advisement by the patient's pediatric electrophysiologist.

Recommendations include returning to low-level dynamic and static activities (eg, golf, bowling, etc) after 3 months of being free of VT or VF requiring device therapy.⁶⁵ Higher-intensity activities can also be considered in discussion with the patient and family in a shared risk arrangement after 3 months without device therapy for VT or VF.⁶⁵ The patient should be counseled on the increased risk of ICD shocks as well as device-related trauma when participating in activities that have a risk of affecting the device.⁶⁵

Patients who have suffered a cardiac arrest from a reversible cause, such as myocarditis or electrolyte abnormality, will often not have an ICD implanted. The most recent recommendations suggest refraining from athletic participation until cleared at a 3-month reevaluation and on the advice of the pediatric cardiologist or electrophysiologist. If the condition causing the cardiac arrest has completely resolved, the athlete may then return to competition.⁶⁵

RECOMMENDATIONS AND PRIMARY TAKEAWAY POINTS FROM THIS POLICY STATEMENT

PCPs, as the preeminent providers of health care to children, should be aware of the features of the clinical history, family history, and physical examination suggestive of a risk for SCA and SCD.

1. All children should be evaluated for conditions predisposing to SCA and SCD in the course of routine health care.¹⁴

2. A thorough and detailed history, family history, and physical examination are necessary to begin assessing SCA and SCD risk.
3. The ECG should be the first test ordered when there is concern for SCA risk. The ECG should be interpreted by a physician trained in recognizing electrical heart disease (ie, a pediatric cardiologist or pediatric electrophysiologist).
 - a. To provide optimal care, ECGs should not be performed in isolation without clinical history; referral to a specialist should be considered.
4. Do not trust the computer interpretation of the ECG.

Recognizing that no single screening strategy will be able to detect all the conditions associated with SCA (primary prevention), it is important to advocate for emergency action plans (secondary prevention) and CPR training in the community. CPR and AEDs are effective for secondary SCA prevention.

Survivors of SCA (and family members of SCA or SCD victims) should have a thorough evaluation to assess the potential of a genetic etiology. Some facilities have specialized centers for SCA. A pediatric SCA center is a children's multispecialty medical facility with expertise in pediatric electrophysiology and inherited channelopathies and cardiomyopathies. This evaluation includes not only molecular genetic testing but also genetic counseling for identifying others who may be at risk.⁶⁶

SUMMARY

The strategy put forth in this policy statement emphasizes the importance of sudden death awareness and prevention that is inclusive of all young people regardless of athletic status. The emphasis shifts from focusing on a single group to expanding the primary and secondary

prevention concepts to a broader group who may achieve similar benefits. There have been many efforts made and published on ways to identify those at risk for SCA and SCD, including clinical (history and physical examination), genetic, and ECG screening. Many SCA and SCD victims cannot be identified before their event, even with testing. Therefore, secondary prevention efforts must not be overlooked by those evaluating large numbers of pediatric patients. Although focusing on prevention efforts in all children may seem to create a burden on PCPs by extending the screening program to more patients, simplification to the aforementioned 4 questions can allow this screening to become incorporated into the routine visit at a minimum of every 3 years. This strategy is intended to increase awareness of SCD prevention in young people and will allow for a healthy lifestyle and reduce the risks of SCA and SCD.

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ABBREVIATIONS

AAP: American Academy of Pediatrics
ACM: arrhythmogenic right ventricular cardiomyopathy
AED: automated external defibrillator
AHA: American Heart Association
BrS: Brugada syndrome
CERP: cardiac emergency response plan
CPR: cardiopulmonary resuscitation
CPVT: catecholaminergic polymorphic ventricular tachycardia
ECG: electrocardiography
EMS: emergency medical services
HCM: hypertrophic cardiomyopathy
ICD: implantable cardioverter-defibrillator
ILR: implantable loop recorder
IVF: idiopathic ventricular fibrillation
LAT: licensed athletic trainer
LQTS: long QT syndrome
PCP: primary care provider
PPE: preparticipation evaluation
QTc: corrected QT interval
SCA: sudden cardiac arrest
SCD: sudden cardiac death
SIDS: sudden infant death syndrome
VF: ventricular fibrillation
VT: ventricular tachycardia

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The guidance in this statement does not indicate an exclusive course of treatment or serve as a standard of medical care. Variations, taking into account individual circumstances, may be appropriate.

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