

## **Sudden Cardiac Arrest in Children and Young Athletes: The Importance of a Detailed Personal and Family History in the Pre-Participation Evaluation**

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## Pre-participation Evaluation Using Patient and Family History

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### **ABSTRACT**

Healthcare providers have become more aware of and concerned about pediatric sudden cardiac arrest. The diseases predisposing a patient to sudden cardiac arrest are all infrequently encountered. However, a detailed and comprehensive patient and family history may reveal warning signs and symptoms that identify a patient at higher risk for sudden cardiac arrest. Since many of these diseases are genetic, extensive family evaluation may uncover a previously undetected cardiac disease process and as well direct the development of a complete family evaluation and treatment plan. Published data document that in many cases preceding warning symptoms and signs are present, but may be misinterpreted or disregarded by medical staff. Attention to the details of patient history, family history, and physical exam is critical to the success of any detection strategy, which can and should be widely applied.

## **INTRODUCTION**

That a child, young adult, or well-trained athlete could suffer sudden cardiac arrest (SCA) seems almost incomprehensible. The numbing pall cast over a community – parents, family, teachers, teammates, and neighbors – is profound. Preventing the tragedy of young-age SCA remains a concern to every healthcare provider evaluating children and athletes and committed to preventing such a tragedy.

This manuscript is intended to review the incidence of SCA in children and young athletes; discuss the different causes of SCA, genetic inheritance, and disease-specific clinical presentations; and emphasize the role for comprehensive personal and family history during the pre-participation evaluation to prevent SCA.

## **INCIDENCE**

The frequency of **SCA** in children and adolescents has been variably reported between 0.8-6.2 per 100,000 per year<sup>1-5</sup>. This is in contrast to the much higher incidence of **SCA** in adults, reported to be 1 per 1,000 adults per year. Recent studies suggest that **SCA** in children and adolescents may be actually increasing<sup>6-7</sup>; the reasons for this increase in frequency are not entirely clear. In the United States, there is no centralized and/or mandatory registry for pediatric and young adult SCA. Therefore, data are compiled usually through media reports, from lay advocacy groups, and/or peer reviewed publications from large referral medical

centers. **It is currently estimated by the Centers for Disease Control that nearly 2,000 patients < 25 years of age will die annually from cardiovascular causes in the United States; this number may surpass 5,000 per year if the age is extended to young adults < 35 years of age<sup>8</sup>.**

Data regarding SCA incidence in a general pediatric population are limited. In 1985 Driscoll<sup>2</sup> reported a review of 33 years of death certificates for all residents of Olmstead County, MN for patients between 1-22 years of age. Sudden and unexpected death occurred in 12/515 certificates reviewed (2.3%), for an overall incidence of 1.3 per 100,000 patient years. In 7/12 cases, deaths were felt to be cardiac-related or probably cardiac related; 3/12 patients had a prior history of syncope, 2 of whom had syncope with exercise and both of whom died during exercise.

A 1996 review by Steinberger<sup>9</sup> reported findings from autopsy specimens from 70 patients < 21 years of age who suffered **SCA**. Patient age was < 1 year in 20/70 patients. Significant cardiac abnormalities were detected in 13 of the 20 infants (65%). Death in half of these patients was felt to be due to coronary artery anomalies. Cardiac abnormalities were detected in 40/50 older patients (80%) and coronary artery anomalies were present in 12 patients (24%).

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Finally, Wisten<sup>10</sup> reviewed the frequency of **SCA** in 15-35 year old Swedish patients between 1992-1999. A national database detected 181 cases of **SCA**, computing to an incidence of 0.093 per 100,000 per year. Preceding symptoms, including chest pain, dizziness, syncope, palpitations, and dyspnea, were present in half the cases. The most common diagnoses at autopsy were: 1) no structural abnormality (21%), possibly representing patients with cardiac electrical abnormalities; 2) coronary atherosclerosis (18%); 3) dilated cardiomyopathy (DCM) (12%); 4) hypertrophic cardiomyopathy (HCM) (11%); and 5) myocarditis (11%).

The likelihood of **SCA** in children and young adults is known to be enhanced by athletic participation. In 1986 Maron<sup>11</sup> reported that HCM was the most common cause of **SCA** in young competitive athletes < 35 years of age. Less common cardiac causes of **SCA** in this patient group included congenital coronary anomalies, ruptured aorta (due to medial necrosis), non-specific left ventricular hypertrophy, and coronary atherosclerosis. Twenty-five percent of the competitive athletes who died suddenly had an underlying cardiovascular abnormality suspected before participation.

In 1998<sup>12</sup>, data from a mandatory insurance program for catastrophic injury or death in Minnesota high school athletes, grades 10-12, revealed 3 deaths due to

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cardiac disease. Based upon the number of sports participants the calculated risk for **SCA** was 1 per 500,000 participants and 1 per 217,400 per academic year.

More recently, intensive search of public media reports and other electronic databases have identified a larger number of cases of **SCA** in U.S. athletes than previously established. The U.S. Sudden Death in Young Athletes Registry has identified approximately 115 cases of **SCA** per year in young competitive athletes<sup>13</sup>. With approximately 5 million competitive high school athletes and 500,000 competitive collegiate athletes in the U.S., a more accurate estimate of the annual incidence of **SCA** in young athletes is about 1:50,000 athletes.

Thus, the incidence of **SCA** in young competitive athletes in the U.S. may be 2-4 times greater than initial estimates and is supported by studies having a mandatory reporting system for **SCA**. Eckart and colleagues reported on 126 non-traumatic sudden deaths in U.S. military recruits (median age 19, range 18-35 years) and found the incidence of exercise-related **SCA** to be 1/9,000<sup>14,15</sup>. In Italy, with a mandatory reporting system for juvenile sudden death, the baseline incidence of **SCA** in young competitive athletes was 1:25,000 prior to the implementation of a national screening program.<sup>16</sup>

Reporting and referral bias may impact the reported incidence as well as causes of **SCA**. The difficulty determining the exact cause of death in patients who suffer primary cardiac electrical disorders (“autopsy negative”) must also be recognized. Many reports actually predate much of what is known now about some of the rare cardiac electric abnormalities including catecholaminergic polymorphic ventricular tachycardia (CPVT), Short QT syndrome (SQTS), and Brugada syndrome, for example.

### **DIFFERENTIAL DIAGNOSIS**

The differential diagnosis for causes of pediatric and young adult **SCA** is detailed in Table 1. In general, these causes of pediatric and young adult **SCA** can be **separated** into structural or functional abnormalities, primary cardiac electrical disorders (usually echocardiographic and autopsy negative), and others. The “other” category includes illicit drugs and stimulants including ephedra, steroids, and cocaine, among others.

## **GENETICS**

Table 1 also designates those disorders which are known to be genetic, and therefore familial. The identification of even a first presymptomatic proband is critical. Several studies have now documented the results of cardiac evaluation of immediate first-degree relatives using more recent genotype techniques. Authors of a 2003 publication<sup>17</sup> reported their evaluation for inherited cardiac disease in cases of sudden arrhythmic death syndrome. Their study revealed 27% of surviving relatives had one or more cardiac symptoms (palpitations 12%, presyncope 6%, syncope 8%, chest discomfort 8%, and dyspnea 7%). There was a 22% incidence of unexpected premature sudden death in addition to the proband in any relative, with a 6% incidence of sudden death within a first-degree relative.

Tester and Ackerman<sup>18</sup> described 49 cases (average age of death  $14.2 \pm 10.9$  years) of autopsy-negative sudden unexplained death. LQTS-associated mutations were discovered in 10 cases; 7 patients had mutations in CPVT genes. Family history of **SCA** or syncope was documented by the medical examiner in 26 (53%) cases but was not specified in 23 cases. There was a personal history of syncope, seizure-like activity, and/or cardiac arrest in 7 cases.

Finally, an investigation in 43 families with one unexplained **SCA** victim < 40 years of age was reported in 2005<sup>19</sup>. A likely cause of death due to an inherited

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disease was found in 17/43 families (40%). Family evaluations discovered 151 presymptomatic disease carriers (average 8.9 per family).

The data presented support the use of a detailed extended family history to identify athletes with familial cardiac disorders that place them at risk for SCA. A 2008 publication discusses the role of family history for evaluating cardiovascular genetic disorders, with a comprehensive table outlining the genes associated with cardiomyopathies and channelopathies<sup>20</sup>. The pedigree as a family history tool is one of the most effective mechanisms for practical application of genetics knowledge into clinical care. A family history template suggested by the U.S. Surgeon General's Family History Initiative is available free at [www.hhs.gov/familyhistory](http://www.hhs.gov/familyhistory).

**Does cardiovascular risk screening work?**

Why should we expect that careful attention to family history, patient history, and physical exam would identify any patients and families at risk for SCA? Despite the widespread use of the sports pre-participation evaluation (PPE), some studies have disparaged the effectiveness of such a PPE process. A 1996 study reviewed the 1985-1995 sudden deaths in 158 United States trained athletes<sup>21</sup>, 134 of whom were felt to have suffered SCA due to a cardiovascular cause. Median age was 17 years (range 12-40 years), with a 90% male incidence. Participation in basketball

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and football accounted for 68% of the sudden deaths. Collapse during or after a training session (78 cases) or during a formal athletic contest (43 cases) was noted. HCM was present in 48 athletes (36%) and was more common in African-American versus white athletes (48% versus 26% of deaths). Anomalous origin of the coronary arteries was detected in 17 athletes (13%). Only 3% of 158 athletes who suffered SCA were suspected of suffering from cardiovascular disease and only one athlete with a specific cardiac anomaly causing SCA was correctly identified by a pre-participation screen. Although the details and adequacy of the PPEs performed were not reported, these authors summarized that “pre-participation screening appeared to be of limited value for identification of underlying cardiovascular abnormalities”. **More recently, investigators in the United Kingdom summarized that family history and personal symptom questionnaire alone were inadequate for the identification of patients and families at risk for SCA<sup>22</sup>.**

However, symptoms in patients with primary structural/functional or electrical cardiac abnormalities may in fact be relatively common, but misinterpreted or disregarded by medical staff. A 1996 report<sup>23</sup> summarized 9 publications detailing the characteristics of 469 sudden deaths from cardiac causes in young people. These studies collectively reported preceding symptoms of dizziness, chest pain, syncope, palpitations, dyspnea, and/or family history of sudden death from a

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cardiac cause in 25-61% in their study population. Deaths were exertion-related in 8-33% of the cases. A report from the Italian Sports Screening program described findings from 55 screened athletes who subsequently suffered **SCA**<sup>16</sup>. PPE identified 24 (44%) with one or more positive findings such as family history for cardiomyopathy, sudden death, or both (n=6), palpitations on exertion (n=10), syncope (n=7), chest pain (n=2), and/or cardiac murmur (n=4). All findings were “considered to be of little or no clinical relevance.” Another retrospective study of 162 individuals (age 15-34) who underwent autopsy evaluation after **SCA** found 92 cases had a preceding history of syncope/presyncope, chest pain, palpitations, and/or dyspnea<sup>24</sup>. In 26 of these subjects there was a family history of **SCA**. A 2005 Australian<sup>25</sup> study about sudden, natural death in persons 5-35 years of age found the most common cardiac cause of sudden death was presumed arrhythmia in those with no or minimal structural heart disease (29%). Sudden death associated with exercise represented 11% of the cases. A history of **SCA** was reported in first-degree relatives in 4.5% of the decedents. Interestingly, this study also reported a 24% incidence of sudden death due to “epilepsy”, an occasional misdiagnosis in patients with LQTS or CPVT. Importantly, symptoms may also be confusing and non-specific, especially in athletes who may overexert until physical exhaustion.

Therefore, careful attention to the details of patient and family history during a pre-participation evaluation may identify a subset of individuals at risk for **SCA** from underlying cardiovascular disease.

### **DISEASE-SPECIFIC CLINICAL PRESENTATION**

**Hypertrophic Cardiomyopathy (HCM).** The clinical presentation of 711 adult patients with HCM is enlightening even during a discussion of pediatric and young adult patients<sup>26</sup>. 305 patients (43%) presented with exertional chest pain or dyspnea, palpitations, fatigue, and/or dizziness/near syncope; 225 were identified during routine medical examination (163/225 as a result of a heart murmur).

Family history concerns prompted evaluation subsequently identifying 91 patients (13%). Eighty Australian HCM children were described in a 2005 publication<sup>27</sup>. Presenting symptoms were congestive heart failure (8%), arrhythmia symptoms (3%), murmurs (53%), and family history of HCM (15%). No patient presented with SCA as a first manifestation of the underlying HCM.

HCM patients may present with respiratory complaints due to decreased left ventricular compliance and/or pulmonary venous hypertension; symptoms may mimic exercise-induced bronchospasm. Failure of empiric drug treatment and/or negative formal pulmonary function studies should prompt extensive cardiac evaluation for HCM.

### **Arrhythmogenic Right Ventricular Cardiomyopathy (ARVC).**

A retrospective report of pre-participation screening in 11 athletes who subsequently died of ARVC revealed that 18% (2/11 patients) had a history of sudden death from heart disease, 54% of patients suffered palpitations on exercise and 45% of patients had a personal history of syncope<sup>1</sup>. A United States experience of ARVC was reported in 2005<sup>28</sup>. This 100 patient study population ranged in age 2-70 years. Familial pattern of ARVC was observed in 32 patients. Palpitations, syncope, and sudden cardiac arrest were the presenting features in 27%, 26%, and 23% of patients, respectively. It is known that palpitations and/or syncope may herald **SCA**, which often occurs during or shortly after activity.

### **Congenital Coronary Artery Abnormalities**

A single right coronary artery, giving rise to an anomalous left coronary artery running an interarterial course, is the most common congenital coronary artery abnormality predisposing to **SCA**. Patients may present with syncope, chest pain, and/or dyspnea. Basso et. al. reported 27 sudden deaths in young athletes identified at autopsy as suffering from congenital coronary artery anomalies<sup>29</sup>. Syncope (most commonly exertional), chest pain, and/or palpitations were present in 10/12 patients for whom clinical information was available. These congenital

coronary artery abnormalities are not genetic and none of the athletes described in this publication had a family history of **SCA** or premature cardiovascular disease.

**Dilated Cardiomyopathy (DCM).** The degree of symptomatology often correlates with the degree of myocardial dysfunction. However, despite echocardiographic findings of severe ventricular dilatation and dysfunction, symptoms may be quite subtle or even apparently absent. Shortness of breath and exercise intolerance are the most common symptoms in older children and generally relate to decreased cardiac output and/or pulmonary venous hypertension with pulmonary edema. Patients may present with a murmur of mitral insufficiency and/or gallop rhythm or subtle findings of congestive heart failure. Palpitations and syncope may herald the presence of complex supraventricular or ventricular arrhythmias which have been regarded as predictors for **SCA** and therefore consideration for early heart transplantation.

Dilated cardiomyopathy may result from previous myocarditis, neuromuscular disorders, or even inborn errors of metabolism. Familial dilated cardiomyopathy is now recognized, and family members should be thoroughly investigated for non-specific history of heart disease or cardiac symptoms including **SCA**<sup>30</sup>.

**Marfan Syndrome.** Marfan syndrome patients may present with chest pain due to aortic dissection and/or rupture or even pneumothorax. However, patients most commonly present with the phenotypic Marfanoid stigmata<sup>31</sup>. This syndrome probably represents, along with HCM, the most important reason for careful cardiovascular physical examination as part of the PPE screening process.

Although this is a genetic disorder of the fibrillin gene, many cases appear to be sporadic with an otherwise negative family history. **SCA** is most commonly due to aortic dissection with rupture, but death due to ventricular arrhythmia has been described<sup>32</sup>. The presence of palpitations should raise the possibility of clinically significant arrhythmias.

**Wolff-Parkinson-White Syndrome (WPW).** WPW syndrome tends to be sporadic and commonly pediatric WPW patients have normal cardiac structure and function. Presentation with palpitations or sustained tachyarrhythmia (usually due to reentrant orthodromic SVT) is typical. However, a small subset of WPW patients may be at risk for cardiovascular collapse, cerebrovascular accident, or **SCA** due to ventricular fibrillation<sup>33,34</sup>. These WPW patients may experience atrial fibrillation, which induces ventricular fibrillation as a result of rapid anterograde conduction to the ventricles over the accessory pathway. The presence of WPW syndrome on ECG should prompt careful electrophysiologic pre-athletic clearance.

**Long QT Syndrome (LQTS).** The clinical findings for 3,343 individuals from 328 families in which one or more family members were identified as affected with LQTS (defined by  $QT_c \geq 440\text{ms}$ ) was reported in 1991<sup>35</sup>. Affected individuals usually presented with syncope during childhood or teenage years, oftentimes misdiagnosed as having benign syncope or seizure disorder. A 2007 report found a family history of **SCA** or syncope documented by a medical examiner in 26/49 cases of sudden unexplained death at an average age of  $14.2 \pm 10.9$  years; LQTS-associated genetic mutations were noted in 10/49 patients<sup>18</sup>. A personal history of syncope, seizure-like activity, and/or cardiac arrest before these sudden unexplained deaths was reported in 7/49 cases. The association between drowning/near drowning<sup>36</sup> and separately with SIDS<sup>37</sup> with LQTS has been documented.

**Catecholaminergic Polymorphic Ventricular Tachycardia (CPVT).** CPVT, like LQTS, is associated with sudden cardiac arrest at a relatively young age. A 1995 study, reporting the results of 21 CPVT patients, found that syncope was a presenting symptom for 20/21 patients with the first syncopal event occurring at  $7.8 \pm 4$  years of age (range  $\overline{3-16}$  years)<sup>38</sup>. Physical effort or emotion typically triggered symptoms and the diagnosis of CPVT was often delayed due to the misdiagnosis of epilepsy, vasovagal events, or neurologic causes. Otherwise, probands are often referred due to life-threatening arrhythmias during physical or

emotional stress. Similar to LQTS, CPVT mutations have been discovered in drowning/near drowning victims.

**Short QT Syndrome (SQTS).** Only recently has the association between SQTS on ECG and familial sudden death been reported. A recent report describes 29 patients with SQTS and personal and/or family history of cardiac arrest<sup>39</sup>. Median age at diagnosis was 30 years (range 4-80 years). 18 patients (62%) were symptomatic; 10 had cardiac arrest (34%), and in 8 patients this was the first clinical presentation (in the first months of life in two patients). Seven patients (24%) experienced syncope. Atrial fibrillation was documented in 9 patients (31%), even in young subjects. As with LQTS and CPVT, SQTS may be associated with **SCA** at very young age.

**Brugada Syndrome.** Although Brugada syndrome generally presents symptomatically at an older age, during the 3<sup>rd</sup> or 4<sup>th</sup> decades of life, families with a more malignant Brugada syndrome phenotype have been described. A 2007 publication described 113 children < 16 years of age (mean  $8 \pm 4$  years) who were affected by Brugada syndrome<sup>40</sup>. Diagnosis of Brugada syndrome was made due to aborted sudden death (1), syncope of unexplained origin (10), symptomatic SVT (1), suspicious **ECG** (1), and family screening for Brugada syndrome (17). Syncope was precipitated by fever in 5 cases. During follow-up one child

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experienced **SCA** but two received lifesaving **implanted cardioverter-defibrillator** shocks for ventricular fibrillation.

**Congenital Complete Heart Block (CHB).** Congenital CHB results most commonly from an autoimmune process resulting in destruction of AV nodal conduction as well as myocardial inflammation<sup>41,42</sup>. In the presence of an adequate junctional or ventricular escape rate, patients may remain asymptomatic though bradycardiac at rest and exhibiting a blunted maximum exercise heart rate. Symptoms of bradycardia may include easy fatigue, exercise intolerance, syncope/near syncope, or seizures. A first Stokes-Adams attack, due to severe bradycardia or asystole, or conversely rather ventricular fibrillation, may prove fatal<sup>43</sup>. The clinical finding of bradycardia may represent sinus node dysfunction or CHB and **ECG** will be required to differentiate these rhythms from physiologic sinus bradycardia in a well-trained endurance athlete.

**Pre-participation Evaluation Form and Cardiovascular Risk Assessment.** The above data support the concept that warning signs and symptoms may be present in patients and/or families affected by cardiac diseases and predisposed to **SCA**. For screening purposes, an American Heart Association document outlines the basic 12-element recommendation for pre-participation cardiovascular screening of competitive athletes<sup>44</sup>. These elements include a) personal history for exertional

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chest pain/discomfort; unexplained syncope/near syncope; excessive exertional and unexplained dyspnea/fatigue, associated with exercise; prior recognition of a heart murmur; elevated systemic blood pressure; b) family history of premature death (sudden and unexpected, or otherwise) before age 50 years due to heart disease, in  $\geq 1$  relative; disability from heart disease in close relatives  $< 50$  years of age; and specific knowledge of certain conditions in family members: hypertrophic or dilated cardiomyopathy, LQTS or other ion channelopathies, Marfan syndrome or clinically important arrhythmias. In addition to the personal and; c) family history elements, physical examination should evaluate carefully for a heart murmur, femoral pulses to exclude aortic coarctation, physical stigmata of Marfan syndrome, and brachial artery blood pressure (sitting position).

Alternatively, the American Academy of Pediatrics, along with five other sponsoring agencies, has endorsed the third edition of the PPE Monograph now used in many settings throughout the United States (<http://amssm.org/ppehistory.pdf>)<sup>45</sup>. This document acknowledges the wide variation in how physicians approach, conduct, and document these PPE examinations. Questions 1-3 deal with denial or restriction of participation in sports for any reason and current on-going medical conditions or medications. Questions 5-14 specifically address signs and symptoms (personal or family history) which may suggest cardiovascular risk. Many states have now endorsed the use of this standardized PPE format, in order

to eliminate unnecessary variability and to more effectively screen for cardiovascular (and orthopedic) risk.

Athletic participation increases the likelihood of sudden death for many of these disorders predisposing to **SCA**. **Corrado et. al., identified a 2.5 times relative risk factor for SCA due to sports activity in athletes versus an age-matched non-athletic population**<sup>46</sup>. The intent of most pre-participation evaluation formats is to evaluate children and young adults before undergoing specific athletic participation. However, as noted specifically for LQTS, SQTS, and CPVT, first symptoms and/or sudden cardiac arrest may occur at young age. Not all children are athletes but all are active and “athletic”. Postponing a detailed pre-participation evaluation cardiovascular risk assessment until high school or college may therefore underdetect patients and families at risk. Therefore, an alternative cardiovascular risk assessment form has been proposed (Figure 1). This form can be initiated for any patient of any age by any care provider at any time. The form incorporates questions from the AAP PPE, as well as questioning of parents and family members about specific genetic disorders known to predispose to **SCA**, accidental deaths, near drowning, congenital deafness, and unexplained seizure disorders.

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The cardiovascular risk assessment process could be used in a continuum of well-child care, including first postnatal visit, pre-kindergarten, pre-middle school, and before high school entry for non-athletes who will not be exposed to a formal pre-participation athletic evaluation.

For any pre-participation evaluation or cardiovascular risk assessment to be successful, examiners must be aware signs and symptoms that may identify patients or families at risk. The use of competent and qualified examiners is still a concern with recent data finding that 35% of states allow non-physician examiners with little cardiovascular training to perform the evaluation<sup>47</sup>.

**SUMMARY**

**SCA** in young athletes and children is caused by a diverse etiology of structural/functional and electrical diseases of the heart. With the use of better and/or mandatory reporting systems, the incidence of **SCA** appears to be considerably higher than initial estimates. Published data verify that many of the cardiac disorders known to cause **SCA** may present with symptoms of syncope, presyncope, chest pain, dyspnea, palpitations, or seizures. A family history of sudden unexpected death, unexplained drowning, near drowning, car accident, or seizures may also be present. Realizing that many of the diseases that cause pediatric **SCA** are genetic, identification of even a first asymptomatic family

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member may reveal a more extensive familial involvement. Attention to the details of patient history, family history, and physical exam (specifically for detection of HCM-associated left ventricular outflow tract obstruction murmur and the physical findings suggestive of Marfan syndrome) is critical to the success of any detection strategy. This process can and should be widely applied. Patients or families suspected at risk by a primary provider **should be referred** to a cardiologist for further evaluation.

Table 1 SCA Differential Diagnosis

Figure 1 Pediatric Sudden Cardiac Arrest Risk Assessment Form

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Table 1: SCA Differential Diagnosis

**Structural/Functional**

- 1) **Hypertrophic Cardiomyopathy (HCM)\***
- 2) Coronary Artery Anomalies
- 3) **Aortic Rupture/Marfan\***
- 4) **Dilated Cardiomyopathy (DCM)\***
- 5) Myocarditis
- 6) Left Ventricular Outflow Tract Obstruction
- 7) Mitral Valve Prolapse (MVP)
- 8) **Coronary Artery Atherosclerotic Disease\***
- 9) **Arrhythmogenic Right Ventricular Cardiomyopathy (ARVC)\***
- 10) Post-operative Congenital Heart Disease

**Electrical**

- 11) **Long QT Syndrome (LQTS)\***
- 12) Wolff-Parkinson-White Syndrome (WPW)
- 13) **Brugada Syndrome\***
- 14) **Catecholaminergic Polymorphic Ventricular Tachycardia (CPVT)\***
- 15) **Short QT Syndrome\***
- 16) Complete Heart Block (CHB)

**Other**

- 17) Drugs and Stimulants
- 18) **Primary Pulmonary Hypertension (PPH)\***
- 19) Commotio Cordis

\* **Familial / Genetic**

Figure 1

### Pediatric Sudden Cardiac Arrest Risk Assessment Form

**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)**

Patient History Questions: Tell me about any of these in your child...	Yes	No
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?		
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 accident, 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: Hypertrophic cardiomyopathy (HCM)		
Dilated cardiomyopathy (DCM)		
Heart Rhythm problems: Long QT syndrome (LQTS)		
Short QT syndrome		
Brugada syndrome		
Catecholaminergic ventricular tachycardia		
Arrhythmogenic right ventricular cardiomyopathy (ARVC)		
Marfan syndrome (aortic rupture)		
Heart attack, age 50 or younger		
Pacemaker or implanted defibrillator		
Deaf at birth (congenital deafness)		
<b>Please explain more about any "yes" answers here:</b>		
<b>Parent Signature:</b> _____		
<b>Physician Signature:</b> _____		
<b>Date:</b> _____		

