Sudden unexpected cardiac death (SCD) in a child or adolescent is a devastating event with serious impact on the family, the school, and the community. This article reviews the epidemiology of SCD in children and adolescents and includes a discussion of its incidence and etiologies. It also discusses strategies for prevention.

Incidence

Various retrospective reviews were undertaken to determine the incidence of SCD in children and adolescents. The reported incidence has varied from 0.8 to 6.2 cases/100,000 population per year [1–5]. This is in contrast to the much higher incidence of SCD in adults, which is reported to be 1 case/1000 population per year. Recent studies suggest that the incidence of SCD in children and adolescents is increasing [6,7]. The reasons for this increase in frequency are not clear.

In 1985, Driscoll and Edwards [2] reviewed the death certificates of all residents of Olmstead County, Minnesota who were between 1 and 22 years of age from January 1950 to October 1982. Of the 515 certificates that were reviewed, deaths in 12 cases (2.3%) were sudden and unexpected; this resulted in an overall incidence of 1.3 cases per 100,000 patient years. Seven of the 12 deaths were cardiac-related or probably cardiac-related. Three of the 12 patients had a
history of syncope, 2 of the 3 patients had syncope with exercise, and both died while exercising.

Maron et al [8] reported in 1986 that hypertrophic cardiomyopathy (HCM) was the most common cause of SCD in young competitive athletes who were younger than 35 years of age; it accounted for about half of the SCDs [8]. They also noted that other cardiovascular abnormalities, which are less frequent, but important, causes of SCD in young athletes, included congenital coronary abnormalities, ruptured aorta (due to medial necrosis), left ventricular hypertrophy, and coronary atherosclerosis. In that review, only 25% of the competitive athletes who died suddenly had an underlying cardiovascular abnormality that was suspected before participation.

In 1992, Waller et al [9] reviewed the incidence of SCD between 1985 and 1990 in Marion County, Indiana during athletic events. Overall, 18 athletic deaths resulted in a frequency of 0.04%. Of the 18 sudden athletic deaths, 88% were of cardiac origin. Waller et al concluded that retrospective screening would have detected most of the cardiac conditions that were responsible for death.

In 1996, Steinberger et al [10] reviewed the autopsy specimens of 70 patients who were younger than 21 years of age and died suddenly. Twenty patients were less than 1 year of age and 50 patients were between 1 and 20 years of age. Significant cardiac abnormalities were present in 13 (65%) of the 20 infants; 10 (50%) had coronary artery anomalies due to abnormal aortic origin. Among the 50 older patients, cardiac abnormalities were found in 40 (80%) and coronary artery anomalies existed in 12 (24%). In both groups, anomalies of the aortic origin of the left main coronary artery were more frequent than right coronary artery anomalies.

Maron et al [11] systematically reviewed the clinical information and circumstances that were associated with sudden death in 158 trained athletes throughout the United States from 1985 through 1995. This analysis included interviews with family members, witnesses, and coaches, as well as postmortem anatomic, microscopic, and toxicologic data. Of the 158 sudden deaths, 24 (15%) were due to noncardiovascular causes. Among the 134 athletes who died of cardiovascular causes, the median age was 17 years (range 12–40), 120 (90%) were male, 70 (52%) were white, and 59 (44%) were black. Deaths that occurred during basketball (47 cases) and football (45 cases) accounted for 69% of the cases. One hundred and twenty-one athletes (90%) collapsed during or after a training session (78 cases) or during a formal athletic contest (43 cases). The most common structural defects noted were HCM (48 athletes, 36%), which was more common in black versus white athletes (48% vs. 26% of deaths). Anomalous origin of the coronary artery was present in 17 athletes (13%). Of the 115 athletes who had a preparticipation medical evaluation, only 4 (3%) were suspected of having cardiovascular disease; the specific cardiovascular abnormality that was responsible for SCD was identified correctly in only 1 athlete.

In another review by Maron et al [12] in 1998, an attempt was made to determine the incidence of SCD in young athletes. Based upon a mandatory insurance program for catastrophic injury or death for Minnesota high-school
athletes, they reviewed the data over a 12-year period from 1985 to 1997 for competitive high school athletes in grades 10 through 12. Three deaths were noted; one from anomalous left coronary artery from the right sinus of Valsalva, one from congenital aortic stenosis with a bicuspid aortic valve, and one from myocarditis. Based upon the number of sports participants, the calculated risk for SCD was 1 in 500,000 participants and 1 in 217,400 participants per academic year.

The National Center for Catastrophic Sport Injury Research (NCCSIR), an organization that tracks voluntary reports of serious injury and death during competition in male and female high school and college athletes, also is a source of data. In 2000, NCCSIR reported 15 deaths in high school athletes nationwide [13]. Two of the episodes were a result of commotio cordis, 12 were due to other nontraumatic causes of SCD, and one was secondary to a brain injury. These data translate to a total annual incidence of 1 case/350,000 high school athletes per year. In addition, over a 10-year period, from July 1983 to June 1993, NCCSIR reported 126 nontraumatic sports deaths among the 5 million high school athletes and 34 deaths in college athletes. This translates to 16 deaths per year in the United States in high school and college athletes. These data are accumulated by voluntary reporting and in addition may not include information that is related to death during noncompetitive athletic endeavors. Therefore, these data may underestimate the true incidence of SCD in children and adolescents.

In 2002, Zheng, et al [14] reported the incidence of SCD by age group. One thousand, one hundred and sixty-six out-of-hospital deaths were reported in the 5- to 24-year-old group. These data were accumulated by reviewing international classification of diseases (ICD) codes that included deaths that were related to congenital heart disease, pulmonary hypertension, and other vascular abnormalities. Therefore, this number is likely to be an overestimation of the true incidence of SCD in children and adolescents each year.

In summary, the true number of SCDs in children and adolescents in the United States each year is not known definitively. Many of the studies that were cited are not comprehensive, do not have the ability to capture all of the data, or include causes of death in addition to what we might define as “unexpected sudden cardiac death.” The true incidence, therefore, is likely to be greater than 15 per year (NCCSIR) but less than the 1166 per year as reported by Zheng et al [14]; a reasonable estimate is in the range of 500 episodes per year of unexplained SCD in children and adolescents in the United States. There would be tremendous benefit for the establishment of a registry, with mandatory reporting, to establish the true incidence of this problem. Such an entity also would enable additional data to be collected and would allow for additional research.

The above studies have allowed us to make other generalizations and conclusions. These studies demonstrate the variability of the presence of antecedent symptoms. As a result, the usefulness of preparticipation screening questionnaires has been challenged, although some investigators have suggested that as many as 50% of patients who had SCD may have had antecedent symptoms (see later discussion and elsewhere in this issue). More than 80% of young patients who
had unexplained sudden death have an underlying cardiac cause that is discovered at autopsy. In the absence of any symptoms, the challenge is to diagnose the underlying condition before death in a cost-effective and accurate manner. Whether this is possible remains in question. Finally, the above studies allow us to conclude that the higher-risk group appears to be boys in their midteens who are participating in competitive athletics. Basketball and football are the highest risk sports. These studies also identify a subset of subjects who are at risk for SCD and in whom further evaluation should be undertaken. These include patients who have exercise-associated syncope and a family history of unexplained sudden death or HCM. Further discussion of screening, including electrocardiographic and echocardiographic screening, is discussed elsewhere in this issue.

**Causes**

Box 1 lists the causes of SCD in children and adolescents. We have classified the lesions into subdivisions that include: structural or functional abnormalities, primary electrical abnormalities, acquired conditions, and postoperative congenital heart disease. The relative frequency of each of these entities is variable.

Many of the causes of SCD are familial and have recognized patterns of Mendelian inheritance (see Box 1). The importance of this concept is obvious. A detailed family history to uncover the presence of a familial cause of SCD or unexplained death at a young age should lead to further investigation of the index patient.

It was suggested that geographic differences in the relative frequencies of the causes of SCD may exist. Basso et al [15] investigated the causes of SCD in young individuals in the Veneto region of Italy from 1978 to 1993. Two hundred cases of SCD in young people (less than 35 years of age) were noted; 163 (81.5%) of the cases were of cardiovascular origin. Twenty-three percent of the deaths were secondary to obstructive coronary atherosclerosis, 12.5% were due to arrhythmogenic right ventricular dysplasia (ARVD), 10% were due to mitral valve prolapse, 10% were due to conduction system abnormalities, 8.5% involved congenital coronary abnormalities, 7.5% had myocarditis, 5.5% had HCM, 5.5% had aortic rupture, 5% had dilated cardiomyopathy, 3.5% had acquired coronary disease, 3% had postoperative congenital heart disease, and 2% had aortic stenosis.

A subsequent study by Thiene et al [16] in 1999 reviewed 49 athletic deaths in the Veneto region of Italy from 1979 to 1996. The incidence of SCD was calculated at 0.75 cases/100,000 population per year in nonathletes and 1.6 cases/100,000 population per year in athletes. Among the athletes, ARVD was the leading cause of death followed by congenital coronary abnormalities. HCM is an uncommon cause of SCD in Italy because affected individuals often are identified by routine preparticipation screening which includes electrocardiography. The reasons for the ostensibly higher incidence of ARVD in Italy remains speculative.
Box 1. Etiologies of sudden unexplained cardiac death in children and adolescents

**Structural or functional abnormalities**

- Hypertrophic cardiomyopathy*
- Arrhythmogenic right ventricular dysplasia*
- Coronary artery abnormalities
  - Left coronary artery arising from the right sinus of Valsalva
  - Right coronary artery arising from the left sinus of Valsalva
  - Single coronary artery ostia
  - Hypoplastic coronary artery syndrome
  - Williams syndrome with coronary ostial stenosis
  - Kawasaki syndrome
- Primary pulmonary hypertension*
- Myocarditis/dilated cardiomyopathy*
- Restrictive cardiomyopathy
- Marfan’s syndrome with aortic dissection*
- Aortic valve stenosis

**Primary electrical abnormalities**

- Long QT syndromes
  - Romano Ward*
  - Jervell Lange Nielsen*
  - Acquired
- Brugada syndrome*
- Wolf-Parkinson-White syndrome
- Primary or idiopathic ventricular tachycardia/fibrillation
- Catecholamine-exercise ventricular tachycardia
- Heart block
  - Congenital
  - Acquired

**Acquired**

- Commotio cordis
- Drug abuse
  - Cocaine, stimulants, inhalants, gasoline, glue, typewriter correction fluid, nitrites (amyl, butyl), emetine
- Secondary pulmonary artery hypertension (Eisenmenbers syndrome)
- Atherosclerotic coronary artery disease
Finally, Wisten et al [17] reviewed the frequency of SCD in 15- to 35-year-olds in Sweden from 1992 to 1999. This national database revealed 181 cases of SCD with an incidence of 0.93 cases/100,000 population per year. Preceding symptoms were present in half of the cases and included chest pain, dizziness, syncope, palpitations, and dyspnea. In this series, 73% of the subjects were male. The most common forensic diagnoses were: no structural abnormality (21%), coronary atherosclerosis (17.7%), dilated cardiomyopathy (12.2%), HCM (10.5%), and myocarditis (10.5%).

**Mechanism of sudden cardiac death**

Although there are several potential underlying causes of SCD (eg, coronary artery compression with ischemia, primary arrhythmia, rapid atrial conduction, trauma during vulnerable electrical phase), it is tempting to invoke ventricular fibrillation (VF) as the final common pathway for SCD. The literature suggests that VF is much less common in children and adolescents than in adults, however. Studies suggest that there is associated VF in approximately 10% to 20% of children and adolescents who have SCD, whereas in adults, more than 50% of SCD is associated with VF [18–20]. The reported incidence of VF in children and adolescents is likely to be underestimated. Many of the epidemiologic studies include infants who had sudden infant death syndrome (SIDS). Many patients who have SIDS may not have a primary cardiac cause for sudden death, and, hence, may not have VF. Most of the remainder of patients may have poorly-documented rhythms or rhythms that were documented late into their catastrophic event; either scenario may miss an earlier episode of VF. Although the true incidence of VF in children is not known, the usefulness of secondary prevention programs for SCD in children and adolescents with the use of community/school-

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<th>Post-operative congenital heart disease</th>
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<td>Transportation of the great arteries (after Mustard/Senning operation)</td>
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<td>Transportation of the great arteries (after arterial switch operation)</td>
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<td>Cardiac transplantation</td>
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*This entity may be familial*
based automated external defibrillators (AEDs) depends upon a higher incidence of VF than is described in the literature.

**Prevention of sudden cardiac death**

SCD in children and adolescents may be prevented by primary prevention programs, secondary prevention programs, or both.

**Primary prevention**

There seems to be theoretic rationale for the optimization of screening; this must include a detailed patient history and family history. Although several of the studies mentioned earlier retrospectively described symptoms in as many as 50% of the children and adolescents who had SCD, preparticipation screening with questionnaires and interviews have not been useful in prospectively uncovering patients who are at risk for SCD [21–23]. Detailed preparticipation sports questionnaires have been developed to address these issues. The efficacy of the questionnaires is variable and depends upon the emphasis and completeness of the questionnaire [21]. Therefore, the potential of such surveys to detect or raise the suspicion of cardiovascular abnormalities that are capable of causing sudden death in competitive student athletes is limited. The accuracy with which the questions are answered also is suspect, especially when the student-athlete understands that participation in competitive sports may be affected by the answers to the questions on the survey. For the child or adolescent who has no symptoms and whose family history is either unknown or noncontributory, the questionnaire is not an effective screening tool.

There has been recent interest in additional routine screening with ECG or echocardiography. The routine screening with ECG in Italy has decreased the frequency of HCM as a cause for SCD. Pilot studies that involve the use of screening echocardiography are currently underway in the United States (see discussion elsewhere in this issue).

**Secondary prevention**

Significant interest exists in school- and community-based AED programs that are targeted toward children and adolescents (see elsewhere in this issue).

**Summary**

SCD in children and adolescents, although less common than in adults, is devastating. It is fair to say, although it is not definitively known, that there are approximately 500 cases per year of unexplained SCD in children and adolescents per year in the United States. The causes are well well-known, although they may not be discovered prior to the episode of sudden death. It is probable
that the final common pathway for many, if not most, of the causes of SCD is VF. A challenge for the future will be the prospective identification of those children and adolescents who are at risk prior to an episode of SCD. Because the identification of all such children may not be possible, there is logic for the development of community/school-based secondary prevention/education programs. There may also be logic for the establishment of a national registry in order to track data that are related to unexpected SCD in children and adolescents in order to understand the true incidence of this problem and its related issues.

References

