Sudden Cardiac Death in Children and Adolescents

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KEYWORDS

• Pediatric • Arrhythmia • Sudden cardiac death • Sudden arrhythmic death syndrome
• Cardiomyopathy • Genetics

KEY POINTS

- Sudden cardiac death (SCD) is rare in childhood; the true incidence is not well understood.
- SCD etiologies in the young include anatomic and functional heart disease, primary arrhythmia syndromes, and other rare conditions.
- Sudden death can be the first symptom in a young person, although warning signs may be present: unexplained syncope or seizure, exertional chest pain or dyspnea, and a family history of early sudden death.
- Diagnosing disorders predisposing to pediatric SCD can be challenging; a strong index of suspicion and referral to experienced specialists are critical.
- Cascade screening of first-degree relatives is indicated after unexplained sudden death in individuals younger than 50 years or when a heritable cardiac condition is identified or suspected.

INTRODUCTION

"Sudden cardiac death has left no age untouched. Sparing neither saint nor sinner, it has burdened man with a sense of uncertainty and fragility."[1] Children, although commonly pictures of health, may be susceptible to sudden cardiac death (SCD). The definition is similar as for adults: sudden unexplained death in an otherwise healthy person within 1 hour of symptom onset, or unwitnessed death within 24 hours after seeing that person alive and well. In pediatrics, etiologies and challenges in SCD sometimes differ from adults.

SCD can be differentiated in the pediatric population based on age (**Table 1**), with variable etiologies. The younger the child, the less likely a cause of death will be identified with standard investigations.[2] When no etiology is found after comprehensive evaluation, including autopsy, toxicology, and histology, unexplained SCD is termed sudden arrhythmic death syndrome (SADS) or sudden unexplained death syndrome (SUDS).

Infant death during the first year of life is a distinct category that includes prenatal factors, congenital malformations, chromosomal abnormalities, premature births, and maternal, social, and environmental factors. An estimated 10% of sudden infant death syndrome (SIDS) is presumed due to an arrhythmia, predominantly congenital long QT syndrome (LQTS). The infant often carries a spontaneous, malignant mutation, typically without antecedent symptoms or known family history.

SCD is tragic for a family and community; there is high awareness of these uncommon but highly publicized events. A cardiac cause may be identified through a thorough postmortem evaluation.

Conflicts of Interest: None.

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Table 1
Pediatric sudden death terminology varies by age

SUID	Sudden unexplained infant death	Death in a child <1 y without obvious cause before investigation • Includes SIDS, accidental suffocation, strangulation in bed • Also called SUDI (sudden unexplained death in infancy)
SIDS	Sudden infant death syndrome	Death in an infant <1 y that cannot be explained after thorough investigation, including • Complete autopsy • Examination of the death scene • Review of the clinical history
SUDC	Sudden unexplained death in childhood	Death in a child >12 mo of age that remains unexplained after a thorough investigation, including • Complete autopsy • Examination of the death scene • Review of the clinical history

The most common pediatric SCD etiologies include congenital heart defects, cardiomyopathies, and inherited arrhythmia syndromes. Their recognition following a resuscitated sudden cardiac arrest (SCA) allows secondary prevention and family screening. Many have autosomal dominant inheritance and thus convey a 50% risk in first-degree relatives.

The true incidence of pediatric SCD is not well understood, limiting the ability to estimate risk and create policies for pre-event diagnosis and prevention. Identifying children at risk before a tragedy remains elusive for numerous reasons:

- Studying SCD in the young prospectively is difficult with rare events in disparate communities with varying reporting practices.
- Etiologies and incidence vary by age, requiring awareness and index of suspicion.
- Many diseases are silent until a sudden fatal event.
- Special pediatric populations raise complexity in evaluation and/or risk.
- Broad screening programs, although ostensibly detect children at risk and prevent SCD, remain in clinical equipoise with debated methods, efficacy, and outcomes.
- Postmortem evaluations vary in pediatric autopsies.
- Cascade screening after heritable SCD diagnosis is not always performed.

EPIDEMIOLOGY

The loss of a previously healthy child shocks a community; grief is propagated via news reports and social media, igniting fear and calls to action to prevent further tragedy. The emotional impact of a child's death is enormous, as are life-years lost. Despite the publicity, however, childhood SCD remains a relatively rare occurrence. According to the Centers for Disease Control and Prevention (CDC), heart disease in general accounts for far fewer childhood deaths than injuries, suicide, and malignancy (**Fig. 1**).[3]

Multiple studies have attempted to define the incidence of childhood SCD, estimated at 0.7 to 6.4 per 100,000 patient-years, far less than in adults.[2,4–6] In 1999, there were 5000 to 7000 childhood sudden deaths in the United States, compared with 300,000 to 400,000 adults,[4] whereas in 2005 the CDC estimated that 2000 people younger than 25 years die per year.[7] Approximately 60% to 75% of childhood SCD victims are male,[2,5,8–14] perhaps due to hormonal differences, level of athletic exertion, and other presumed variables.

One limitation in interpreting SCD studies in the young is lack of a uniform upper age limit. There is general consensus that infant death is a separate phenomenon; therefore, the lower cutoff is 1 year. Studies loosely define young people as those younger than the age at which atherosclerotic disease predominates, and thus includes individuals up to 30 to 45 years.

Existing data are largely based on regional populations in areas with disparate tracking of sudden deaths, thus affecting the incidence numerator. Most regions lack comprehensive health records with mandatory reporting of sudden deaths and rely on multiple and often less reliable sources: media and Internet searches, catastrophic