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Title page

Title: Sudden Cardiac Death in Childhood: Peaks in teenagers.

Running title: Sudden Cardiac Death in Children peaks in Teens

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We have no conflicts of interest to declare.

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Nonstandard Abbreviations and Acronyms

CHD: congenital heart disease

SADS: sudden arrhythmic death syndrome

SCDC: sudden cardiac death in children

Text

Sudden cardiac death in children(SCDC) is a rare but devastating event estimated to occur at 0.7-10.1 per 100,0001,2. Previous studies have highlighted sudden arrhythmic death syndrome(SADS), and cardiomyopathy as predominant causes1,2. These causes are genetic and have major implications for surviving family. We aimed to assess the age, gender and causes of SCDC from a specialist UK national cardiac pathology referral centre.

This is a descriptive cohort study conducted in the Cardiac Risk in the Young Cardiovascular Pathology centre based in St. George’s, University of London with IRB and ethical approval (10/H0724/38). Supporting data is available upon request. The centre receives referrals from the UK where a cardiac cause of death is suspected or no cause is found at the original autopsy. We acknowledge that there may be a referral bias toward more complex cases. All referrals underwent full autopsy and toxicology to exclude non-cardiac causes. SCDC was defined as death within 24 hours of being well in individuals aged >1-17. Information including symptoms were provided by referring coroners, family questionnaires and general practitioners. Hearts were assessed according to guidelines as previously published3. Categorical data are presented as number(%) and continuous data as mean±SD.

Between 2013 and 2022, there were 791 deaths of children with cardiovascular codes and we received 415(52%) cases of SCDC. There were 8108 referrals of sudden cardiac death between 1994-2023 and 624(8%) were SCDC. Average age is 12±5years with 413 males and 211 females(2:1). Figure 1 shows the age referral pattern by sex. SCDC peaks at age 1 and between 13-17. SCDC in females starts to increase from 9 whereas male SCDC starts to increase from 10. Death occurred associated with exertion in 128(21%). Cardiac symptoms were reported in 71(11%) with syncope/seizure/convulsion in 42(7%), palpitations in 14(2%), breathlessness in 11(2%), and chest pain in 10(2%). Abdominal pain was reported in 12(2%).

SADS(393, 63%), where the heart is morphologically normal, is most common in all age groups followed by cardiomyopathy(97, 15%), myocarditis(29, 4%) and congenital heart disease(CHD, 27, 4%). Anomalous coronary artery(16, 2%), valve disease(13, 2%), commotio cordis(11, 2%), and idiopathic infarction(11, 2%) made up similar proportions. Rarer causes include cardiac tumour(5, 1%), Wolff-Parkinson-White(4, 1%), ischemic heart disease due to coronary artery atheroma(2, <1%), vasculitis(2, <1%), transplant vasculopathy(2, <1%) and hypertensive heart disease (1, <1%).

Cardiomyopathies include hypertrophic cardiomyopathy(25, 4%), arrhythmogenic cardiomyopathy(22, 4%), idiopathic fibrosis(14, 2%), idiopathic hypertrophy(12, 2%), dilated cardiomyopathy(10, 2%) and metabolic cardiomyopathy(5, 1%).

SADS increased as a proportion and CHD decreased as a proportion of SCDC with increasing age. Cardiomyopathy appears prominent at age 1 and older age group. Myocarditis made up a higher proportion of SCDC between 2-12 years. Commotio cordis only appears in older children active in sport.

In this study SCDC peaks at age 1 and between 13-17. SCDC is very rare between 2-8 and increases dramatically in teenagers. This may suggest an influence of hormonal change raising the risk of SCDC especially in SADS and cardiomyopathy. During the pubertal growth phase, cardiac enlargement and increasing blood pressure place additional demand upon the heart which may result in an increased risk of arrhythmia and SCDC4. Alternatively, these deaths may be related to factors such as increased participation in strenuous sports or disease progression as in arrhythmogenic cardiomyopathy5.

There has been increasing interest in SCDC with link to heritable channelopathies and cardiomyopathies, risk stratification, genetic testing and management. The nationwide Danish study reported 87 SCDCs which agreed with main causes being SADS and cardiomyopathies. Our study reported less cardiac symptoms(11% vs 59%), but this is partially explained by the fact that they reported on all symptoms including stomach pain, back pain and nausea1. Stomach pain was their most common prodromal symptom however this reported at a lower frequency in our cohort(2% vs 12%). We report a higher proportion of SCDC associated with exertion(21% vs 14%). They had more CHD in their cohort(4% vs 20%). In contrast to our study and another UK study2, they found no hypertrophic cardiomyopathy.

A recent UK study of 151 SCDC from two paediatric centres had similar findings but contrasting found more CHD than cardiomyopathy which may reflect specialist paediatric referral practice and including infants2. Full autopsy and cardiac examination in all cases of SCDC is essential in order to make the correct diagnosis.

In this cohort study, a larger proportion of SCDC were among teenagers raising a potential association with age. Most SCDC is not associated with exertion(21% with exertion) or reported cardiac symptoms(11% with symptoms). In this cohort, SADS(63%) and cardiomyopathy(15%) accounted for a majority of SCDC, suggesting heritable conditions were the main cause of SCDC. Thus, families in which an inherited cardiac condition is identified in a child, need screening to detect and treat these conditions to prevent further deaths. Based on the findings in this cohort, related children should be screened prior to 13.

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Disclosures

There are no conflicts of interest to disclose.

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Figure 1 legend: Age, sex and causes of sudden cardiac death in children. Of 7464 sudden cardiac deaths, 624 (8%) were children. Sudden cardiac death in children peaks at age 1 and between 13-17. Sudden arrhythmic death syndrome (SADS) increased as a proportion and congenital heart disease (CHD) decreased as a proportion of SCDC with increasing age. Cardiomyopathy (CM) appears prominent at age 1 and older age group. Myocarditis made up a higher proportion of SCDC between 2-12 years. Commotio cordis (CC) only appears in older children active in sport. (ACA=coronary artery abnormality; II=idiopathic infarction).

For twitter:

Sudden cardiac death in children peaks in teens caused by heritable cardiomyopathy and SADS. Cardiological relative screening should be prior to 13.