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Autopsy results and factors associated with sudden cardiac death in young individuals with congenital heart disease – a nationwide study

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ABSTRACT

Objectives: Sudden cardiac death (SCD) is a leading cause of mortality among individuals with congenital heart disease (CHD), and risk stratification remains challenging. This study aimed to describe the underlying structural cardiac abnormalities in a national cohort of SCD victims with CHD, their socioeconomic status, and interactions with the healthcare system before death. **Methods:** The Swedish study of Sudden Cardiac Death in the Young, 2000–2010, included SCD victims under 36 years, along with population-based controls and their parents. Of 903 SCD victims, 39 with autopsy-defined CHD were included in this study, together with 195 controls. Information on socioeconomic variables and healthcare contacts was gathered from Swedish national registers. **Results:** The median age for SCD was 24 years, and 64% were male. The CHD was undiagnosed before death in 31% of the cases, of whom 8 had coronary anomalies. Moderate to complex CHD was observed in 41%. Structural abnormalities of the ventricles were prevalent, with left ventricular hypertrophy present in 56% and fibrosis in 64%. The cases had a higher frequency of hospital admissions within 6 months before SCD compared to controls (OR 14.1,95% CI 3.80–52.44), $p < 0.001$. No socioeconomic differences were observed. **Conclusions:** This study identified a broad spectrum of underlying anatomical defects, with ventricular structural abnormalities being a common autopsy finding. The majority of cases had moderate to severe lesions. An increased frequency of healthcare contacts prior to death was noted, which may be a variable needing more attention as a predictor for a higher risk of SCD.

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Introduction

In recent decades, the prevalence of adults with congenital heart disease (CHD) has risen, attributed to advancements in surgical techniques, medical care, and enhanced follow-up systems. Sudden cardiac death [1] (SCD) is a leading cause of mortality among individuals with CHD [2,3]. The annual incidence of SCD in the CHD population ranges from 0.28 to 2.7 per 1,000 patient-years, influenced by factors such as study design, age range, and lesion definitions [2,4].

The underlying causes of SCD in young individuals with CHD have been explored in numerous studies, revealing that multiple pathophysiological processes and triggers are involved [4,5]. The primary cause of SCD in these patients is malignant arrhythmia [6], with structural abnormalities increasing the risk. Some congenital heart defects are associated with a higher risk [7], although this risk spans the entire spectrum of CHD. Non-arrhythmic causes, such as myocardial infarction, pulmonary embolism, and aortic dissection, are less common [2,3,7]. Additionally, SCD can sometimes be the first presentation of an underlying congenital heart defect [4,8]. However, the specific characteristics that put certain individuals with CHD at higher risk remain unclear.

A Swedish study of SCD in young individuals (SUDDY) [1] demonstrated that those who experienced sudden death exhibited more symptoms and had more frequent healthcare interactions compared to controls. Socioeconomic status (SES) is associated with cardiovascular disease, and a low educational level, often used as a marker for SES, has been linked to less favourable outcomes [9–11]. The connection between SES and CHD-related mortality, as well as the relationship between low maternal education and increased infant mortality, has also previously been documented [12,13].

This study aimed to characterise a national cohort of young SCD victims with CHD, emphasising the prognostic significance of underlying structural cardiac abnormalities, the frequency of medical contact, and socioeconomic status prior to death.

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Method

Definition of the study individuals

Our study group consisted of participants from the SUDden cardiac Death in the Young cohort [1], which identified 78 individuals with CHD and SCD over an 11-year period (2000–2010). From this group, 39 individuals who had undergone a post-mortem examination (either clinical or forensic autopsy) and for whom reports were available were included in our study. For each of these 39 individuals, we confirmed the underlying CHD diagnosis and the cause of SCD by reviewing death certificates and autopsy protocols, as illustrated in Figure 1.

Structuring of autopsy reports and definition of variables

Forensic and clinical autopsies were conducted at various medical centres in Sweden using a gold-standard approach [14]. A structured review of the autopsy reports systematically extracted anatomical variables. These variables were selected based on clinical experience regarding the impact of structural heart abnormalities.

Structural malformations, including ventricular hypertrophy and dilatation, as well as fibrosis and signs of acute inflammation or infarction, were recorded. If a heart exhibited multiple structural malformations, each was documented separately. Valvular defects and abnormalities in the great arteries were also recorded and graded as mild, moderate, or severe. If a structure was described as normal or if a specific pathology was ruled out in the report, it was categorised as normal. Any missing information about a specific structure was labelled as missing data.

The congenital pathologies identified in the autopsy reports were categorised into seven groups based on the predominant structural abnormality to maintain anonymity and minimise recognition. These groups included atrial septal defect (ASD), ventricular septal defect (VSD), VSD with other pathology, aortic stenosis, aortic stenosis with other pathology, mitral valve pathology, and other lesions. The "other lesions" category encompassed coronary anomalies and other moderate to severe lesions [7] not classified elsewhere, as shown in Figure 2.

Collection of controls

In the SUDDY study, five controls from the general population, selected by Statistic Sweden (SCB), were included for each case. Data were also collected from the parents of both cases and controls [1]. Our study involved a comparison of the individuals with 195 population-based controls, with each case matched to five controls based on sex, age, and residential area.

Registers and study period

The National Board of Health and Welfare in Sweden administers several national registers to support the development of healthcare and social services. National quality registers, maintained by healthcare professionals, enable the monitoring of care quality and outcomes. All Swedish residents, whether born in the country or immigrating, are assigned a unique 10-digit personal identification number. This number facilitates the linking of data from various registers, databases, and medical records. For this study, we retrieved data from the National Patient Register (NPR) and the Swedish Prescribed Drug Register (SPDR) using the personal identification numbers of both study participants and controls. Additional data were obtained from the Longitudinal Integrated Database for Health Insurance and Labor Market Studies (LISA), managed by Statistic Sweden, as well as from the SWEDCON registry, which contains information on patients with CHD. The study period was defined as 180 days before the date of death (the reference date) for study participants, with the same time interval applied to the controls.

The Swedish longitudinal integrated database for health insurance and labor market studies (LISA)

LISA is a register of socioeconomic variables that has been available since 1990 for all individuals aged 16 and older residing in Sweden. For this study, we extracted data on the highest completed educational degree for both study participants and controls from the year prior to the reference date. Educational attainment was categorised into three levels: compulsory school, upper secondary school, and university. We also recorded the highest educational level attained by the parents of both cases and controls. To assess the marital status of the parents, we utilised the family ID created by the LISA register. If the family IDs for both parents matched, their marital status was classified as cohabiting; if they differed, it was classified as non-cohabiting.

The Swedish National Patient Register (NPR)

The NPR register contains data on hospital admissions since 1987 and outpatient visits since 2001. For this study, we collected information on whether individuals had been admitted to a hospital or visited an outpatient clinic during the 180-day study period. We also gathered data on hospital admissions and outpatient visits attributed to cardiovascular causes, as indicated by the primary diagnosis using ICD-codes, detailed in Supplemental Table 1.

The Swedish Prescribed Drug Registry (SPDR)

Established in 2005, the SPDR contains data on dispensed medications. We utilised ATC-codes from the Anatomical Therapeutic Chemical classification system, as outlined in Supplemental Table 2, to gather information on cardiovascular medications dispensed during the reference period. If no registration was found, it was assumed that the individual was not receiving cardiovascular therapy. If the reference date fell before the establishment of the registry, it was set as missing information.

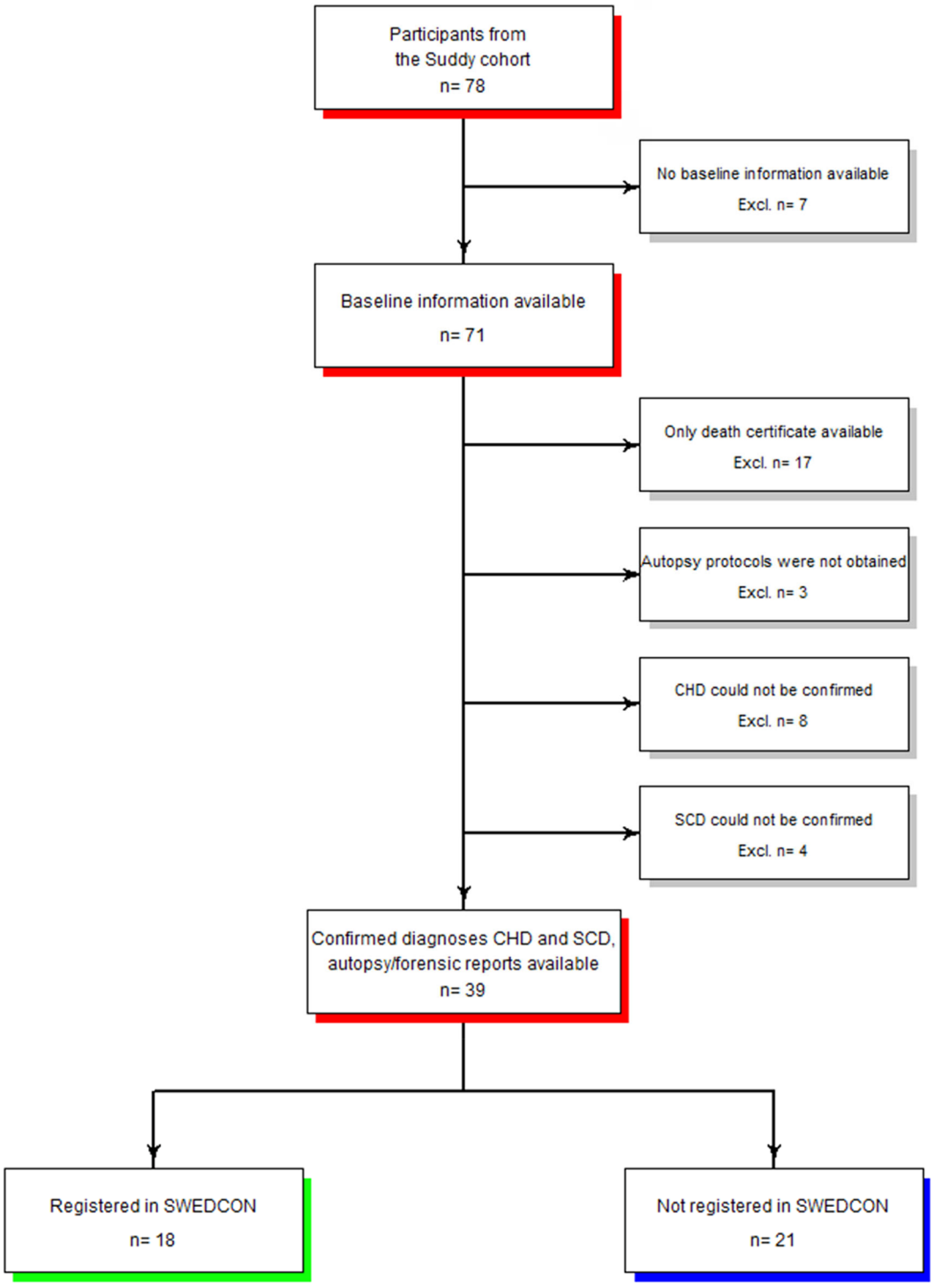

Figure 1. Flow chart of the inclusion process. The SUDDY cohort contained 78 sudden cardiac death (SCD) victims in Sweden with congenital heart disease (CHD) between the years 2000 and 2010. Death certificates, autopsy and forensic reports of these individuals had been collected within the scope of that study. Finally, 39 participants from the SUDDY cohort with available autopsy reports and confirmed CHD and SCD diagnoses were included in this study. The study individuals were later divided according to being registered or not in the national quality register for CHD, the SWEDCON register.

The Swedish registry of congenital heart disease (SWEDCON)

Since 1998, cardiologists managing adult patients with CHD have been able to register their patients in SWEDCON. The registry includes detailed information on outpatient visits for patients over the age of 18 years, encompassing medical history and clinical status. A paediatric section of the registry has been available since 2009. We searched the registry to determine if the study individuals were registered. For those who were, we extracted data from their last visit, including NYHA classification, QRS-width from a standard 12-lead ECG, blood pressure, and weekly exercise level.

Ethical statement

Data about the study participants were integrated with the SUDDY cohort database. This research was approved by the

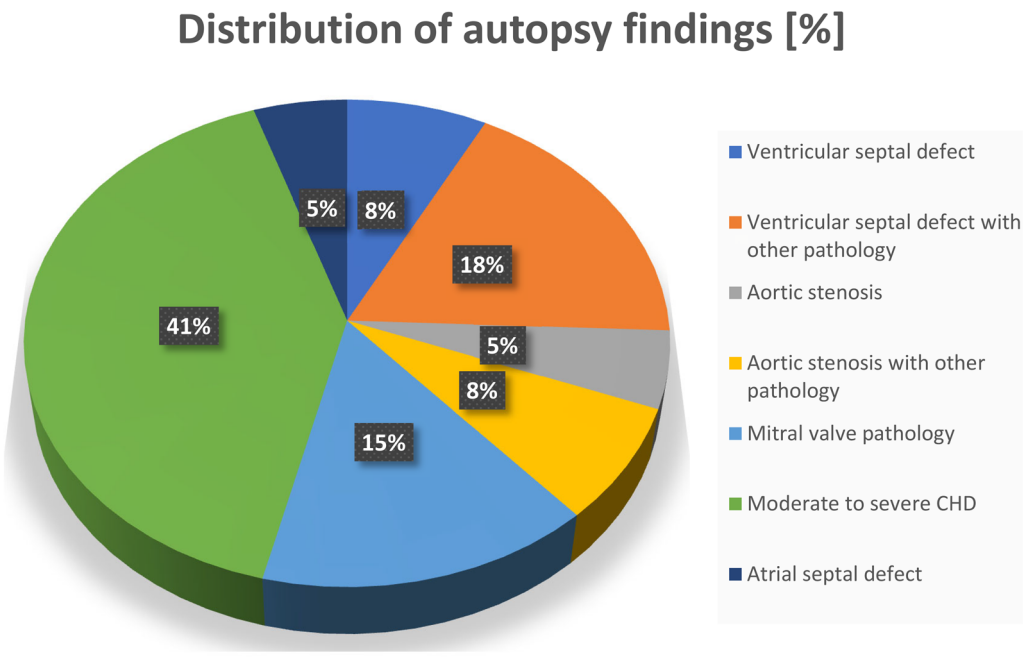

Figure 2. The underlying pathologies, according to the autopsy protocols divided into seven main groups: ventricular septal defect (VSD) ($n = 3$), VSD with other pathology ($n = 7$), atrial septal defects (ASD) ($n = 2$), aortic stenosis ($n = 2$), aortic stenosis with other pathology ($n = 3$), mitral valve pathology ($n = 6$) and moderate to severe lesions, including coronary anomalies ($n = 16$).

Swedish Ethical Review Authority in Uppsala/Umeå, with approval numbers 2012-161-31 M/2017-431/2017-430, 2014-156-32 M and 2020-06095.

Statistics

Descriptive statistics were used for baseline characteristics. Conditional logistic regression models, accounting for the matching of cases and controls, were applied to analyse the association between having a congenital heart defect and suffering from sudden cardiac death, as well as the association with socioeconomic status (SES) and morbidity. A chi-square test was used to compare autopsy findings between cases registered in SWEDCON and those not registered. For all analyses, a two-tailed P-value < 0.05 was considered statistically significant. All analyses were done in R [15].

Language review assistance

An AI/machine learning tool, ChatGPT 4o [16], was used for language review and spelling of the manuscript. The authors reviewed and edited the content after using the tool and take full responsibility for the content of the article.

Results

Baseline information of the study individuals

The study cohort consisted of 39 individuals with congenital heart lesions, including 14 women (36%) and 25 men (64%). Eighteen (46%) of these individuals were registered in SWEDCON, comprising 8 women and 10 men. The distribution of age at death is shown in Figure 3. Fifteen individuals had at least one outpatient visit recorded in SWEDCON. However, none had complete data on all parameters of interest from their last recorded visit. NYHA class was registered in 11 individuals; 8 were classified as NYHA class I, while the remaining were classified as NYHA class III. Weekly exercise, documented in 12 individuals, was reported as none for 7 individuals, less than 3 h for 2 individuals, and more than 3 h for 3 individuals. The mean systolic blood pressure was 127 mmHg (range 100–180 mmHg), and the mean diastolic blood pressure was 80 mmHg (range 60–115 mmHg) ($n = 12$). The mean ECG width recorded was 128 msec (range 90–158 msec) ($n = 10$).

Autopsy results

All of the 39 study individuals underwent an autopsy. A forensic autopsy was conducted in 26 cases (67%), while a clinical autopsy was performed in 13 cases (33%). Forensic autopsies were more frequently performed in individuals without registration in SWEDCON compared to those who were registered ($p = 0.002$). The distribution of underlying pathologies, categorized into the seven main groups of structural abnormalities, differed between individuals registered in SWEDCON and those not registered, as shown in Supplemental Table 3. In 12 cases (31%), the congenital heart defect was diagnosed post-mortem. In this group, coronary anomalies were identified in 8 individuals; 3 had valve anomalies, and 1 was diagnosed with Ebstein's anomaly.

Autopsy results; structural abnormalities

The distribution of congenital structural abnormalities observed during autopsy is illustrated in Figure 2. The most common

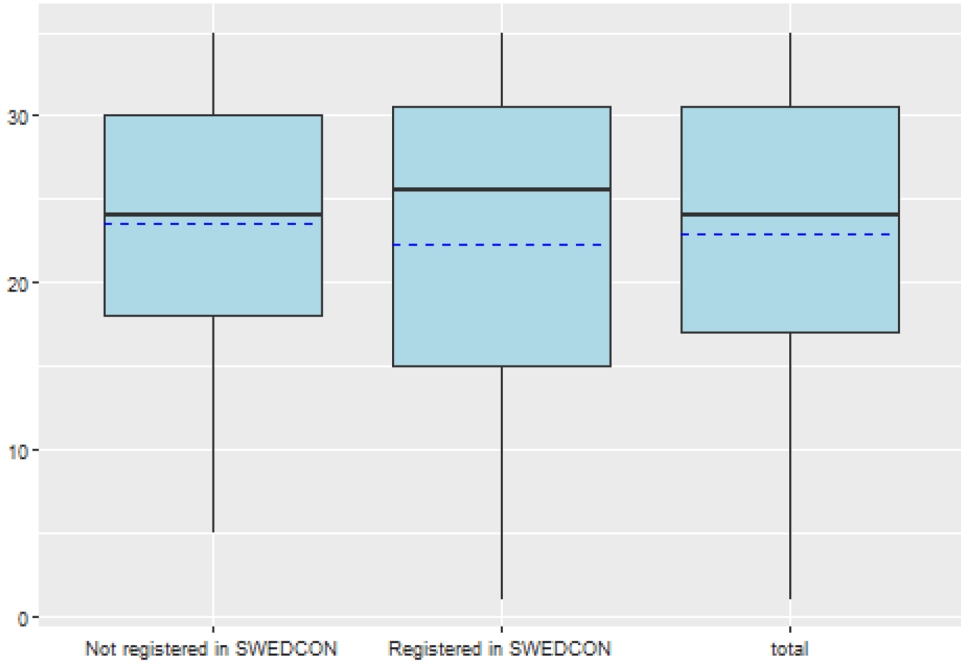

Figure 3. The distribution of age at death, overall and for those registered in SWEDCON and not registered, respectively. The horizontal lines within each box represent the median, the dotted line represents the mean, and the upper and lower edges of the box represent the first and third quartiles. The whiskers extend to 1.5 times the interquartile range.

Table 1. Structural abnormalities of the heart found at autopsy for the group registered in SWEDCON and not. Significant p value ($p < 0.05$) is marked with an asterisk in the table.

	Total	Not registered in SWEDCON ($n = 21$)	Registered in SWEDCON ($n = 18$)	p-value
Left ventricular dilatation, %	41	48	33	0.136
Right ventricular dilatation, %	36	33	39	0.238
Left ventricular hypertrophy, %	56	48	67	0.252
Right ventricular hypertrophy, %	36	10	67	<0.001*
Heart weight, g (mean ± SD)	447 ± 178	413 ± 153	490 ± 203	0.165
Acute inflammation, %	41	33	50	0.263
Fibrosis, %	64	71	56	0.248

finding was moderate to severe heart lesions, identified in 16 cases (41%). In 2 of these cases, there was also aortic valve pathology, including severe fibrosis and a bicuspid valve; however, these were categorised within the moderate to severe lesions group due to other underlying complex conditions. A mechanical valve was included in the group of aortic stenosis with other pathology. In addition to the six cases with mitral valve defect, a more discrete mitral valve pathology was noted in three individuals who had additional pathologies, leading to their classification in other groups. No cases of mitral valve stenosis were recorded. Pathologies affecting the tricuspid and pulmonary valves were less common. The pulmonary valve was affected in three cases, all of which were registered in SWEDCON. The tricuspid valve was affected in 5 cases, with 4 out of 5 also registered in SWEDCON. Seven specific phenotypic patterns could be classified in 12 cases (31%), shown in Supplemental Table 4.

Specific autopsy results

A thrombus was identified in 5 cases, with 3 located within the ventricle and 2 found in the pulmonary artery and the basilar artery in the brain.

Left ventricular hypertrophy was the most prevalent structural abnormality, occurring in 22 cases (56%). This was the only structural abnormality more common than having a normal myocardium. Left ventricular dilation was observed in 16 cases (41%), while right ventricular hypertrophy was noted in 14 cases (36%), with right ventricular dilatation also found in 14 hearts (36%). Fibrosis was present in 25 cases (64%), and acute inflammation or infarction was seen in 16 cases (41%).

There was a difference in the proportion of right ventricle hypertrophy between individuals registered in the SWEDCON register and those who were not ($p < 0.001$, Table 1).

The median weight of the hearts was 422 g (IQR 350.5 g, 599,3 g) ($n = 34$). The minimum weight recorded was 73,7 g, while the maximum weight was 800 g.

Abnormal anatomy of the coronary arteries was found in 14 cases (36%), with pathologies ranging from thin arteries to completely missing arteries. Coronary anomalies were the sole defect in 7 of the 14 cases, while the remaining 7 cases had coronary anomalies in conjunction with other pathologies; of these, 4 were registered in SWEDCON. Narrowing of the coronary arteries of any degree was noted in 10 cases (26%), with 3 cases exhibiting more advanced atherosclerosis. Atherosclerosis of the aorta was found in 11 cases (28%), while hypoplastic aorta and necrosis in the aortic wall were described in 7 and 1 cases, respectively. Additionally, 7 cases (18%) had anatomical abnormalities in the pulmonary artery.

Hospitalisation and outpatient visits

Within the 180 days prior to death, study individuals experienced a significantly higher frequency of hospital admissions, with 23% compared to 2% among controls (OR 14.1, 95% CI 3.80–52.44, $p < 0.001$). Cardiovascular disease was the reason for admission in 5 cases (13%), including 1 admission due to ischemic heart disease, 1 due to cerebrovascular disease, and 3 due to "other heart disease." Only 1 cardiovascular outpatient visit was recorded among the cases, and no admissions or outpatient visits for syncope were found. Notably, none of the controls had any medical contact for cardiovascular causes within the reference period, as shown in Table 2.

Dispensed drugs

There was a significant difference between the two groups in the dispensing of cardiovascular drugs during the reference period (OR 7.1, 95% CI 1.22–40.84, $p = 0.025$). The total number of dispensed cardiovascular drugs was 12 for the study group compared to 3 for the control group. In the study individuals, the dispensed cardiovascular drugs were categorised as follows: platelet aggregation inhibitors ($n = 2$), anticoagulation therapy ($n = 3$), diuretics ($n = 3$), beta-blocking agents ($n = 1$), ACE-inhibitors/ARBs ($n = 1$), and other cardiovascular therapies ($n = 2$). In contrast, the control group received beta-blocking agents in 2 cases and ACEI/ARB inhibitor in one case.

Socioeconomic variables

The distribution of educational levels among individuals over 18 years, as shown in Table 3, was similar between study individuals and controls ($p = 0.43$). The majority of study individuals had secondary upper school as their highest level of education, which was also the case for the control group (OR 0.87, 95% CI 0.29–2.61).

The educational level distribution remained comparable between study individuals and the control group when including all individuals ($p = 0.43$), with both groups predominantly holding secondary upper school qualifications (OR 0.87, 95% CI 0.29–2.61).

The distribution of educational levels among the parents of study individuals and controls showed no significant difference ($p = 0.88$). The majority of parents in both groups had secondary upper school as their highest level of education, with 49% for study individuals and 52% for controls (OR 0.75, 95% CI 0.25–2.25).

There was no significant difference in the distribution of cohabiting vs. non-cohabiting parents between the groups ($p = 0.22$), with most parents of both study individuals and controls cohabiting (OR 0.63, 95% CI 0.30–1.32).

In the subgroup of individuals over 18 years, there were no differences in either parental educational level ($p = 0.78$) or in the distribution of cohabitation status ($p = 0.12$).

Discussion

In this study on sudden cardiac death in young individuals with CHD, autopsy results revealed a wide range of defects. Left ventricular hypertrophy was a common structural abnormality, and most defects were classified as moderate to severe lesions. Notably, in 31% of cases, SCD was the first indication of an underlying CHD. The study individuals exhibited increased healthcare contact before death; however, no significant differences in socioeconomic status were found compared to controls.

Table 2. Hospitalisation and outpatient visits within 180 days before reference date, and prescribed drugs within the same period. Significant p-values ($p < 0.05$) are marked with asterisks in the table.

	Study group $n = 39$	Control group $n = 195$	OR	95% CI	p value
Admission to hospital, any cause, *n* (%)	9 (23)	4 (2)	14.1	3.80–52.44	<0.001*
Admission to hospital, cardiovascular cause, *n* (%)	5 (13)	0 (0)			
Outpatient visit, any cause, *n* (%)	13 (38)	23 (14)	3.6	1.61–8.11	0.002*
Outpatient visit, cardiovascular cause, *n* (%)	1 (3)	0 (0)			
Any medical contact, *n* (%)	17 (50)	25 (15)	5.5	2.46–12.40	<0.001*
Any medical contact, cardiovascular cause, *n* (%)	6 (17)	0 (0)			
Any prescribed cardiac therapy, *n* (%)	4 (27)	2 (3)	7.1	1.22–40.84	0.025*

Table 3. Educational level of the study individuals, controls and their parents, as well as marital status of their parents.

	Study group $n = 39$	Control group $n = 195$	OR	Conf int	p-value
Educational level, individuals >18 years					0.43
Compulsory school, *n* (%)	7 (25)	28 (20)	1		
Secondary upper school, *n* (%)	16 (57)	72 (52)	0.87	0.29–2.61	
University, *n* (%)	5 (18)	38 (28)	0.45	0.11–1.84	
Educational level, parents					0.88
Compulsory school, *n* (%)	5 (14)	20 (11)	1		
Secondary upper school, *n* (%)	18 (49)	95 (52)	0.75	0.25–2.25	
University, *n* (%)	14 (38)	69 (38)	0.83	0.25–2.69	
Marital status, parents					0.22
Non-cohabitant, *n* (%)	17 (47)	67 (37)	1		
Cohabitant, *n* (%)	19 (53)	115 (63)	0.63	0.30–1.32	

Cohort composition

The age range of the study group, which spans 34 years and shows a predominance of males, aligns with previous findings, indicating that the risk of SCD exists across all ages and primarily affects men [1]. Our study also shows that SCD occurs throughout the whole span of CHD but is more prevalent among moderate to complex lesions, which is line with previous research [4,5, 8,17].

Risk assessment by clinical parameters registered in SWEDCON

Predicting the risk of SCD in patients with CHD is challenging. While lesions-specific algorithms provide some guidance, no general algorithms exist to predict risk, primarily due to the heterogenicity of congenital heart defects [2,6].

Previous studies have shown that a broad QRS-complex is associated to SCD in certain forms of CHD [18], with individuals who experience SCD having a wider QRS-complex (greater than 140 ms) compared to controls [2]. In our study, however, the mean QRS duration was only 128 ms, and none of the individuals exceeded 160 ms on their last registered ECG. This underscores the complexity of identifying specific predictors for SCD in CHD. Some studies indicate that changes in QRS width may serve as a more reliable predictor of SCD [2,19], although further research is necessary to confirm this relationship. Additionally, self-reported exercise capacity may not accurately reflect the limitations of individuals with CHD, as they often underestimate their symptoms [20,21]. Adaptation to the hemodynamic changes associated with their heart defect, along with slow disease progression, may explain why the majority of study individuals were classified as asymptomatic (NYHA class 1) during their last outpatient visit before SCD. However, CHD patients in NYHA class 1 have been shown to have a lower peak VO2 compared to matched controls, and impaired exercise capacity is associated with increased risk of hospitalisation and death [21]. Therefore, even those classified as NYHA class 1 should be considered for SCD risk assessment.

In this study, 46% of the individuals were registered in SWEDCON. The cohort included coronary anomalies, which often remain undiagnosed prior to death and thus are not recorded in SWEDCON. There was an overrepresentation of right ventricle hypertrophy in the SWEDCON group ($p < 0.001$). Most tricuspid valve pathologies and all pulmonary valve pathologies were also registered in this group, indicating that more advanced CHD cases were registered in SWEDCON during the study period. Individuals not registered in SWEDCON may not have received care at medical centres that had access to the register, though registration has become more widespread over time.

Autopsy result in sudden cardiac death cases

In CHD, long-term hemodynamic effects on the myocardium can lead to fibrosis and structural changes in both surgically corrected and uncorrected cases, potentially serving as substrates for malignant arrhythmias [8]. Although autopsies reveal these myocardial changes, they rarely pinpoint the exact cause of death [22]. In this study, we assumed that death was related to the underlying CHD unless indicated otherwise. Thrombosis was found in only a few cases involving the heart, great vessels, or brain, which might suggest alternative causes of death [3,4].

The prevalence of coronary anomalies ranges from 0.21 to 5.79% and has been associated with myocardial ischemia and SCD [23]. Previous studies indicate that coronary anomalies account for a small percentage of SCD cases in young individuals, particularly among athletes [8]. An overview of sudden cardiac death in young US individuals highlights coronary anomalies as a common cause of SCD and states that most deaths related to coronary anomalies occur during exercise, and approximately half of the individuals experience symptom weeks prior death [24]. A Danish study found that coronary anomalies were responsible for 6% of SCD cases in this population [22]. While our study observed a higher percentage of coronary anomalies, potentially due to broader inclusion criteria and small sample size, it supports the notion that these anomalies are a common cause of SCD in young individuals with CHD, often diagnosed post-mortem.

Most SCD victims in our study exhibited left ventricular hypertrophy and fibrosis. Previous studies, particularly those focused on Tetralogy of Fallot (ToF), have identified extensive fibrosis as a necropsy finding that may contribute to malignant arrhythmias [25]. Left ventricular fibrosis has also shown to be a common autopsy finding among SCD-CHD cases from a small cohort in Australia [17]. Structural myocardial changes can lead to ventricular dysfunction, thereby increasing the risk of lethal arrythmias. Routine follow-up care for CHD patients typically includes echocardiography and magnetic resonance imaging to detect ventricular hypertrophy and focal myocardial fibrosis [26]. Fibrosis detected in the right ventricle using late gadolinium enhancement (LGE) has been shown to predict arrhythmias [27], indicating that cardiac remodelling and fibrosis are important factors in SCD risk assessment.

Despite the young age of our study population, atheromatous changes were noted in both the aorta and coronary arteries in several cases. Previous small studies have shown intimal hyperplasia in the coronary arteries of young CHD patients, suggesting early atherosclerosis [28–30]. Multiple pathophysiological mechanisms contribute to coronary artery pathology in CHD patients, and evidence indicates that these individuals face an increased risk of premature acquired cardiovascular disease [30,31]. Moreover, modifiable risk factors like obesity and hypertension are more prevalent among those with CHD [31]. Thus, it is crucial for caregivers to recognise this heightened risk and to take proactive steps to mitigate any adverse conditions.

Morbidity correlated to sudden cardiac death

This study demonstrates greater morbidity among the SCD group compared to controls, aligning with findings from other studies [1,22,24]. This supports the theory that symptoms indicative of deteriorating hemodynamic function may arise before SCD, potentially aiding in the identification of at-risk

patients [7]. Additionally, the results indicated increased healthcare contact for all medical causes, suggesting that any decline in health may precede SCD. The higher percentage of prescribed medications in the SCD group further reinforces the idea of increased morbidity. Chromosomal abnormalities, copy number variations and chromosome rearrangement are associated with CHD and also to SCD [32–34]. Chromosomal abnormalities but also pathology in single genes can be associated with both syndromic and non-syndromic CHD forms [4]. Extracardiac malformations, might be a reason for the CHD patients to seek healthcare in different departments, and knowledge of the increased risk for SCD is important for all caregivers. Syndromic CHD makes this deterioration in health even more difficult to notice. To deepen the understanding of underlying genetic abnormalities in patients with CHD might help in reducing the risk for SCD [32].

Socioeconomic status impact on sudden cardiac death

This study highlights socioeconomic status (SES) as a potential factor in assessing SCD risk among CHD patients. While the association between low SES and SCD has been investigated, the relationship between CHD, low educational attainment, and SCD remains underexplored.

Children with CHD are known to face risks for poorer neurocognitive outcomes [35–37], even in less severe defects [38]. Previous research has shown inconsistent findings regarding educational achievement in CHD patients, likely reflecting the heterogeneity of the conditions [36]. Some studies have linked CHD to lower educational achievements and shown that educational level can correlate with the severity of the defect [36,39,40]. Our results suggest a trend toward lower educational levels among CHD patients compared to controls, particularly at the university level, though these differences were not statistically significant.

Earlier studies have indicated that SES is a risk factor for mortality among children with CHD, with lower maternal educational levels associated with poorer survival [13]. In line with these findings, our results indicate a trend toward lower educational levels among parents of SCD victims compared to controls, although the difference was not statistically significant. Gaining a deeper understanding of both the educational level of patients and their parents could enhance risk assessment for sudden cardiac death.

Additionally, cohabitation has been suggested as a protective factor for children with CHD [41]. While our study did not find significant differences in parental cohabitation, the odds were lower for parents of SCD victims. The absence of parental cohabitation might therefore represent a risk factor worth considering for younger patients. Further research is needed to explore the socioeconomic factors influencing SCD risk in individuals with CHD.

Strength and weaknesses

This study is, to our knowledge, one of the few to investigate socioeconomic factors in relation to SCD risk among CHD patients. It also adds to the literature on autopsy findings in this population by offering additional data on structural heart abnormalities. However, there are limitations to consider: the National Patient Register (NPR) includes hospital activity only since 1987 and outpatient data since 2001, while the SWEDCON registry does not provide complete coverage, which may result in missing data. Additionally, the small sample size constrained the statistical power of the analysis.

Conclusions

In conclusion, this national study highlights the multifaceted factors influencing the risk of SCD in individuals with CHD. We found a wide range of underlying anatomical defects among young individuals who experienced SCD, with most exhibiting structural abnormalities in their ventricles. Notably, SCD was the first presentation of an underlying cardiac defect in almost one-third of cases, with coronary anomalies being the predominant findings. Furthermore, these young individuals with CHD demonstrated increased healthcare contact before their deaths, underscoring the importance of vigilant monitoring and risk assessment in this vulnerable population.

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Disclosure statement

No potential conflict of interest was reported by the author(s).

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Data availability statement

The data that support the findings of this study are available from the corresponding author upon reasonable request.

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