Sudden cardiac death in young people

What is sudden cardiac death?

Sudden cardiac death (SCD) is an unexpected death due to cardiac causes occurring in a short time period (generally within one hour of the onset of symptoms) in a person with known or unknown cardiac disease. Most cases of SCD are related to cardiac arrhythmias.

SCD may also be caused by non-cardiac conditions - eg, pulmonary embolism.[1]

How common is sudden cardiac death? (Epidemiology)[2] [3]

- Most studies report an incidence of sudden cardiac death in the young as 1-2 cases per 100,000 person-years.[4]
- One review found an annual incidence of 1.3 cases per 100,000 in those aged 1-35 years of age. 72% involved boys or young men. Those aged 31-35 years had the highest incidence of sudden cardiac death (3.2 cases per 100,000 persons per year), and those aged 16-20 years had the highest incidence of unexplained sudden cardiac death (0.8 cases per 100,000 persons per year).
- The most common explained causes of sudden cardiac death were coronary artery disease (24% of cases) and inherited cardiomyopathies (16% of cases). Unexplained sudden cardiac death (40% of cases) was the predominant finding among all age groups, except for those aged 31-35 years, for whom coronary artery disease was the most common finding.
- Younger age and death at night were independently associated with unexplained sudden cardiac death compared with explained sudden cardiac death.

- A clinically relevant cardiac gene mutation was identified in 31 of 113 cases (27%) of unexplained sudden cardiac death in which genetic testing was performed. During follow-up, a clinical diagnosis of an inherited cardiovascular disease was identified in 13% of the families in which an unexplained sudden cardiac death occurred.
- In general, the risk of SCD in young people approximately doubles during physical activity and is two to three times higher in athletes compared to non-athletes. However, the incidence of SCD in young athletes is in fact very low.[5] [6]
- One US review of SCD among young athletes found an incidence of 1:50,000 in college athletes and between 1:50,000 and 1:80,000 for high school athletes, with certain subgroups at particularly high risk, including men, basketball players, and African Americans. Although initial reports suggested that the most common cause of SCD was hypertrophic cardiomyopathy (HCM), more detailed investigations of athletes, non-athletes, and military indicate that the most common finding on autopsy in young individuals with SCD is a structurally normal heart (autopsy-negative sudden unexplained death).
- Premature atherosclerotic disease is also an important cause in young adults, as well as congenital cardiac abnormalities.
- There is an increased risk of SCD associated with cocaine abuse.[7]

Aetiology[8] [9]

The Cardiac Risk in the Young (CRY) Centre for Cardiac Pathology is based at St George's University of London. A large cohort study of 756 adolescents concluded that Sudden cardiac death may occur during adolescence, even in apparently healthy individuals and athletes. A structurally normal heart at autopsy and myocardial diseases are the prevalent findings in this population. Coronary anomalies, arrhythmogenic cardiomyopathy, and commotio cordis are common causes of death in young athletes. The strong association of cardiomyopathies and coronary anomalies with exercise-induced SCD reinforces the need for early diagnosis.[10]

In the USA, the National Registry of Sudden Death in Athletes was established in the 1980s and reported on 1,866 sudden deaths in individuals under 40 years of age during a 27-year observational period.[11]

Their data showed that 36% of all sudden deaths in this registry were attributed to confirmed cardiovascular causes, of which the most frequent are hypertrophic obstructive cardiomyopathy (36%), congenital anomalies of the coronary arteries (17%), myocarditis (6%), arrhythmogenic right ventricular cardiomyopathy (4%) and channelopathies (3.6%).[12]

In 4% of sudden deaths in the 16-64 age group, post-mortem examination failed to identify a cause; these cases are diagnosed as having sudden arrhythmic death syndrome (SADS).[13] [14]

- Hypertrophic obstructive cardiomyopathy (HOCM).
- Dilated cardiomyopathy.
- Arrhythmogenic right ventricular cardiomyopathy (ARVC).
- Cardiac ion channelopathies - eg, congenital long QT syndrome (LQTS), Brugada's syndrome, short QT syndrome.
- Catecholaminergic polymorphic ventricular tachycardia (CPVT).
- Valvular heart disease (with or without infective endocarditis) - eg, aortic stenosis, mitral valve prolapse.
- Cyanotic heart disease - eg, Fallot's tetralogy, transposition.
- Acyanotic heart disease - eg, ventricular septal defect, patent ductus arteriosus.
- Cardiac arrhythmias - eg, Wolff-Parkinson-White syndrome.
- Coronary heart disease: acute myocardial infarction, congenital anomaly of coronary arteries, coronary artery embolism, coronary arteritis.
- Myocarditis.
- Myotonic dystrophy.
- Kawasaki disease.
- Commotio cordis (traumatic blow to the chest wall).

Major causes of sudden cardiopulmonary death include pulmonary embolism, aortic dissection and ruptured aortic aneurysm.

Differential diagnosis

Other causes of sudden death in young people include:[15]

- Serious infection - eg, meningitis, encephalitis.
- Epilepsy.
- Asthma.
- Pulmonary embolism.
- Intracranial haemorrhage.

Investigations[16]

- The background history of the victim of sudden death should be elicited, including preceding symptoms, previous medical history and circumstances of death.
- The family history must be established, including any histories of unexplained syncope, sudden death or muscle weakness.
- The results of a post-mortem examination (including toxicology screen and blood analysis - eg, electrolytes) may provide a clear indication of the cause of death and any specific investigations that should be offered to first-degree relatives.
- Post-mortem CT angiography combined with image-guided biopsy have a potential role in the detection of the cause of death after acute chest pain.[17]

Finding a sudden death where there is no cardiac anomaly at post-mortem could point towards SADS. Criteria to explore this potential cause of death include negative general autopsy findings, both macroscopic and histological, along with negative toxicology and a morphologically normal heart. In such cases, it is strongly recommended that the heart is retained in its entirety and either sent for expert cardiac opinion or examined histologically in detail locally in an attempt to elucidate any underlying cause.[18]

Investigation of sudden cardiac arrest survivors

- Survivors of cardiac arrest require a comprehensive clinical assessment including a detailed presenting history with witness statements, family and drug histories.
- Baseline electrolyte and metabolic testing to look for reversible causes of cardiac channel instability, along with markers of cardiac injury. Further investigations should be undertaken if findings are suggestive of cardiac involvement of systemic disease, such as amyloid, sarcoid, autoimmune disease or infection.
- Structural and electric testing should initially include coronary angiography, echocardiography, and resting ECG. Further imaging with cardiac magnetic resonance imaging (MRI) and drug provocation should also be included.
- Coronary imaging, usually with coronary angiography, is required to exclude coronary heart disease, including to rule out congenital coronary anomalies in younger people.
- Subsequent testing includes treadmill testing and a signal-averaged ECG. Signal-averaged ECG testing is primarily used to look for evidence of late potentials, which is helpful in the screening of ischaemic cardiomyopathy, subclinical arrhythmogenic right ventricular cardiomyopathy and Brugada's syndrome.
- Drug provocation to unmask a primary electric cause of cardiac arrest plays a key role when the diagnosis remains unclear. Provocation testing protocols, including sympathomimetic or sodium-channel blocking drug infusions, are primarily used to unmask phenotypes of long-QT syndromes, Brugada's syndrome and CPVT.
- Advanced imaging (such as gated cardiac MRI or CT scanning and nuclear imaging - eg, thallium or technetium 99^{m}Tc scintigraphy) should be considered unless a clear diagnosis has been obtained.
- When the diagnosis still remains unclear, further testing is exploratory and unlikely to provide a definitive diagnosis. Additional tests may include electrophysiology studies with voltage mapping and cardiac biopsy.

- Genetic testing is indicated when an inherited phenotype (arrhythmogenic right ventricular cardiomyopathy, Brugada, CPVT or long-QT syndromes) is detected, both for diagnosis and to aid family screening.

Investigation of first-degree relatives

- The management of families depends on the outcome of thorough assessment of the index cardiac arrest survivor.
- The focus is on exclusion of known phenotypes/genotypes or, alternatively, the blind workup of a relative of an undiagnosed cardiac arrest.
- In practice, investigations are confined to an ECG and echocardiogram in most cases
- In situations in which the index case diagnosis is unclear, a tiered approach to screening is usually offered, including ECG, echocardiogram, exercise testing, signal-averaged ECGs and Holter monitoring.
- Cardiac MRI, electrophysiology studies and cardiac biopsy are reserved for when the diagnosis is unclear.

Sudden cardiac death prevention

- If a diagnosis has been made then advice on testing of other relatives is appropriate.
- Pre-participation cardiovascular screening of young competitive athletes by 12-lead ECG (in addition to history and physical examination) has been recommended both in Europe and the USA. [19]
- The detection of cardiac abnormalities, such as hypertrophic cardiomyopathy, dilated cardiomyopathy, or arrhythmogenic right ventricular dysplasia offers the prospect of treatment that will improve symptoms and greatly reduce the risk of sudden death.
- Patients with a substantial risk of sudden death associated with cardiac arrhythmias usually need an implantable cardioverter defibrillator (ICD). [20]

- The National Institute for Health and Care Excellence (NICE) recommends there is adequate evidence for the benefit of the insertion of a subcutaneous ICD for the prevention of sudden cardiac death for patients with arrhythmias and those at risk of sudden cardiac death. [21]

Further reading

- Tsuda T, Fitzgerald KK, Temple J; Sudden cardiac death in children and young adults without structural heart disease: a comprehensive review. Rev Cardiovasc Med. 2020 Jun 30;21(2):205-216. doi: 10.31083/j.rcm.2020.02.55.
- Cardiac Risk in the Young (CRY) Centre for Cardiac Pathology.

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