Sudden cardiac death in the young

The annual incidence of SCD in the general population is estimated to be less than 1 in 1000 but it is devastating for those left behind.

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Sudden cardiac death (SCD) is defined as ‘death due to natural causes within an hour of the onset of symptoms, in the absence of any other cause, and assumed or proven to have a cardiac cause.’ When a young person dies it has catastrophic effects on the family. Post mortem results of SCD in the young have shown a number of underlying conditions, such as cardiomyopathy, myocarditis and coronary heart disease. There are also many unexplained deaths (approximately 20-30%) in people under 35 years of age where there is no structural abnormality found at post mortem and these are classed as sudden arrhythmic death syndrome (SADS).

The annual incidence of SCD in the general population is estimated to be less than 1 in 1000. Over 5,000 people suffer sudden cardiac death in Ireland each year, of which 60 to 80 are under the age of 35 years. Review of Irish post mortem data suggests that SCD in a population of 14-35 years occurs in 3 to 4 per 100,000, with males being 7 times more likely to be affected. It also suggests that in up to 50% of cases of SCD in the young, the cause may be inherited.

High risk sub-groups
Since prospective cardiac assessment of the general population is not currently feasible, the initial focus is to target high-risk subgroups

- History of sudden death in close family member with suspected cardiac cause or unknown cause
- Family history of inherited cardiac disease
- Signs and symptoms that may indicate cardiac cause

Signs and symptoms

- Chest pain with activity that limits completion of activity
- SOB on exertion that happens repeatedly on less than maximal exertion
- Dizziness syncope – especially on exercise
- Exercise intolerance
- Sometimes no symptoms – could be found during routine medical examination
- Unexplained/uncontrolled seizure-like episodes
Hypertrophic cardiomyopathy (HCM) is a genetic disease associated with a risk of ventricular tachyarrhythmias and sudden death, especially in young patients.

Conditions that can be detected on cardiac testing include:
- Cardiomyopathy: dilated cardiomyopathy (DCM), hypertrophic cardiomyopathy (HCM), arrhythmogenic right ventricular cardiomyopathy (ARVC).
- Long QT syndrome (LQT)
- Brugada syndrome
- Catacholaminergic polymorphic ventricular tachycardia (CPVT)
- Wolf Parkinson White syndrome (WPW)
- Connective tissue disorders (e.g. Marfans).

Hypertrophic cardiomyopathy
Hypertrophic cardiomyopathy (HCM) is a genetic disease associated with a risk of ventricular tachyarrhythmias and sudden death, especially in young patients. The heart muscle becomes thick and fibres are arranged haphazardly increasing the risk of fatal arrhythmias. The thickened muscle interferes with the heart’s normal contraction and relaxing, which results in reduced cardiac output. 25-30% of patients have the ‘obstructive’ form with relative obstruction to blood flow from the left ventricle to the aorta.

LQT
Long QT syndrome is an association of cardiac rhythm disorders and QT prolongation or abnormal T waves on ECG. Abnormal corrected QT measurements are defined as being >440ms in men and >460ms in women for a diagnosis to be made. Some medications may increase the risk of arrhythmia in LQT syndrome. A patient’s resting ECG is not always abnormal in LQT patients.

Brugada syndrome
Brugada syndrome was first reported in 1992 by Josep and Pedro Brugada. It is an arrhythmogenic disease that is characterised by ST elevation in the right precordial leads (V1 – V3) with incomplete or complete right bundle branch block (RBBB) pattern on the ECG. It is associated with a high incidence of syncope and sudden death in patients with a structurally normal heart. The patients resting ECG is often significantly abnormal in patients at high risk of arrhythmia. Gene carriers of Brugada syndrome may be unmasked during Ajmaline provocation testing.

CPVT
Catecholaminergic polymorphic ventricular tachycardia is an arrhythmogenic disorder of the heart characterized by a reproducible form of polymorphic ventricular tachycardia induced by physical activity, stress, or catecholamine infusion, which can deteriorate into ventricular fibrillation. Resting ECG is normal or near-normal in patients with CPVT. Diagnosis is made on exercise testing and epinephrine provocation testing.

Wolff-Parkinson-White syndrome
Wolff-Parkinson-White (WPW) syndrome is a heart condition in which there is an extra electrical pathway (circuit) in the heart which can lead to tachycardia. Resting ECG is usually abnormal in patients with WPW.

Figure 1 – Family pedigree with 4 generations
If a person is at risk they can be treated with either medication or an internal cardiac defibrillator.

- Evaluation of young persons who have symptoms
- Support for families during their tests and counselling service provided if required
- Referral for genetic testing if required.

**Conclusion**
SCD in the young occurs in Ireland approximately once a week and for the families it is devastating. Post mortems will reveal any structural abnormalities within the heart but electrical conditions cannot be detected after death and are known as ‘SADS’. The nurse can play an important role in educating and counselling individuals and family members during this difficult period.

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**References**
1. Task Force for Sudden Cardiac Death, 2004