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Prognosis in patients with hypertrophic cardiomyopathy (HCM) is generally good but a small percentage of patients die suddenly because of abnormally fast heart rhythms called ventricular arrhythmias.

Once a ventricular arrhythmia becomes sustained (longer lasting), the heart is unable to pump normally, rapidly leading to death. A smaller proportion of sudden deaths are probably caused by abnormally slow heart rhythms.

The annual risk of sudden cardiac death (SCD) in contemporary medical literature is less than one per cent per year and fortunately only a small proportion of HCM patients die suddenly. Despite intense research in this field, SCD remains a complex and incompletely understood clinical problem. This article looks at assessing the risk.

Prevention of SCD

Avoiding exercise

Intense exertion may bring on ventricular arrhythmias and SCD. International guidelines recommend that patients with HCM avoid competitive sports and strenuous physical effort. Even though the effectiveness of this approach is unclear, on balance the medical community feels that this is a reasonable recommendation.

Drugs

Drugs which help control heart rhythm (such as amiodarone and sotalol) do not provide adequate protection from SCD and should not be relied on for its prevention. These drugs are still used in HCM patients as they are effective in controlling other heart rhythm abnormalities such as atrial fibrillation.

Implantable cardioverter defibrillators (ICDs)

These devices, similar to pacemakers, monitor each heart beat and deliver an electric shock if a potentially life threatening ventricular arrhythmia develops. The electric shock stops the ventricular arrhythmia and restores rhythm. HCM patients treated with an ICD rarely suffer SCD. The devices are currently the most effective means of preventing SCD. Unfortunately ICDs have drawbacks:

- fitting an ICD involves an operation that carries a small risk of damaging the heart or lungs;
- the battery becomes flat so replacement is needed every five to six years;
- the ICD is a foreign body and there is a small risk of infection;

- ICD hardware malfunction may require further operations;
- ICDs can fire inappropriately after mistakenly identifying ventricular arrhythmia. Inappropriate shocks are painful and distressing and affect approximately a fifth of patients; and
- lifestyle modifications are needed and there are some driving restrictions (see DVLA regulations).

All these complications are rarely life threatening and the ICD is unequivocally the most successful treatment for preventing SCD in HCM.

Deciding on ICD treatment

When deciding whether an ICD should be fitted, patients and their medical team need to consider:

- the risk of sudden death and the implications of no treatment; and
- the benefits and the risks of treatment with an ICD.

Specialist nurses and doctors should provide information to allow patients to reach an informed decision but the nature of the problem means that uncertainties are almost always encountered.

The perception of risk also varies between patients and doctors, and the threshold for treatment may depend on non-medical circumstances such as a young family or lifestyle modification after ICD implantation. Discussion with other patients who already have an ICD may be helpful when considering treatment.

Assessing the risk of SCD

Ascertaining the magnitude of SCD risk is essential so that patients at high enough risk, who would benefit most from an ICD, can be identified. All HCM patients should have a comprehensive assessment.

In the small number of people who survive a cardiac arrest there is a high risk of recurrence. Doctors agree ICDs are beneficial in these patients.

The vast majority of HCM patients do not have a prior episode. The risk assessment involves a clinical review, an ultrasound scan of the heart (echocardiogram), an exercise test on a bicycle or treadmill and a heart rhythm monitor over 24 to 48hrs.

The risk of SCD should then be reassessed every one to two years, or after changes in symptoms such as fainting.



Guidelines in the USA (published in 2011), and the 2003 European guidelines, recommend assessment of five risk factors associated with SCD.

- Unexplained collapse with loss of consciousness or an episode of fainting with no obvious medical explanation. Fainting after having blood taken or because of a slow heart rate is not considered a marker of high risk. A sudden faint with no warning is more suspicious.
- A history of SCD in a first degree relative (parent, sibling or child). A family tree will be drawn up and relatives who died suddenly at a young age identified. This is sometimes limited by lack of information.
- Short episodes of ventricular arrhythmia during heart rhythm monitoring. Most patients are not aware of them but they are a marker of risk particularly in a young patient or during exercise.
- Extreme thickening of the main pumping chamber of the heart seen in an echocardiogram. Sometimes an magnetic resonance scan (MRI) of the heart may be needed.
- Failure of blood pressure to rise during exercise on a treadmill or bicycle test. Normally blood pressure increases with exercise and in proportion to the intensity of exercise. In some HCM patients this does not happen and this has been linked to SCD. Medication that lowers blood pressure and heart rate should be discontinued prior to the test. It is of limited value in the very young and in older patients.

With a few exceptions, none of these five risk factors are strong enough to predict SCD on their own. To overcome this problem, guidelines recommend the use of ICDs in patients with multiple risk factors (such as family history of SCD as well as unexplained fainting) or in some patients with a single risk factor.

This approach has been in clinical use since 2003 and is partially successful. Research which critically appraised the robustness of this approach demonstrated several limitations which lead to the development of a new method for risk stratification.

HCM Risk-SCD

To overcome the limitations above, a collaboration of European investigators developed a new method of assessing the risk of SCD in 2013. This is called HCM Risk-SCD and since its publication has been endorsed by the 2014 European Society of Cardiology guidelines as the preferred method for risk stratification.

As well as the risk factors listed above, age, the size of the left atrium of the heart and obstruction to the blood flow out of the heart are also considered to estimate the risk of SCD within five years.

Unlike the existing guidelines which classify patients into high and low risk, this model provides a numerical estimate of risk as a percentage. The new method works similarly to car insurance prices where companies use data from previous customers to calculate the risk associated with a particular applicant. In the same way that a young driver with a powerful car pays a higher insurance than a middle aged driver with a more modest car, a young HCM patient with unexplained fainting has a higher risk of SCD than an older patient without fainting. The five year estimate of SCD risk can be used by patients and doctors to consider whether the risks of having an ICD outweigh the benefits.

HCM Risk-SCD can be used in most patients with HCM. However, in patients with Anderson-Fabry disease or other rarer causes of HCM, this method should not be used. It is also not clear how to use it in patients who have had treatment for obstruction (an alcohol septal ablation or myectomy*) after their initial assessment. Patients with a history of cardiac arrest should receive an ICD irrespective of their HCM Risk-SCD.

Since 2014, this method of risk stratification has been tested in more than 7000 HCM patients around the world and the results of several studies suggest that HCM Risk-SCD provides accurate predictions in most patients.

Conclusions and future directions

Predicting the risk of SCD, irrespective of method or heart condition, is challenging. HCM Risk-SCD represents an improvement in the management of HCM patients, but it is not perfect and currently it is not possible to predict all cases of SCD.

HCM Risk-SCD will also be updated and additional risk factors, such as genetic information, will be included to improve its predictive accuracy. Improvements in ICD technology will also help reduce ICD complications and help tip the balance in favour of treatment in some patients.

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