key points

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Inherited heart conditions are the most common cause of sudden cardiac death in those under the age of 35 and the leading cause of non-traumatic death in young athletes. Hypertrophic cardiomyopathy (HCM) is the most common inherited heart disease affecting 1 in 500 of the population. Some patients may exhibit severe left ventricular hypertrophy, others may show nothing more than an abnormal ECG. Left ventricular hypertrophy most commonly manifests in the second decade of life. Sudden death is rare and usually affects patients in the first three decades whereas older patients present with heart failure, atrial fibrillation and stroke.

Arrhythmogenic right ventricular cardiomyopathy is a rare, autosomal dominant heart muscle disorder which affects between 1 in 1,000 and 1 in 5,000 of the population. The current hypothesis is that abnormal desmosomal proteins lead to myocyte detachment during exertion and promote arrhythmias. The diagnosis usually requires a plethora of electrical and imaging tests.

Dilated cardiomyopathy (DCM) is characterised by a dilated left ventricle with impaired function that cannot be explained by ischaemic heart disease, hypertension or valvular heart disease. At least 25% of cases of DCM are familial. DCM may be associated with multisystem conditions such as muscular dystrophy. Chemotherapy and certain other drugs, alcohol abuse and myocarditis may also lead to a dilated and poorly contracting left ventricle.

In many cases the first manifestation of an inherited cardiomyopathy can be a sudden cardiac arrest. Other presentations include chest pain or breathlessness during exertion, palpitations and syncope. In many of the cardiomyopathies, the diagnosis can be made with a standard ECG and echocardiogram. However if the diagnosis is not certain or the cardiologist wishes to look at the heart structure in greater detail, a cardiac MRI may be performed.

For symptomatic patients with HCM a beta-blocker is the first-line treatment and a calcium channel blocker such as verapamil can be used if a beta-blocker is contraindicated. If patients are felt to be high risk, an implantable cardioverter-defibrillator is recommended. Dilated cardiomyopathy is treated with conventional heart failure treatment usually with a beta-blocker and an ACE inhibitor in the first instance.