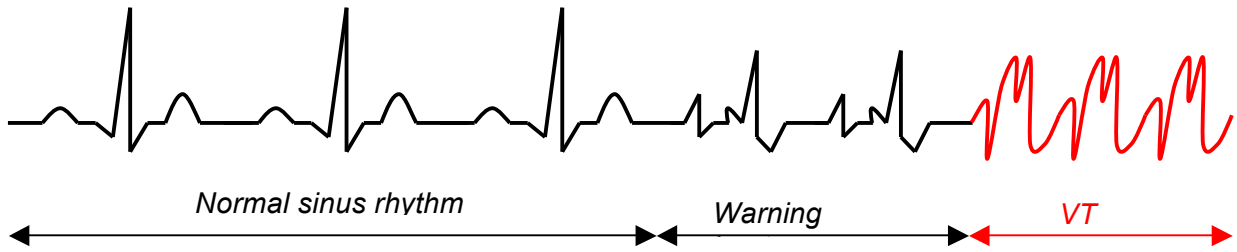
Catecholaminergic Polymorphic Ventricular Tachycardia (CPVT)

Catecholaminergic Polymorphic Ventricular Tachycardia (CPVT) is a rare condition that affects the heart of otherwise fit and healthy people. It causes the heart to beat abnormally fast (ventricular tachycardia), usually at times of exercise (particularly swimming) or high emotion. It can result in dizziness, sudden loss of consciousness or even death. It most commonly occurs in children and young adults, typically in the first or second decade of life. At least a third of those people with CPVT have affected relatives so the condition is said to be 'genetic', 'familial' or 'inherited'.

When the heart needs to work harder (e.g. during exercise or when a person is emotional), the body releases adrenaline and noradrenaline (known as catecholamines); these cause the heart to beat faster and increase blood pressure. This response increases the amount of blood and oxygen getting to areas that need it. People with CPVT have an abnormal response to adrenaline, which causes the heartbeat to become fast and irregular (ventricular tachycardia [VT]). If this rhythm lasts longer than a few seconds, the brain does not receive enough blood and this can lead to fainting, collapse and sudden death.

It is a difficult condition to diagnose because if tests are done when a person is not stressed it give normal results. These tests include electrocardiogram (ECG) and echocardiogram (ultrasound of the heart). The diagnosis is usually made by detecting extra beats or bursts of fast rhythm arising from the bottom part of the heart (ventricles) during an exercise test or on a 24-hour ECG (Holter monitor).



Exercise ECG of CPVT

How it is inherited?
CPVT is usually inherited as an autosomal dominant condition but can sometimes be inherited as an autosomal recessive condition. This means both copies of the gene must have an alteration for the condition to occur (generally meaning both parents must be carriers). See section on Genetic Inheritance for more information.

Genetic testing
Genetic testing is now commercially available and involves screening two genes that can cause the condition. The gene alteration will be identified in approximately 70% of families.

Treatment

Treatment depends on the severity of symptoms. People with CPVT must avoid swimming and competitive sport. Beta-blocker medications are taken regularly and a small key-hole operation to cut the nerves to the heart can be helpful (left cervical sympathectomy). Those who are felt to be at high risk of cardiac arrest may benefit from an ICD (implantable defibrillator).

It is recommended that people with CPVT see their cardiologist at least once a year.

For more information

To find out more about genetic heart conditions and the latest research news, please visit the Australian Genetic Heart Disease Registry at www.heartregistry.org.au or the Cardiac Inherited Diseases Group (New Zealand) at www.cidg.org