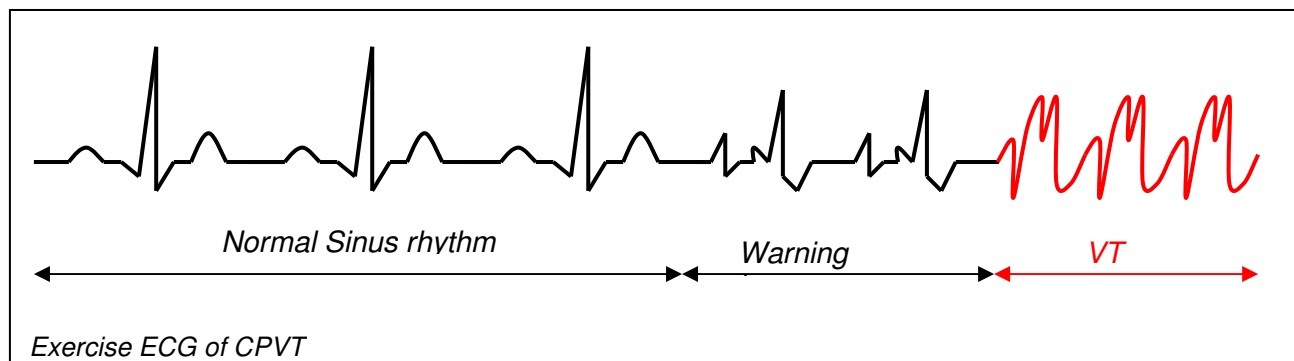


Catecholaminergic Polymorphic Ventricular Tachycardia (CPVT)

Catecholaminergic polymorphic VT is a rare condition that affects the heart of otherwise fit and healthy people. It causes the heart to beat abnormally quickly (ventricular tachycardia), usually at times of exercise (particularly swimming) or times of 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. In at least a third of cases, it is familial (inherited), being passed down through the generations.

When the heart is under stress, eg. during exercise or emotional stimulation, the body releases adrenaline and noradrenaline (known as catecholamines), which cause the heart to beat faster and increase blood pressure. This response allows blood and oxygen to get to the required areas faster. People with CPVT have an abnormal response to adrenaline, such that it causes the heartbeat to become fast and irregular (ventricular tachycardia (VT)). If this irregular rhythm lasts for longer than a few seconds then not enough blood reaches the brain and can lead to fainting, collapse and sudden death.

It is a difficult condition to diagnose, because all of the tests taken at a time of rest are normal- including the electrocardiogram (ECG), and the echocardiogram (ultrasound of the heart). The diagnosis is usually made by detecting extra beats, or runs of fast rhythm arising from the bottom part of the heart (ventricles) during an exercise test or on a 24 hour ECG (holter monitor).



How it is inherited

CPVT is generally inherited as an autosomal dominant disease (see section on Genetic Inheritance for more information). There are some families that pass on this condition as an autosomal recessive trait, which means both copies of the gene must have an alteration for the disease to occur (generally meaning both parents must be carriers).

Genetic testing

Genetic testing is available but not straightforward. Three genes have been identified to cause catecholaminergic polymorphic VT in about 70% of cases.

Treatment

Treatment is adjusted according to severity. Those affected 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 (intracardiac defibrillator).

All people with CPVT should see their Cardiologist as a minimum once a year.

A more detailed review is available at www.cidg.org