FACTSHEET

CPVT - Catecholaminergic Polymorphic Ventricular Tachycardia

What is CPVT?

Catecholaminergic Polymorphic Ventricular Tachycardia (CPVT) is a rare heart rhythm disorder, which can cause periods of abnormally fast heart rhythm called arrhythmias. The condition affects around 1 in every 10 000 people. These arrhythmias are more likely to occur when the stress hormones, adrenaline and noradrenaline (also known as 'catecholamines'), are released into the body. Catecholamines normally increase heart rate and blood pressure in response to physical or emotional stress. This helps the muscles of the body receive the extra oxygen they need. People with CPVT do not respond to catecholamines in the usual way. Their heartbeat can become very fast and irregular, causing an arrhythmia called ventricular tachycardia (VT).

What are the symptoms?

Symptoms of CPVT are most likely to occur at times of strenuous physical activity. If the fast heart rhythm occurs, not enough blood is pumped from the heart. This can lead to a fall in oxygen supply to the body and brain and a loss of consciousness (fainting episode). If this episode is not recognised and treated immediately it may be life-threatening. Symptoms of CPVT usually occur for the first time during late childhood and adolescence.

How is CPVT diagnosed?

People with CPVT do not have any structural abnormalities in their heart. A routine ECG (a test of the heart's electrical activity, performed when a person is at rest) is also usually normal for people with CPVT. Therefore, CPVT can be difficult to diagnose, and sometimes the episodes of collapse can be mistaken for other conditions. Additional tests are often needed to diagnose CPVT. These tests may include:

- An exercise stress test: ECG monitoring is performed during exercise (usually while running on a treadmill).
- An adrenaline challenge: This involves admission to hospital for cardiac monitoring whilst intravenous adrenaline is administered in a safe environment.

Detailed family history or genetic testing may also help with diagnosis of CPVT.

What is the cause?

Some children are born with CPVT because they have inherited a gene change associated with CPVT from one or both of their parents. Parents may have been unaware they had the condition because they may not have had any symptoms.

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CPVT can also result from a gene change that has happened after conception. If this is the case, the gene change can be passed on to the affected child's own children in the future, but their siblings are usually unaffected.

There are currently four genes known to be associated with CPVT. Changes in the *RYR2* (Ryanodine receptor 2 gene) gene causes CPVT in approximately 50% of those with CPVT. CPVT can be inherited in different ways in families. Sometimes the way CPVT is inherited can be determined by the family history or genetic testing. Genetic testing can be used to confirm a clinical diagnosis of CPVT and if the gene change is known potentially allow testing of other family members. If no changes in the CPVT genes are found by genetic testing this does not rule out the diagnosis of CPVT, as not all of the causes of CPVT are known.

For more information on the genetic aspects of CPVT please refer to the following information sheets:

- Australian Genetic Heart Disease Registry - CPVT: http://www.heartregistry.org.au/wp-content/uploads/2012/12/AGHDR_CPVTInfoSheet_2012.pdf
- Centre for Genetics Education – Primary Arrhythmogenic Disorders: http://www.genetics.edu.au/Publications-and-Resources/Genetics-Fact-Sheets/FS58PRIMARYARRYTHMOGENICDISORDERS.pdf

How is it treated?

The treatment for CPVT is aimed at reducing the risk of the fast rhythm. People with CPVT can be affected to varying degrees and so treatment is tailored to the individual.

Lifestyle modifications:
Your Cardiologist will provide information specific to your child regarding what lifestyle changes or modifications your child may need to adhere to. For most, this will mean avoiding strenuous activity, especially competitive sports and swimming.

Medications:
First line treatment is usually medication. The medications used are beta-blockers. Beta blocker medications block the effect of adrenaline on the heart, preventing it from beating faster. The most commonly used in CPVT are propranolol, atenolol and nadolol.

Implantable cardioverter defibrillator:
If beta-blockers are not effective in managing your child's heart rhythm or your child has already had a significant cardiac event, your child may need to have an operation to implant an implantable cardioverter defibrillator (ICD). This device requires implantation under the skin, underneath either the left collar bone or the ribcage. An ICD is similar to a pacemaker but is also able to deliver an electrical impulse if it senses that the heart is beating in a potentially dangerous rhythm. This can help return the heart to normal rhythm and get it pumping again.

Automatic External Defibrillator:
An automated external defibrillator (AED) is a portable device that is used to deliver an electric shock in order to make the heart rhythm normal again. In this way it is similar to an ICD, however the AED is an external machine that has adhesive pads that need to be applied to the chest in order for the shock to be delivered. The current devices are all pre-programmed to analyse the rhythm once the pads are applied and will audibly instruct the user what actions are required. It is best to speak to your Cardiologist about whether this device is recommended for your child.

How we can help?

The Heart Centre for Children is dedicated to supporting children diagnosed with CPVT and their families. The Inherited Arrhythmia Clinic (IAC) at the Heart Centre for Children, The Children's Hospital at Westmead, offers a comprehensive range of services for families with this diagnosis and other arrhythmias. Please refer to the IAC factsheet available at: www.heartcentreforchildren.com.au.

Contacts:

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