Arrhythmia Alliance
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Catecholaminergic Polymorphic Ventricular Tachycardia (CPVT)

What is CPVT?

Catecholaminergic Polymorphic Ventricular Tachycardia (CPVT) is an inherited arrhythmia (heart rhythm disorder). CPVT is sometimes known as a channelopathy. This is because CPVT is caused by an abnormality of ion channels which allows calcium to accumulate in heart muscle cells. This can lead to abnormal heartbeats - usually during exercise or when epinephrineis released in stressful situations. These extra beats can arise from the top chambers of the heart (atrial ectopic beats) or, more frequently, from the bottom pumping chambers of the heart (ventricular ectopic beats). If there are runs of fast beats from the atria or ventricles these are known as atrial or ventricular tachycardia. In CPVT the heart usually has a normal structure. It is thought that about 1 in 10,000 people have CPVT, although the exact numbers are unknown.

Symptoms

Episodes of ventricular tachycardia (VT) can cause lightheadedness, dizziness and loss of consciousness (syncope). Symptoms usually start in childhood but can appear in young adults for the first time.

Most syncopal episodes in childhood are benign, however those who suffer syncope during exercise or in response to an epinephrine stimulus should be further investigated. Sometimes these episodes can be mistaken for a seizure/epilepsy as they can look very similar and as a consequence many children are treated with antiepileptic drugs.

In a child not responding to antiepileptic drugs a diagnosis of CPVT should be considered. Unfortunately, sometimes the first presentation can be sudden cardiac arrest - an episode of VT cannot be sustained for a long period of time and may result in the heart completely stopping (cardiac arrest). Atrial tachycardia usually causes a sensation of inappropriate racing of the heart (palpitations). This is not usually dangerous but can be unpleasant.

Diagnosis

In patients presenting with sudden cardiac arrest in the absence of structural cardiac disease, CPVT should be considered in the differential diagnosis. Clinical diagnosis is made based on family history, exercise or emotional stress-induced symptoms and, significantly, response to exercise or catecholamine (epinephrine) infusion, however not all episodes are triggered by epinephrine.

A cardiac ultrasound (echocardiogram) and resting EKG are usually normal.

Genetics

The most common cause of CPVT results from mutations in the cardiac ryanodine receptor gene (RYR2). Less common causes are in other genes such as Calsequestrin, Calmodulin and Triadin. These genes code for proteins that handle calcium which helps maintain a regular heartbeat.

Familial inheritance has been seen in about a third of cases of CPVT. RYR2 causes autosomal dominant inheritance meaning if you inherit the abnormal gene from only one parent, you can get the disease.

First-degree relatives should be evaluated with EKG, Holter monitoring and exercise stress testing. Genetic testing can sometimes be helpful. Identification of a genetic abnormality that is the definite cause of the condition (a

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"pathogenic" mutation) can allow other family members to be tested.

Analysis might identify silent carriers of CPVT-related mutations, and it may be recommended that even symptom-free carriers are treated with medication such as beta-blockers. Sometimes, a genetic change is discovered and it is unclear if this is the definite cause. This is known as a "variant of uncertain significance " or VUS. A VUS cannot be used for family screening.

Treatment

Once diagnosed, treatment is usually with a beta-blocker. Beta-blockers decrease the activity of the heart by blocking the action of hormones such as epinephrine, which would normally increase in times of exercise or emotional stress. Subsequently, the number of episodes of VT is reduced. A high dose is often required.

Flecainide works on the sodium channel and may inhibit cardiac ryanodine receptor-mediated calcium release. Flecainide appears to work very well in many people and is rapidly becoming a preferred drug in many people with CPVT.

Missing even a single dose of beta-blocker can be potentially dangerous. Internal cardiac defibrillators (ICDs) are sometimes fitted in addition to medication to 'shock' the heart back into normal rhythm if VT occurs. Survivors of cardiac arrest or high-risk patients with a strong family history of sudden death are more likely to be offered an internal cardiovertor defibrillator. ICD treatment without use of beta-blockers is not advised as a shock from the defibrillator can lead to an epinephrine surge and multiple runs of ventricular tachycardia known as an "electrical storm".

A left cervical sympathectomy may be offered to some patients, e.g., those in whom beta-blockers are contraindicated, when an ICD cannot be implanted, or where there is recurrent VT in patients with an ICD despite maximal medical treatment. A cervical sympathectomy is an operation carried out through a small incision under the arm. This blocks a group of nerves that produce and deliver epinephrine to the heart. These nerves are not essential to normal heart function but sympathectomy can be very helpful in preventing serious arrhythmias.

Additionally, some sports may be restricted, e.g., competitive swimming, as participation can result in an episode of tachycardia.

Arrhythmia Alliance offers helpful resources on ICDs and left cardiac sympathetic denervation (LCSD) for those exploring treatment options.

To view our patient resources, scan the QR code below:

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