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Catecholaminergic Polymorphic Ventricular Tachycardia (CPVT)

Inherited Heart Disease Clinic

How does the heart work?

The heart is a hollow organ made up of muscle. It has 4 chambers: 2 at the top (**atria**) and 2 at the bottom (**ventricles**).

Blood flows from the body into the top chambers. The blood is pumped to the bottom chambers, and is then pumped back out to the body.

In a normal heartbeat, an electrical signal starts in the upper chamber and travels through the heart muscle to the bottom chambers. This electrical activity is called the heart rhythm.

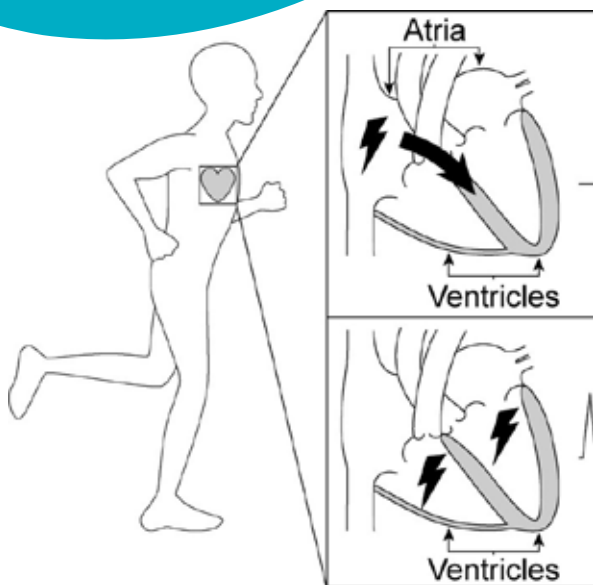
Heart cells have channels that work like doors. These channels open and close to let electrical signals flow through the heart cells.

When you exercise or have a strong emotional reaction (like fear), your body releases adrenaline. Adrenaline affects the heart cell channels by causing more electrical signals in the top chambers. This makes your heart beat faster.

What is Catecholaminergic Polymorphic Ventricular Tachycardia?

CPVT affects the way heart cell channels respond to adrenaline. It causes extra heartbeats to start in the bottom chambers of your heart. This can make your heart beat too fast.

Your heart might beat so fast that it cannot pump blood out to your body. These abnormal heart rhythms can make you collapse (faint). **If the rhythm does not stop, it can cause sudden death.**



What causes CPVT?

CPVT can be caused by changes in your genes that create the fault in the heart cell channels. CPVT can be hereditary (passed from parents to their children).

Sometimes the cause of CPVT is not known.

It is important to tell your health care provider if you had a relative who died suddenly at a young age. This could have been caused by CPVT.

What are the symptoms of CPVT?

Symptoms may include:

- › Fainting
- › Blackouts (passing out or not being able to remember a certain amount of time)
- › Seizures (brief, abnormal, excessive [too much] discharge of electrical energy in the brain)
- › Palpitations (feeling like your heart is jumping, racing, or fluttering)

Normal heart rhythm



CPVT



How is CPVT diagnosed?


Health care providers use different tests to diagnose CPVT. You may have some of these tests:

Electrocardiogram (ECG/EKG): A recording of your heart rhythm for 10 to 20 seconds.

Exercise stress test: A recording of your heart rhythm and blood pressure while you run on a treadmill.

Holter monitor: A recording of your heart rhythm for 24 hours (1 day).

Your cardiologist (heart doctor) may want you to have other tests when they are diagnosing you. They will talk with you about this, if needed.



Genetic testing and family screening

If your cardiologist thinks changes in your genes caused your CPVT, they may ask if you would like to talk with a genetic counsellor about genetic testing.

Your cardiologist may also want to ask other members of your family to test for CPVT.

They may ask you to help by giving letters to your family members.

How is CPVT treated?

There is no cure for CPVT, but there are treatments that can control fast heart rhythms.

Treatments may include:

- › a pacemaker called an **internal cardioverter defibrillator** (ICD). The ICD identifies fast heart rhythms and slows your heart rhythm down to a safer speed.
- › medications called beta-blockers.
- › other medications to control your heart rhythm.

Sometimes, heart surgery may be needed to lower the amount of adrenaline going to the heart.

If you have CPVT:

- Your health care provider will talk with you about what activities are safe for you, and what activities and places you should avoid.
- **Do not** do strenuous (hard) exercise. This can cause you to black out.
- **Do not** take part in competitive sports.

- **Do not** swim alone. Swimming can cause you to black out and may lead to drowning.
- **Do not** use recreational drugs (substances that affect your body's nervous system), especially stimulants. This includes energy drinks.
- **Tell your family members that you have CPVT.** It can be hereditary and they may need to be checked for it.
- **Tell your loved one(s), and any support person(s) that you have CPVT.** Tell them what to do to help you if you have symptoms.

More information about CPVT:

The Canadian Sudden Arrhythmia Death Syndromes (SADS) Foundation

- › www.sads.ca
- › Includes a booklet you can download called *Cardiac Channelopathies* that has information about CPVT.

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The information in this pamphlet is to be updated every 3 years or as needed.