Arrhythmogenic Right Ventricular Cardiomyopathy (ARVC)

Inherited Heart Disease (IHD) Clinic
How does the heart work?
The heart is a hollow organ made of muscle. It has 4 chambers: 2 at the top (atria) and 2 at the bottom (ventricles). Blood flows from outside the heart into the top chambers, is pumped to the bottom chambers, and is then pumped back out to the body (see pictures).

The pumping of the heart chambers is controlled by electrical signals that pass through the heart muscle. This electrical activity is called the heart rhythm.

What is ARVC?
In arrhythmogenic right ventricular cardiomyopathy (ARVC), the heart muscle is replaced by fat and scar tissue. This can make it harder for the heart to pump blood out to the body.

ARVC usually affects the right side of the heart. The fat and scar tissue causes the right side of the heart to stretch. In very bad cases, a weak heart may cause fluid to build up outside of the heart, in the lungs (causing shortness of breath), or the ankles or belly (causing swelling).

ARVC also affects the electrical activity of the heart and can cause fast, dangerous heart rhythms.

What causes ARVC?
ARVC can be caused by a change in your DNA (genes). These changes can be hereditary (passed on from parents to children).

Sometimes the cause of ARVC is not known.
What are the symptoms of ARVC?
Symptoms may include:
- lightheadedness, fainting spells,
  palpitations (feeling like your heart is jumping, racing, or fluttering), shortness of breath, tiredness, and pain, swelling, or tightness in the chest.

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 ARVC.

How is ARVC diagnosed?
A number of tests are used to diagnose ARVC. Your health care provider may arrange for you to have some of these:

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

**Signal averaged electrocardiogram (ECG):** A more detailed recording of your heart rhythm for 5 to 10 minutes.
Holter monitor: A recording of your heart rhythm for 24 hours (1 day).

Echocardiogram (Echo): An ultrasound is used to measure the size of the heart chambers and how well the heart is pumping.

Cardiac MRI: A scan using magnets and radio waves. It can measure the size of the heart chambers, how well the heart is pumping, and show fat and scar tissue in the heart muscle.

Your cardiologist (heart doctor) may also suggest other tests. They will talk with you about this, if needed.
Genetic testing and family screening
If your cardiologist suspects that a change in your DNA could have caused your ARVC, they may ask if you would like to talk with a genetic counsellor about genetic testing. Your cardiologist may also want to invite other members of your family to be tested for ARVC. They may ask you to help by passing letters on to your family.

How is ARVC treated?
There is no cure for ARVC, but there are treatments available to control fast heart rhythms and to make you feel better.

Treatments may include:
› medications
› a special pacemaker called an internal cardioverter defibrillator (ICD) (to identify and treat fast heart rhythms)
› a heart transplant – this is rare

High levels of exercise may make ARVC worse and should be avoided. Please talk with your health care provider about what activities are safe for you.

For more info, visit:
Hearts in Rhythm Organization (HiRO)  
› www.heartsinrhythm.ca/arvc/info/  
The Canadian Sudden Arrhythmia Death Syndromes (SADS) Foundation (includes booklet on ARVC that you can download)  
› www.sads.ca
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