Commentary
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Arrhythmogenic Right Ventricular Cardiomyopathy Overview

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Arrhythmogenic right ventricular cardiomyopathy, or ARVC, is a type of cardiomyopathy that affects the ventricles (lower pumping chambers) of the heart and causes arrhythmias (abnormal heart rhythms). It affects the right ventricle, and often also affects the left ventricle. For this reason it is sometimes called arrhythmic cardiomyopathy (as the main symptoms are arrhythmias). It doesn't affect the atria (upper chambers) of the heart. In ARVC there is a defect in the proteins that join the cells of the heart muscle myocytes) together. This means that the proteins do not develop properly and cannot keep the muscle cells together. When this happens the muscle cells detach and die, the area of the heart becomes inflamed, and the lost cells are replaced with fibrous scar tissue and fat deposits. This affects the structure of the heart muscle, and it becomes thin and stretched. This causes two main problems:

The electrical pathways through the heart that coordinate the heart beat may be affected, causing arrhythmias; and the thin walls of the ventricles of the heart are unable to pump blood as effectively as normal.

Diagnosis of ARVC

There is no single diagnostic test for ARVC. The diagnosis is made using a combination of clinical, electrocardiographic and radiological features, as defined by the (horribly complicated).

Clinical Features

- ARVC causes symptoms due to ventricular ectopic beats or sustained ventricular tachycardia (with LBBB morphology) and typically presents with palpitations, syncope or cardiac arrest precipitated by exercise.
- The first presenting symptom may be sudden cardiac death.
- Over time, surviving patients also develop features of right ventricular failure, which may progress to severe biventricular failure and dilated cardiomyopathy.
- There is usually a family history of sudden cardiac death.

Electrocardiographic Features

ARVD is associated with characteristic ECG abnormalities: Epsilon wave (most specific finding, seen in 30% of patients)

- T wave inversion in V1-3 (85% of patients)
- Prolonged S-wave upstroke of 55ms in V1-3 (95% of patients)
- Localized QRS widening of 110ms in V1-3
- Paroxysmal episodes of ventricular tachycardia with LBBB morphology (e.g. right ventricular VT).

How is ARVC treated?

A variety of medicine may be used to help treat ARVC. Some of these may be needed only when the disease is more severe. They include:

- Medicines to control your heartbeat and rhythms, such as beta-blockers
- Medicines to help prevent abnormal heart rhythms (antiarrhythmics)
- Water pills (diuretics) to reduce swelling (edema)
- Medicines to reduce the workload of the heart, like ACE inhibitors
- Blood thinners (anticoagulants) to prevent blood clots

Catheter ablation is another option for certain people with ARVC. This is a procedure used to treat certain types of abnormal heart rhythms. Ablation involves threading a catheter through a vein in the groin up to the heart. There, the doctor sends heat to destroy the cells that are starting abnormal heartbeats.

Many people with ARVC need an implantable cardioverter defibrillator (ICD). An ICD is a small electronic device placed under the skin and attached to the heart. It uses electrical shocks to treat life-threatening arrhythmias. This can help prevent sudden death. Your healthcare provider will review your symptoms and test results to determine whether you need an ICD. You may need heart transplant if the damage to your heart has become severe. However, this is rare.