

Other treatments for arrhythmia

◆ **DC cardioversion** - A controlled electric shock may be effective in returning the heart to its normal rhythm.

◆ **Catheter ablation** - Patients with recurrent ventricular tachycardia, who do not respond to antiarrhythmic drugs, or have unacceptable side effects, may be offered catheter ablation. Abnormal electrical circuits within the heart muscle are identified during an electrophysiological study. This is a procedure similar to an angiogram.

Screening relatives - It is worthwhile for all immediate family members of a patient with ARVC to receive screening. This includes parents, siblings, and children.

Questions may be asked about any history of sudden death or heart problems within the family. A family tree is often constructed. Gene identification studies are currently in progress. Patients and relatives may be requested to provide a blood sample for genetic analysis. Since ARVC demonstrates incomplete penetrance, a positive genetic test does not necessarily mean that the person will develop the condition. However, regular clinical checks are recommended for gene carriers to assess for signs of the disease.

Exercise - The general advice to patients with suspected ARVC is as follows. Participation in competitive sports and endurance training is discouraged, but recreational activity may continue, provided that it does not involve extreme physical exertion. There is no specific restriction on the type of activity; we simply recommend that you do not push yourself to the limit.

Fainting during strenuous activity calls for prompt medical attention

Prolonged standing in hot conditions and very hot baths or showers may also predispose to fainting and should be avoided. Severe diarrhoea and/or vomiting interfere with the absorption of medication and may cause dehydration and chemical imbalance. Medical advice should be sought.

GLOSSARY

ANGIOGRAPHY: An internal x-ray of the heart and blood vessels, which may be taken at the time of cardiac catheterisation, (see below). In particular, this test assesses the coronary arteries, (the blood vessels which supply the heart muscle).

ARRHYTHMIA: An abnormal rhythm or irregularity of the heart beat. The heart beat may be either too fast (*tachycardia*) or too slow (*bradycardia*). Arrhythmias may cause symptoms such as breathlessness or light-headedness.

CARDIAC CATHETERISATION: A special test used for many forms of heart disease, which is sometimes performed in ARVC. At cardiac catheterisation a fine tube is passed from a blood vessel, (usually in the top of the leg), to the heart, using x-ray guidance. The structure and function of the heart can then be assessed.

ECHOCARDIOGRAM: A non-invasive test where images of the heart can be obtained through the use of high frequency sound waves.

ELECTRICAL CARDIOVERSION: An arrhythmia, such as atrial fibrillation, may be stopped by the application of an electric shock to the chest. If this procedure is necessary, it is carried out under general anaesthesia.

ELECTROCARDIOGRAM (ECG): A non-invasive test where a tracing of the passage of electrical impulses through the heart muscle is obtained.

ELECTROPHYSIOLOGICAL STUDY or EPS: In this test catheters are introduced into the heart as in cardiac catheterisation, (see above). These catheters can record and stimulate the electrical activity of the heart.

EXERCISE (STRESS) TESTING: Exercise capability may be tested using either a treadmill or a stationary bicycle. During an exercise test a doctor and technician will monitor a patient's symptoms, ECG, blood pressure and, sometimes, breathing.

HOLTER MONITOR: A continuous recording of the heart beat over 24 to 48 hours. Adhesive electrodes are placed on the chest, wires from these go to a special cassette recorder, which is worn on a belt. A Holter monitor detects irregularity of the heartbeat, otherwise known as arrhythmia.

IMPLANTABLE CARDIOVERTER DEFIBRILLATOR (commonly abbreviated to ICD): A specialised device, which is implanted in a similar way as a pacemaker. It can recognise when a heart rate is excessively fast and responds by either pacing the heart or delivering a small electrical shock to restore the normal heart rhythm. The ICD can also serve as a conventional pacemaker to deliver the necessary impulses when the heart rate is too slow.

IMPLANTABLE LOOP RECORDER: This is a small device, which can be implanted to assist the diagnosis of an arrhythmia. The patient activates the device when symptoms occur to record any rhythm disturbance.

PALPITATION: An uncomfortable awareness of an irregularity or change in the heartbeat. Palpitation may be due to a normal heartbeat made more prominent by anxiety or exercise, or may be caused by an arrhythmia.

PRE-SYCOPE: Symptoms causing a near faint, but without actually losing consciousness.

SIGNAL AVERAGE ECG: A more sensitive type of ECG designed to record more detailed information of the heart's electrical activity.

SYCOPE: Loss of consciousness.

VENTRICLES: The main pumping chambers of the heart, one on the right side and one on the left.

VENTRICULAR FIBRILLATION: An arrhythmia arising in the ventricles where the electrical activity of the heart is chaotic. The heart's pumping action is uncoordinated resulting in a lack of output of blood. Emergency resuscitation is required to correct this arrhythmia.

FOR MORE INFORMATION VISIT



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ARVC

(Arrhythmogenic Right Ventricular
Cardiomyopathy)

Arrhythmogenic Right Ventricular Cardiomyopathy (ARVC) is an inherited heart muscle disorder where damaged heart muscle is gradually replaced by scar tissue and fat. A recognised cause of sudden death in the young, ARVC may cause abnormal electrical heart rhythms and weakening of the pumping action of the heart.

In many cases, the disease does not limit the quality or duration of life. However, a proportion of people with ARVC develop complications, all of which are treatable. Evaluation and follow-up by a cardiologist is therefore recommended.

How does ARVC affect the heart?

Heart muscle cells are replaced by fibrous and fatty tissue. Initially this appears to be patchy and predominantly affects the right ventricle. A progressive pattern of disease is often seen. It is now recognised that the left side of the heart can also be affected.

One of the most significant advances in our understanding of ARVC came with the identification of disease-causing mutations in the genes encoding the proteins plakoglobin and desmoplakin. This discovery paved the way for further gene identification studies, which are currently in progress. But how exactly do the mutant proteins affect the heart to cause the disease?

This will be an active area of research for years to come. Plakoglobin and desmoplakin are key components of the junctions responsible for linking cells to one another.

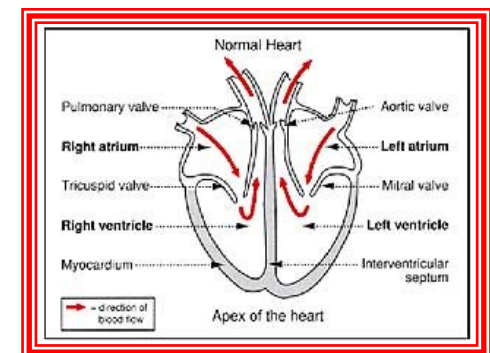


Figure 1 - The Normal Heart: Structure and function

The Heart

The heart is a four-chambered muscular pump. The two smaller chambers, or atria, contract first, filling the larger ventricles. The right ventricle pumps blood into the lungs, where it absorbs oxygen. Oxygenated blood returns to the left atrium and, from there, the left ventricle. The left ventricle pumps the oxygenated blood to the rest of the body. Blood is then returned by the veins to the right atrium and then the right ventricle to repeat the cycle.

Abnormal heart rhythms

Arrhythmia is the medical term for any abnormal heart rhythm. Ventricular premature beats are not dangerous in themselves and are relatively common in healthy people. They can result from simple things like coffee, cigarettes, lack of sleep, anxiety or alcohol. However, ventricular premature beats may also be a sign of an underlying heart condition, particularly when occurring in large numbers. People with ventricular premature beats may experience palpitation (an awareness of the heart beating out of rhythm or fluttering).

It can cause a sudden fall in blood pressure and may cause loss of consciousness. Fainting or passing out is referred to as syncope. The sensation of being about to faint, frequently perceived as light-headedness or dizziness, is termed pre-syncope.

Ventricular fibrillation represents complete electrical chaos within the ventricles. The heart is completely unable to pump during ventricular fibrillation.

Cardiac arrest occurs and prompt resuscitation is necessary if the person is to survive. Applying an electric shock (defibrillation) may help the heart to return to a normal rhythm.

ARVC has a genetic basis

Our DNA contains a set of coded instructions for the protein building blocks of life. This blueprint is copied from one generation to the next. Occasionally, a spelling mistake occurs during the copying process. The resulting genetic alteration is termed a mutation. We inherit two copies of most of our genes, one from each of our parents. Every child of a person with a faulty gene has a 50% chance of inheriting it.

The most common pattern of inheritance of ARVC appears to be autosomal dominant with incomplete penetrance. Autosomal dominant transmission means that a single copy of the abnormal gene is sufficient to cause the disease.

On the other hand, a person with the abnormal gene will not necessarily develop any features of the disease. This latter phenomenon is termed incomplete penetrance.

What are the symptoms of ARVC?

Palpitation. People describe this in different ways:

- ♥ 'My heart gave such a flutter'
- ♥ 'I could feel my heart racing suddenly'
- ♥ 'My heart keeps missing beats'
- ♥ 'I could sense my heartbeat was irregular'
- ♥ 'I could feel my heart thumping away/ pounding against my chest'
- ♥ 'I felt this discomfort in my chest'

Light-headedness (Pre-syncope)

- ♥ 'I felt light-headed/ giddy/ dizzy/ woozy/ faint'
- ♥ 'I felt like I was about to faint/ pass out'
- ♥ 'My head was spinning'
- ♥ 'I had a funny turn'
- ♥ Fainting (Syncope) - By definition, an episode where loss of consciousness occurs

The above symptoms may indicate rhythm disturbances and often occur during strenuous activity. Chest pain, breathlessness, sweating, and nausea may also accompany arrhythmia. More rarely, ARVC can present with the symptoms of weakening of the heart muscle pump, e.g.

- ◆ Shortness of breath on exertion
- ◆ Breathlessness on attempting to lie flat
- ◆ Waking up in the middle of the night with difficulty in breathing
- ◆ Swelling of ankles or, in more severe cases, the abdomen

Unfortunately, sudden death may be the first manifestation of ARVC in some people. This highlights the importance of screening the family members of patients with ARVC.

Diagnosis of ARVC

A single test is seldom able to confirm or rule out ARVC. Establishing the diagnosis is often difficult because many of the findings are subtle and nonspecific – that is, they are seen in other conditions and are not particular to ARVC.

The relevant investigations are outlined below:

The 12-lead electrocardiogram or ECG - Various ECG abnormalities are included in the diagnostic criteria for ARVC. However, many patients with ARVC have a normal recording or only minor changes.

Signal averaged ECG - A positive signal averaged ECG is one of the minor criteria for ARVC.

24 or 48-hour 'Holter' monitor - The Holter monitor is a tape that is fitted to a patient for 24 or 48 hours, and provides a continuous recording of the heart rhythm.

Exercise test - Strenuous activity may trigger arrhythmias in ARVC. The response to physical exertion is assessed by monitoring the heart rhythm while the patient exercises on a treadmill or bicycle.

Echocardiogram - The echocardiogram is an ultrasound scan of the heart. Safe, relatively fast, and available at most hospitals, it is usually the first choice for imaging the heart.

Cardiac catheterization - This is an invasive procedure for which the patient is admitted to hospital. This study is often indicated in the investigation of collapse or chest pain, when coronary artery disease may be responsible. The angiogram is the best way of looking for the narrowings in the coronary arteries that cause heart attacks and angina.

Electrophysiological (EP) study - Invasive electrophysiological testing involves the insertion of catheters into the heart for two main purposes:
(1) To obtain a detailed study of the electrical activity of the heart
(2) To attempt to provoke arrhythmia under controlled conditions

An EP study may also be offered to patients who have suffered multiple fainting spells, or a previous cardiac arrest, and have no evidence of arrhythmia on ECG, Holter monitor, or exercise test.

Implantable loop recorder (ILR) - The ILR is a thumb-sized device implanted just beneath the skin that is capable of monitoring and recording the heart rhythm. Day case admission is arranged, and the device is inserted on the left side of the chest under local anaesthetic.

Management of ARVC - Although no cure currently exists for ARVC, there are many effective ways of averting and treating its complications. The management of ARVC has four specific aims:

- ◆ Identifying people at risk of sudden cardiac death and offering protective measures
- ◆ Preventing or controlling arrhythmia
- ◆ Improving the functioning of the heart muscle pump
- ◆ Screening family members for the condition

What can be done to prevent sudden death? - The implantable cardioverter-defibrillator (ICD) is the best available protection against sudden cardiac death

Medication to control arrhythmia - The following medication is commonly used to suppress ventricular arrhythmia in ARVC:

- ◆ Beta-blockers. (Examples: atenolol, bisoprolol, metoprolol and carvedilol) In simple terms, beta-blockers interfere with the action of adrenaline on the heart. Since adrenaline seems to play a role in causing rhythm disturbances in ARVC, beta-blockers are often the first-line treatment. They are usually well tolerated.
- ◆ Amiodarone - Amiodarone is a powerful anti-arrhythmic agent that is effective against many different rhythm disturbances. However, careful observation is required for a number of possible drug reactions.
- ◆ Sotalol - Anti-arrhythmic agents are classified by their mechanism of action into four categories. Beta-blockers comprise class II, while amiodarone belongs to class III. Sotalol shares some of the properties of both class II and class III agents and is recommended by some authorities for the treatment of ARVC.