

2. ARRHYTHMOGENIC RIGHT VENTRICULAR CARDIOMYOPATHY ^{1,2}

Arrhythmogenic Right Ventricular Cardiomyopathy (ARVC) is probably the second most common cause of unexpected sudden death in the young. The incidence of this condition is now thought to be higher than previously believed, due to the availability of better diagnostic techniques and general awareness of the disorder among cardiologists. It is anticipated that more information regarding the condition will be available in the coming years, to help to recognise the pathology. The disorder is characterised by a progressive replacement of normal right ventricular muscle cells by fibrous tissue and fat. Initially this may only involve specific regions of the right ventricle but later on it becomes global and may even involve the ventricle as well. ARVC was first recognised in the late 1970's and the Cardiovascular Group at the University of Padova in Italy have developed particular expertise in the condition.

What causes ARVC?

The precise cause of ARVC is not known. The condition is usually familial and is passed on in the genes from one generation to the next. The pattern of inheritance is autosomal dominant such that the child of an affected parent will have 50% chance of inheriting the abnormal gene. The disease affects men and women equally and has been recognised in people of diverse ethnic origin. The Northern region of Italy seems to have highest prevalence of the disease.

Symptoms

Clinical presentation is usually with symptoms of arrhythmia and occasionally with sudden death. Typical arrhythmia symptoms are of *rapid heart beat* associated with *light-headedness* or *fainting episodes*. Unlike most cardiomyopathies, *shortness of breath* and *chest pains* are unusual symptoms and tend only to occur in older patients.

How is ARVC diagnosed?

The diagnosis of ARVC can be extremely difficult and usually requires specialist expertise. There are no 'gold standard' investigations as there are available for diagnosing HCM. The investigations are similar to those performed in the diagnosis of hypertrophic and other cardiomyopathies and include an electrocardiogram and a two dimensional echocardiogram. The features, however, are usually very subtle, as in the early stages the condition is often confined only to the right ventricle. Other methods of imaging the right side of the heart are sometime useful and these include magnetic resonance imaging. Despite all these investigations, the condition can still be missed.

What treatment is available?

The majority of patients with this condition are asymptomatic for many years unless arrhythmias develop. Treatment in the majority then aims to prevent or at least control arrhythmia with drugs. When drug treatment is unsuccessful, an implantable cardioverter defibrillator and other specialised treatments may be necessary, these include:

- *Catheter Ablation* – This involves the delivery of electrical energy via a catheter inserted in the groin to the area in the right side of the heart where the arrhythmia is originating from. The overall effect is to create a small scar which is incapable of transmitting any arrhythmia.
- *Surgical Ablation* – In individuals where the arrhythmias are originating from many different sites in the right side of the heart. Occasionally it is necessary to perform open-heart surgery. During the operation, the electrical pathway of the arrhythmia can be mapped, identified and destroyed.

¹ The following medical information has been kindly provided by CRY and is available for download in the website: <http://www.c-r-y.org.uk>.

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