



## Review

# Arrhythmogenic right ventricular dysplasia/cardiomyopathy. A review and update

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Received 7 February 2005; received in revised form 19 May 2005; accepted 4 July 2005

**Abstract**

The arrhythmogenic right ventricular dysplasia/cardiomyopathy is an important cause of sudden arrhythmic death, often exertional, in young individuals and athletes. Although the aetiology remains partially unknown, genetic abnormalities have been demonstrated. Reported prevalence is 1 in 5000 individuals but it is considered there are many non-diagnosed cases.

The characteristic pathologic finding is the progressive fibro-fatty replacement of the right ventricular myocardium. The clinical manifestations vary from asymptomatic patients with an episode of sudden cardiac death as first symptom to chronically symptomatic patients with recurrent palpitations and/or right or biventricular failure.

Approximately a third of the patients show the characteristic Epsilon wave in the 12-lead ECG which is a useful screening test. Signal-averaged ECG frequently demonstrates late potentials. The two-dimensional echocardiography, magnetic resonance imaging, computerized tomography and right ventricular cineangiography show morphologic abnormalities in the right ventricle.

Therapy is directed to prevent and/or treat malignant ventricular tachyarrhythmias with medications, implantable cardioverter defibrillator and radiofrequency ablation in selected cases.

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*Keywords:* Cardiomyopathy; Sudden death; Arrhythmia; Right ventricle

**1. Introduction**

Arrhythmogenic right ventricular dysplasia/cardiomyopathy (ARVD/C) is a cardiac disease that affects mainly the right ventricular myocardium. It has been increasingly recognized as a major cause of sudden cardiac death in young individuals and athletes.

Dalla Volta and co-workers described it first in 1961 in Italy. However, Frank and Fontaine coined the term arrhythmogenic right ventricular dysplasia in 1978 [1]. Recently, based on its nature of progressive myocardial disease of unknown aetiology, ARVD has been more appropriately included among the cardiomyopathies in the classification proposed by a task force of the World Health Organization/International Society and Federation of Car-

diology [2]. Two international registries in USA and Europe are under way aiming to determine the clinical, pathological and genetic aspects of ARVD/C, validate the diagnostic criteria and define strategies for disease management and sudden death prevention [3–5].

Although there are many topics that require further investigations, much more information about this new cardiomyopathy is now available.

**2. Aetiology**

Although a genetic predisposition is recognized and abnormal genes have been identified during the last two decades [6–12], the precise aetiology of ARVD/C remains to be established. Several authors reported a familial presentation [13–19] with symptomatic and asymptomatic members and different degrees of myocardial compromise.

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It is typically inherited as an autosomal dominant trait with incomplete penetrance and variable expression [18]. We described an autosomal recessive form of the disease associated with anterior polar cataract in several members of a family [19]. Interestingly, ARVD/C and the anterior polar cataract genes are located in contiguous loci on the chromosome 14. An autosomal recessive form associated with palmoplantar keratosis is called Naxos disease [6]. A locus for Naxos disease was located on chromosome 17q21 [6,20]. A mutated gene produces a protein called plakoglobin involved in the genesis of the disease [21]. Plakoglobin is a component of desmosomes and participates in maintaining tight cell to cell junction. It was found a homozygous 2 base pair deletion in this gene in affected individuals. A recessive mutation in the desmoplakin gene was described as the aetiology of Naxos disease [22].

Nine loci associated with ARVD/C have been found. Disease loci for the autosomal dominant form have been mapped to chromosomes 14q23–24 (ARVD1), 1q42–q43 (ARVD2), 14q12–q22 (ARVD3), 2q32 (ARVD4), 3q23 (ARVD5), 10p12–p14 (ARVD6), 10q22 (ARVD7), and 6p24 (ARVD8) [7–12,23–25].

Some involved genes have also been identified. The gene at locus 6p24 (ARVD8) encodes desmoplakin [26] which is a component of desmosomes and adherens junctions. Abnormal cell junction is followed by cell death and fibro-fatty replacement.

Clinical and pathological evidence support the existence of a degenerative process (atrophy or dystrophy) attributable to a progressive death of myocardial fibers. Mallat and Valente [27,28] proposed that apoptosis (programmed cell death) could be the underlying mechanism in the pathogenesis of this disease. It is unknown how the apoptotic mechanism is initiated but may be related to altered intracellular calcium concentration. It was also suggested that superimposed viral infections (myocarditis) could explain the severity and poor prognosis in some patients.

### 3. Incidence and prevalence

The incidence is unknown since this pathological process can be totally asymptomatic in the young with the first manifestation being an episode of sudden death. However, a prevalence of approximately 1 in 5000 people [29] has been estimated but it could be higher because of the existence of many non-diagnosed or misdiagnosed cases. It has been found in up to 20% of sudden deaths in young people and it is considered as the most common cause of exercise-related sudden death among young Italian athletes. Although the first symptoms usually appear during adolescence or early adulthood, patient ages range from 4 to 73 years. Fontaine et al. reported an approximate 80% predominance in males

[30]. The incidence of familial presentation varies from 15% to 50% in the literature [13–17]. Interestingly, there is a higher frequency in regions such as the Veneto, in Italy. However, cases of ARVD/C have been found worldwide.

### 4. Anatomic and pathologic features

Macroscopic findings frequently observed in ARVD/C include dilatation of the right ventricle, aneurysmal dilata-tions of the infundibular, apical and subtricuspid areas (the so-called triangle of dysplasia), and replacement of normal myocardium with fibro-fatty tissue and thinning of the right ventricular wall [15–31]. Although the ventricular wall appears thinner, there is no contact between epicardium and endocardium as in the Uhl's anomaly.

The interventricular septum and the left ventricle are typically spared until late stages of the disease. The pathological process advances from the subepicardium to the endocardium. Therefore, an endocardial biopsy in a suspected case may be normal.

Microscopic findings include the replacement of external and medial layers of right ventricular myocardium and, occasionally, the left ventricular myocardium by fibro-fatty tissue (Fig. 1). This abnormal tissue is located around or interspersed among the normal fibers. There is a coexistence of normal and partially degenerated strand of fibers that provide the substrate for slow conduction and reentrant ventricular arrhythmias observed in these patients. To remark, only fibrosis around or between surviving myocardial fibers is diagnostic of ARVD/C.

It was postulated that fibrosis results from a super-imposed myocarditis on a genetically predisposed abnormal myocardium. Coxsackie B and enteroviral RNA viruses were identified in biopsy samples [32].

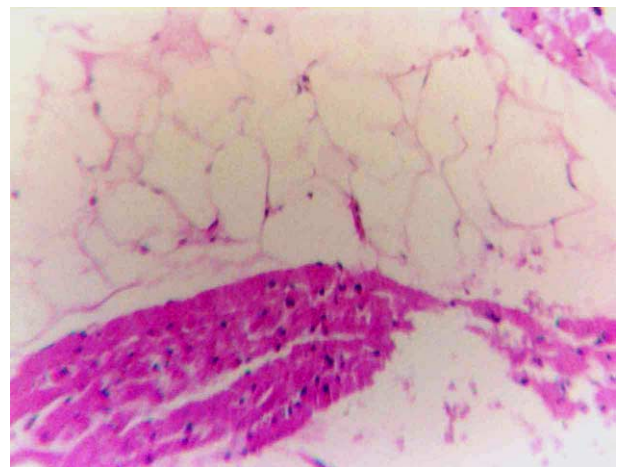


Fig. 1. Histological pattern of the right ventricular wall in a 48-year old woman with ARVD/C. Profuse fatty tissue around spared strands of cardiomyocytes. (From Dr. Farré, Sanatorio Parque, Rosario, Argentina).

The coronary arteries are usually spared but thickening of the media may be observed. This narrowing of the lumen could explain the atypical chest pain referred by some patients. In rare cases, complete obstruction of the lumen is present.

The term “biventricular dysplasia” is used when there is involvement of the left ventricle [33]. In that case, the apical and inferior areas are mainly affected.

## 5. Clinical presentation

Diagnostic criteria have been proposed by the working group on pericardial and myocardial diseases of the European Society of Cardiology and the Scientific Committee on Cardiomyopathies of the International Society and Federation of Cardiology [34]. The presence of two major criteria or one major with two minor criteria or four minor criteria from different groups (Table 1) supports the diagnosis of ARVD/C. The use of these criteria in the clinical practice is strongly recommended but the clinician must consider that mild forms of the disease may not be encompassed by these criteria.

The clinical presentation varies from silent forms with an exercise-related episode of syncope or sudden death as first manifestation to biventricular cardiac failure that requires cardiac transplantation. Right ventricular failure is usually observed in older patients. However, we assisted a few young patients with right heart failure as the presenting form of the disease. Common symptoms include palpitations during physical activity, fatigue, dizziness, atypical chest pain and one or more episodes of syncope.

Fifty percent of the patients have normal physical examination. When present, abnormal physical signs may include asymmetric chest wall, giant a waves on the neck inspection, tricuspid regurgitation murmur in patients with severe dilatation of right ventricular chambers and right-sided S3–S4. The S2 can be split when right ventricular ejection is prolonged.

The presence of ventricular arrhythmias including isolated premature ventricular contractions and non-sustained or sustained VT with left bundle branch block morphology represent a particularly frequent (50%) and characteristic form of presentation. As mentioned, ventricular fibrillation (VF) and sudden death can be observed during exercise or physical activity. However, episodes of sudden death during sleep were also seen in patients with normal physical examination.

Supraventricular arrhythmias including atrial premature contractions, flutter, tachycardia and fibrillation were reported in 25% of patients with ARVD/C and ventricular arrhythmias. Supraventricular tachyarrhythmias alone are uncommonly observed.

There is an overlapping of clinical manifestations with Brugada Syndrome, and discrimination between both cardiomyopathies may be particularly difficult because

Table 1

Criteria for diagnosis of right ventricular dysplasia (from McKenna WJ, et al. [34])

I Global and/or regional dysfunction and structural alterations*	
MAJOR	
Severe dilatation reduction of right ventricular ejection fraction with no (or only mild) LV impairment.	
Localized right ventricular aneurysm (akinetic or dyskinetic areas with diastolic bulging).	
Severe segmental dilatation of the right ventricle.	
MINOR	
Mild global right ventricular dilatation and/or ejection fraction reduction with normal left ventricle.	
Mild segmental dilatation of the right ventricle.	
Regional right ventricular hypokinesia.	
II Tissue characterization of walls	
MAJOR	
Fibro-fatty replacement of myocardium on endocardial biopsy.	
III Repolarisation abnormalities	
MINOR	
Inverted T waves in right precordial leads (V2 and V3) (people aged more than 12 years, in absence of right bundle branch block).	
IV Depolarisation/conduction abnormalities	
MAJOR	
Epsilon waves or localized prolongation (>110 ms) of the QRS complex in right precordial leads (V1–V3).	
MINOR	
Late potential (signal averaged ECG).	
V Arrhythmias	
MINOR	
Left bundle branch block type ventricular tachycardia (sustained and non-sustained) (ECG, Holter, exercise testing).	
Frequent ventricular extrasystoles (more than 1000/24 h) (Holter).	
VI Family history	
MAJOR	
Familiar disease confirmed at necropsy or surgery.	
MINOR	
Familial history of premature sudden death (<35 years) due to suspected right ventricular dysplasia.	
Familial history (clinical diagnosis based on present criteria).	

\*Detected by echocardiography, angiography, magnetic resonance imaging, or radionuclide scintigraphy. ECG: electrocardiogram; LV: left ventricle. I acknowledge the BMJ Publishing Group (Br Heart J 1994; 71:215–218) for the permission to reproduce this material.

ARVD/C may at times mimic Brugada syndrome and structural abnormalities may only be found at time of autopsy.

In summary, ARVD/C is a polymorphic disease with different clinical presentations. Clinical suspicion arises from the presence of right precordial ST–T changes on the ECG and a familial history of sudden death. After non-invasive and invasive diagnostic tests are performed, it is strongly recommended a genetic diagnosis if available.

### 5.1. Risk of exercise

As stated, ARVD/C is a frequent cause of sudden cardiac death in athletes. Higher frequency is noted in selected populations such as in northern Italy. One report showed [35] that most of the athletes who died suddenly presented one or more risk factors on previous screening. Recent studies on

risk stratification agree that (a) history of syncope, (b) appearance of symptoms at a young age, (c) severely progressed disease to the right ventricle, and (d) left ventricular involvement are related to sudden cardiac death [36,37]. The presence of exercise-induced ventricular arrhythmias suggests the involvement of catecholamines. Increased sensitivity to catecholamines could be caused by an abnormal cardiac sympathetic function. The islands of fibrofatty tissue would generate macro-reentry circuits, the electrophysiological substrate for malignant ventricular tachyarrhythmias [38]. Interestingly, 24 h Holter monitoring shows a progressive increase in the sinus rate before the onset of VT which suggest progressive sympathetic stimulation.

As consequence, patients with ARVD/C should not participate in competitive sports and avoid any activity that triggers symptoms as palpitations, presyncope or syncope.

## 6. Non-invasive tests

The chest X-ray is usually normal unless right ventricular enlargement and failure are present. The 12-lead ECG shows abnormal changes in 90% of the cases, but some authors reported lower rates [39]. The presence of abnormal findings depends on the stage and severity of the disease. The most common finding is a negative T wave in V1–V3 and sometimes through V6. However, because of children and young women have T-wave inversions in right precordial leads, this is consequently a diagnostic criterion only in adults and patients without right bundle branch block or right ventricular enlargement. The characteristic finding is the presence of Epsilon waves, which can be observed in 30% of the cases. They are located at the beginning of the ST segment or the end of QRS complex in right precordial leads and represent ventricular post-excitation, known as late potentials which are recorded on the signal-averaged ECG and are considered the electrophysiological substrate for ventricular tachyarrhythmias. The signal-averaged ECG is also useful for screening purpose or detection of ARVD/C in family members. Serial ECG recordings in a patient referred to our Service showed Epsilon waves after years and when the right ventricle was severely damaged (Fig. 2).

Depolarization delays, either complete or incomplete right bundle branch block and QRS prolongation are also observed. The selective QRS prolongation in V1–V3 longer than 110 ms compared with lead V6 and the Epsilon waves are considered major diagnostic criterion. Low voltage QRS in limb leads, P wave higher than 2.5 mV, and ventricular premature beats with left bundle branch morphology may be present.

### 6.1. Echocardiogram

Echocardiography is the first line imaging approach in evaluating patients with suspected ARVD/C or in screening

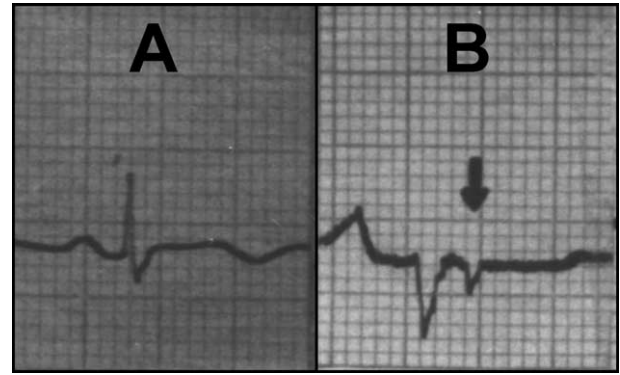


Fig. 2. V2 leads recordings from a man affected by ARVD/C. (A) At the age of 10, a non-specific ST–T change is observed. (B) At the age of 27, the same lead recorded with double standard amplitude (20 mm/1 mV) shows a different QRS morphology and a negative Epsilon wave on the ST–T.

family members. It is useful to evaluate right and left ventricular size and functions that are considered major and minor criteria for the diagnosis of ARVD/C. However, specificity and sensitivity are not high. Echocardiographic studies may demonstrate right ventricular enlargement or multiple areas of dilatation, and dyskinetic or hypokinetic zones mainly in the infero-basal and apical regions [40,41]. Increased thickness of the moderator band, physiological trabeculations of the right ventricular apex and dilatation of right ventricular outflow tract were also described [42]. Kisslo considered that the most important diagnostic parameters were right ventricular end-diastolic and end-systolic diameters, and the ratio of the right ventricular and left ventricular end-diastolic and end-systolic diameters [43]. Nevertheless, this ratio is now considered not a sensitive criterion and the measurement of the absolute diameter of the right ventricle difficult to obtain. Recently, Yoerger et al. reported the results of detailed echocardiograms performed in 29 patients who were newly diagnosed with ARVD/C. They found significant right atria and right ventricular enlargement and decreased RV function. They pointed out that the right ventricular outflow tract was the most commonly enlarged dimension in their patients [44].

The left ventricle may show severe hypokinesia and low ejection fraction, as described by Pinamonti et al. [45] in a patient with left ventricular involvement.

It must be emphasized that the majority of the patients have structural abnormalities focalized in the infero-basal region.

Doppler-echocardiography may demonstrate abnormal diastolic function of right ventricle and tricuspid regurgitation with normal jet velocity.

### 6.2. Magnetic resonance imaging (MRI) and computerized tomography (CT)

MRI and CT provide useful information regarding the structural abnormalities previously described. The MRI

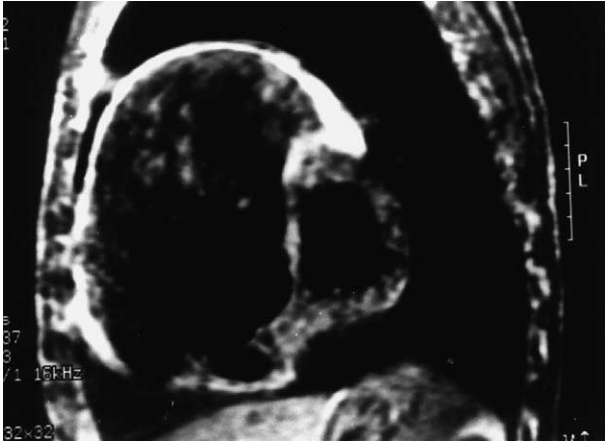


Fig. 3. Magnetic resonance imaging in a 27-year old man with a history of palpitations and right cardiac failure (same patient that in Fig. 2). Lateral view shows a severely dilated right ventricle with intense signal due to replacement of normal myocardium by fibro-fatty tissue (high density).

appears to be the best imaging technique as it provides information about volumes, wall motion right ventricular abnormalities, but principally because it shows intramyocardial fat deposits seen as an intense signal (Fig. 3) compared with the normal myocardium [46]. MRI provides the advantage of distinguishing fat from muscle but as the right ventricular wall is thinner than normal, adequate assessment is sometimes difficult. Bomma et al. demonstrated a high frequency of misdiagnosis of ARVD/C when abnormalities are detected solely on MRI. They showed an over-reliance on the presence of intramyocardial fat and wall thinning as the main causing factor of the misdiagnosis [47]. There is a significant interobserver variability probably due to lack of experience with ARVD/C and some have suggested that the use of MRI for the diagnosis of ARVD/C should be limited to centers with experience on this disease. In summary, although MRI is a very useful test, its specificity is decreased by all these factors.

CT demonstrates an increase in epicardial adipose tissue delineated by densitometric analysis of the image. This test also shows other morphologic abnormalities described above such as localized or diffuse compromise and dilatation of the right ventricle, thinning of the wall and hypokinesis, prominent trabeculations with low attenuation.

## 7. Invasive tests

### 7.1. Cineangiography

Right ventricular cineangiography is considered by many experts the most useful invasive test to diagnose ARVD/C and evaluate right ventricular function when performed and interpreted with a strict methodology. Some technical aspects of right ventricular angiography should be considered, such as: (a) the use of special catheters; a Berman balloon is recommended to avoid triggering premature

ventricular contractions; (b) the use of digitalized ventriculography (50 frames/s) provides optimal analysis, (c) placement of the catheter tip at a non-arrhythmogenic position, (d) the use of two orthogonal views or rather the combination of the 30° right anterior oblique and 60° left anterior oblique views which gives a more complete analysis of the segments preferentially involved in ARVD/C including infundibulum, anterior RV free wall, apex, and the inferior wall, particularly subtricuspid area, e) the slow dye injection after a test injection of 10 ml saline solution is recommended.

Good quality angiograms allow global and regional analysis of morphology and motion. Dyskinetic or akynetic zones in the infundibular, apical or subtricuspid regions have specificity greater than 90% [48]. Increased end-diastolic volume with abnormal and generalized wall motion, persistence of dye during more than 20 beats or multiple outpouchings of the inferior right ventricular wall or in aneurysms, tricuspid or mitral valve prolapse, isolated diastolic protrusions and systolic dyskinesia of the outflow tract are also found. It is also emphasized the finding of transversely arranged hypertrophic trabeculae thicker than 4 mm separated by deep fissures, and coarse trabeculae in the apical region distal to the moderator band. Although the right ventricular volume is almost always increased, it is often difficult to assess right ventricular enlargement because of the complex geometric shape of this chamber. It should be remembered that a negative angiogram does not rule out ARVD/C [49] and the borderline between normal and pathology is very hard to define in some cases. The positive predictive value of a right ventricular angiogram is more than 85%, with a negative predictive value of approximately 95% [50].

### 7.2. Invasive electrophysiologic study (EPS)

The EPS with programmed ventricular stimulation with or without isoproterenol infusion is performed to reproduce VT or VF, induce supraventricular tachycardia and evaluate antiarrhythmic drugs efficacy and implantable cardioverter defibrillator therapy. A continuous infusion of isoproterenol induced VT in most patients with ARVD/C and represents a highly sensitive test for its diagnosis. This finding is consistent with the observation that VT in these patients is usually preceded by a heart rate increase, suggesting that a change in the sympathetic tone acts as a trigger [51].

However, the usefulness of EPS is controversial. Di Biase et al. were only able to induce sustained VT in patients with spontaneous VT [52]. Thus, EPS would not be a valuable test for risk stratification in patients with ARVD/C.

### 7.3. Endomyocardial biopsy

The presence of abnormal structural findings confirms the diagnosis. However, the endomyocardial biopsy is

frequently negative because of the characteristic patchy infiltration and the progression of the disease from outer to inner myocardial layers. The site of biopsy is critical. As it is usually done in the right ventricular septum for safety reason, and the typical pathologic changes are present in the right ventricular free wall, the samples show frequently normal myocardial tissue.

Right ventricular free wall biopsy involves the risk of cardiac perforation if an abnormal area is punctured.

## 8. Prognosis

Long-term follow-up studies in large populations are not available yet, making the prognosis in symptomatic patients difficult to define. There is even less information about clinical outcome on asymptomatic affected individuals. However, it is known that the natural history of the disease is related to the electrical instability that can precipitate fatal arrhythmic events at any time during the patient's life.

Studies in small groups of patients demonstrated different mortality rates. A follow-up of 33 patients (range 1.5–12 years) showed that the clinical course of this disease is quite different [53]. Berder et al. reported 4% mortality in a 4.5 years follow-up [54]. In contrast, Pinamonti et al. described 19% mortality after an average follow-up of 8.2 years [55]. A long-term follow-up of 15 patients with ARVD/C reported that the incidence of sudden death is 1% per year in patients with empiric unguided antiarrhythmic drug treatment [56] and 3% without treatment. It is noteworthy that cardiac arrhythmias may disappear spontaneously in some patients [57].

As the ARVD/C is a progressive disease that involves partially or globally the right ventricle which becomes more diffusely damaged with time, the existence of right ventricular failure implies progression and severity of the disease. Although macroscopic or histological changes of the left ventricle were found in many patients, left cardiac failure is uncommon and it is present only in later stages of the disease. The biventricular failure mimics a dilated cardiomyopathy of any origin, adding complications such as thromboembolic events and atrial fibrillation which imply a poorer prognosis.

According to some authors, the occurrence of myocarditis or biventricular compromise with congestive heart failure adds an additional 1% risk of mortality per year [58,59].

### 8.1. High risk patients

Recent studies on risk stratification agree that patients with the following characteristics have a higher risk of sudden death: (a) history of syncope; (b) appearance of symptoms at a young age; (c) severely progressive disease to the right ventricle and (d) left ventricular involvement [36,60].

A baseline study for risk evaluation includes a detailed clinical history, 12-lead ECG, 24 h Holter monitoring, exercise stress test and signal-averaged ECG.

## 9. Therapy

The therapy is directed to treat the heart failure if present and/or prevent sudden cardiac death. Patients with ventricular or biventricular cardiac failure should receive the standard therapy including diuretics, angiotensin converting enzymes inhibitors, and digitalis as well as anticoagulants.

The main goal of the antiarrhythmic strategy is to prevent the appearance or to avoid recurrences of ventricular tachyarrhythmias. It should be remembered that all identified or suspected patients are at risk of sudden death even if they have no symptoms or diagnosed ventricular arrhythmias. For that reason, one of the main clinical dilemmas is whether or not to consider prophylactic treatment in patients with minor or no symptoms of ARVD/C diagnosed during a family screening or cardiovascular evaluation for other reasons (primary prevention). Management strategies of patients with ARVD/C should be individualized and therapeutic strategies based on the experience of each medical team.

As mentioned, patients with ARVD/C have to avoid competitive sports and any activity that cause symptoms of palpitations, presyncope or syncope.

### 9.1. Antiarrhythmic drug treatment

It is the first choice treatment for patients with well tolerated and non-life threatening ventricular arrhythmias. Amiodarone has been extensively used and seems to be quite effective. One study reported that sotalol is the most effective drug in the treatment of inducible and non-inducible VT with an efficacy of 68% and 82%, respectively [61]. Therefore, the initial therapy with sotalol is recommended for primary prevention in patients with ARVD/C. If sotalol is ineffective, response to other drugs is unlikely and therefore a non-pharmacologic therapy should be indicated to these patients. The evaluation of drug efficacy may be non-invasively achieved by 24 h Holter monitoring and/or stress testing to demonstrate reduction in arrhythmic events.

### 9.2. Non-pharmacological treatment

#### 9.2.1. Implantable cardioverter defibrillator

The implantable cardioverter defibrillator (ICD) is the most effective therapy to prevent sudden death. It is the first choice treatment in patients with life-threatening ventricular arrhythmias and unsuccessful or non-tolerated drug therapy. In patients with syncope and non-inducible VT, the ICD is indicated because of drug therapy cannot be tested.

It is also recommended in resuscitated patients with VT or VF, patients with poorly tolerated VT and severe compromise

of the right ventricle. It is considered a class I recommendation for secondary prevention and a class IIa recommendation for primary prevention [62]. Sotalol can be added to reduce the number of ICD discharges, improving the patients' quality of life and prolonging the lifetime of the battery.

There are possible complications related to the catheter implantation. Firstly, as the right ventricular wall is thinner than normal, perforation and cardiac tamponade are more likely. Secondly, the fibro-fatty replacement of the right ventricular myocardium may lead to inadequate sensing of ventricular tachyarrhythmias and ICD failure.

Recently, Wichter et al. published their experience of long-term follow-up and complications in 60 patients with ICD implantation. They observed an improvement in long-term prognosis by ICD therapy in high-risk patients with ARVD/C. However, a considerable cumulative incidence of lead-related complications such as higher pacing thresholds and lower amplitude of endocardial potentials during extended long-term were noted [60]. Fontaine and Prost-Squarcioni highlighted the high incidence of long-term complications in Wichter's patients, adding his observation of a high defibrillation threshold based on his experience [63].

### 9.3. Radiofrequency ablation

Radiofrequency ablation (RFA) should be reserved for patients with well-tolerated and haemodynamically stable VT who do not respond to antiarrhythmic drugs. A report including 50 patients showed that success rate was 32%, 45% and 66% after one, two, and three ablation sessions [64]. The low success rate could be explained by the existence of several arrhythmogenic foci and the appearance of new ones in a progressive cardiomyopathy. As other groups, we noted that ablation of one focus may unmask a different arrhythmogenic area. Although there are recurrences of VT in up to 60% of the cases were reported, RFA is a therapeutic option for a well selected group of patients.

### 9.4. Surgery

In a few patients, surgical isolation of the right ventricular wall was made to prevent transmission of abnormal rhythms to the left ventricle [65]. However, right ventricular failure was observed in the long-term follow-up of these patients [66]. In 1977, Fontaine et al. reported their experience with ventriculotomy in an attempt to control VT episodes [67], but recurrence of the arrhythmia after surgery was quite frequent. Although cardiac transplant remains as an option for patients with drug refractory heart failure, all the surgical techniques are rarely used.

## 10. Conclusion

ARVD/C is an increasingly reported cause of sudden death, principally in apparently healthy young individuals

and people engaged in competitive sports. The suspicion of the disease in a patient arises from subtle clinical signs, ECG changes and a familial background of sudden death. Non-invasive and invasive tests confirm or rule out the diagnosis in most cases. New therapeutic approaches as ICD implantation help to prevent fatal outcomes in many patients who otherwise have a poor prognosis.

Collaborative studies on this new form of cardiomyopathy are providing more information about its partially unknown etiology, long-term prognosis and efficacy of new therapeutic options.

## Aknowledgements

I thank very much E. Hulse, MD from Children's Mercy Hospital, Kansas City for revising and making suggestions to this manuscript.

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