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# Arrhythmogenic Right Ventricular Cardiomyopathy in the Elderly: A Case Report

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**ABSTRACT**

Arrhythmogenic right ventricular dysplasia also known as arrhythmogenic Right Ventricular Cardiomyopathy is one of the leading causes of arrhythmic cardiac arrest in young people and athletes, but is probably under diagnosed in the older population.The clinical course of arrhythmogenic right ventricular dysplasia is characterized by the occurrence of arrhythmic events, which can cause sudden death, and the impairment of ventricular systolic function, which can lead to death from heart failure. Diagnosis has been facilitated by a set of clinically applicable criteria.The treatment is aimed at reducing the risk of sudden cardiac death properly risk-stratifying patients that would benefit from an implantable cardioverter defibrillator .Antiarrhythmic drugs and catheter ablation are also commonly used to improve quality of life by preventing ventricular arrhythmias, exercise restriction to prevent disease progression.This report presents case of elderly man with newly symptomatic arrhythmogenic right ventricular dysplasia.



## Introduction

Since the first description of arrhythmogenic right ventricular dysplasia (ARVD/C) in 1982, there have been major advances in the diagnosis and management of the disease.

ARVD/C is a heritable heartmuscle disorder that predominantly affects the right ventricle , ARVD/C was once considered a disease of the young, but may be underrecognized in the older population. The diagnosis of ARVD/C should be considered in individuals of all ages.

This article presents the oldest patient with newly diagnosed symptomatic ARVD/C at the ages of 83 presenting with malignant dysrhythmias.

## Case Report

We describe the case of an 83 -year-old man with no cardiovascular risk factors , who consulted for palpitations and weakness , the physical examination has objectified the hemodynamic collapse With tachycardia at 245 beats/min, low blood pressure of 50/32 mmHg, a respiratory rate of 24 breaths/min, profuse sweats and cold extremities. The electrocardiogram (ECG) showed monomorphic ventricular tachycardia “Fig. 1”.The unstable patient was immediately treated with synchronized direct current cardioversion and the administration of intravenous amiodarone

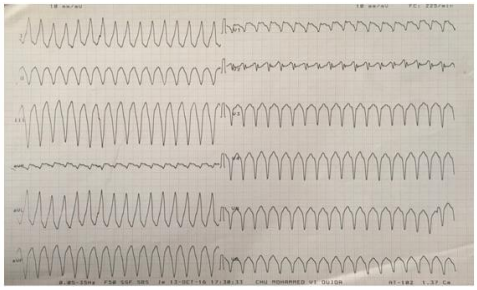

**Figure 1. Electrocardiogram showed monomorphic ventricular tachycardia.**

The cardiac exam revealed an irregular rhythm with a spontaneous turgescence of the jugular veins. The restored rhythm was in atrial fibrillation with a right bundle branch block, left anterior fascicular bloc and negative T-waves in V1 to V5 “Fig. 2”.

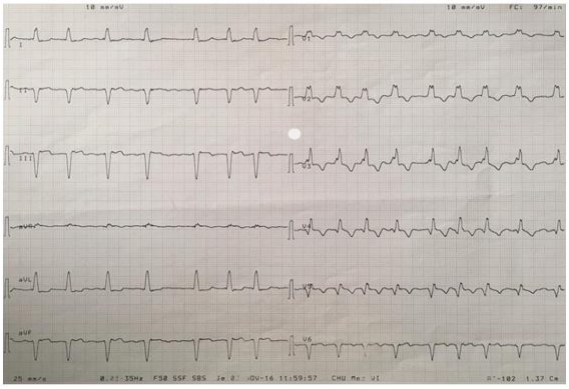

**Figure 2. electrocardiogram after cardioversion: Arial fibrillation with a right bundle branch block, left anterior fascicular bloc and negative T-waves in V1 to V5.**

On admission, the patient’s hemoglobin, white blood cell count, and platelet count were 14,5 g/dL, 11.000/mm3, and 229.000/mm3, respectively. Serum level of potassium was normal at 4.5 mmol/l High-sensitive cardiac troponin test was 109 µg/L(normal value < 26 µg/L).

Laboratory tests also showed normal renal function with urea and creatinine plasmatic levels of 0.44 g/l and 9,76 mg/l, respectively. There were no notable changes in the chest X-Ray.

The echocardiogram showed a preserved function of the left ventricle (LV) , dilated right ventricle (RV) (diastolic diameter of 46 mm), RV dysfunction with tricuspid annular plane systolic excursion (TAPSE) of 9 mm and moderate tricuspid regurgitation, he had an estimated pulmonary artery systolic pressure of 30 mmHg.

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During hospital, Holter monitor detected atrial fibrilation, with 709 premature ventricular beats, and 7 episodes of unsustained VT during 24 hours.

A thoracic CT scan revealed cardiomegaly in the right cavities without any signs of pulmonary embolism. Cardiac magnetic resonance imaging (MRI) was performed showing dilation of the RV with ratio of RV end-diastolic volume to bodysurface area : 115 ml/m² , RV ejection fraction : 40%, Regional RV dyskinesia ( apex and Pulmonary infundibulum) with akinesia of the inferior wall High-intensity areas indicated fatty infiltration of the RV inferior wall and apex "Fig. 3 and 4".

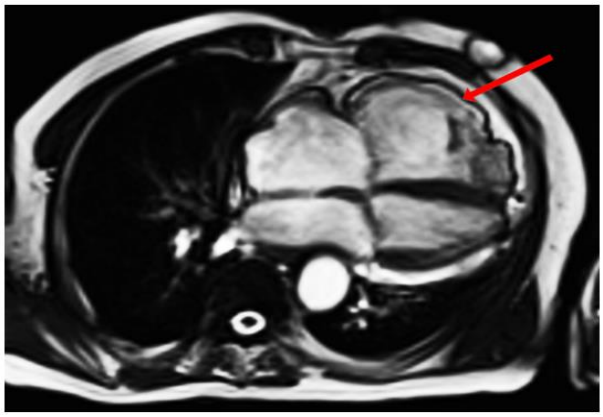

**Figure 3. Delayed contrast-enhanced MRI show delayed enhancement (arrows) confirming presence of fibrosis. Note right ventricular enlargement. True fast imaging with steady-state precession cine reveals localized dyskinesia ( apex and Pulmonary infundibulum ) with akinesia of the inferior wall with markedly diminished ejection fraction.**

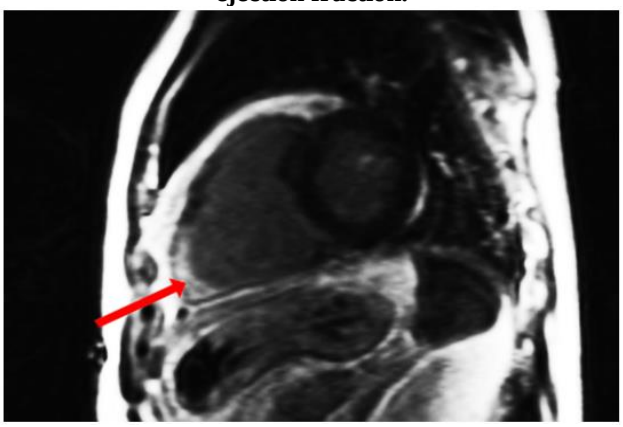

**Figure 4 . Two chamber views arrows indicating a massively dilated right ventricle and fat and fibrotic infiltration.**

The implantable cardioverter defibrillator (ICD) was refused by patient, pharmacological therapies: Betablocker ( Bisprolol) and Amiodarine were well tolerated . Also, oral anticoagulants (acénocoumarol) were used for atrial fibrillation with target International Normalized Ratio (INR) of 2.0 to 3.0.

In subsequent consultations, the evolution appears to be favorable: the patient is asymptomatic with no recurrence, the ECG shows an atrial fibrillation with a heart rate of 70 beats/min, the INR is between 2 and 3 under 3mg of acénocoumarol.

**Discussion**

Right ventricular (RV) dilatation and pathological fibrofatty changes of the RV myocardium leading to dysrhythmias characterize ARVD/C . [1]

In 2002, Daniel More [2] presents the two oldest patients with newly diagnosed symptomatic ARVD/C at the ages of 73 and 74, respectively, presenting with progressive right-sided heart failure and ultimately leading to SCD from malignant dysrhythmias .

ARVD/C is a disease process classically affecting young men and presenting with a wide spectrum of symptoms, including SCD. While the majority of patients affected are under 40 years of age, the syndrome is probably under diagnosed in the older population. This condition should be considered in individuals who present with chest pain, syncope, palpitations, SCD, or signs of chronic progressive heart failure and have evidence of ARVD/C on ECG. [2]

The disease is typically transmitted with an autosomal dominant pattern of inheritance, although rare auto somal recessive forms have been described. [3]

ARVD/C is one of the leading causes of arrhythmic cardiac arrest in young people and athletes. [4] Typically occurs between the second and fourth decades of life. Although the disease was described until the ninth decade . [4-5]

Ferreira [5] report case of the oldest patient ever diagnosed with ARVD/C described in the literature : An-84-year-old woman , who admited for dizziness and unsteadiness , which the diagnosis of ARDV/C was made by autopsy .

The disease is more malignant in men than in women, a finding that can be explained either by a direct influence of sex hormones on the mechanisms involved in the phenotypic expression of the disease or by sex-based differences in the amount or intensity of exercise. [6]

The genetics of ARVD/C support the hypothesis that this is a disease of desmosomal dysfunction. Although the pathogenesis of ARVD/C is not fully understood, it has been hypothesized that abnormalities in the desmosomal proteins may lead to impaired mechanical coupling between individual cells, which then causes myocyte uncoupling resulting in inflammation, fibrosis, and adipocytosis. This is especially true in situations that increase myocardial strain, such as exercise. [7]

ARVD/C is histopathologically characterised by progressive fibrofatty replacement of cardiomyocytes, primarily in the right ventricle. [8]However, histopathologically and functionally the left ventricle ( LV) is affected in many cases and both ventricles are similarly affected by desmosomal and gap junctional protein redistribution. [9]

In the past 25 years, increased insight into the development of the disease as well as the discovery of pathogenic gene mutations involved led to the current understanding that ARVD/C is a genetically determined cardiomyopathy. The molecular genetic substrate for the disease is mainly acknowledged in genes encoding desmosomal adhesion proteins in the intercalated disk . [10]

Specific evaluations are recommended in all patients suspected of ARVD/C : detailed history and family history, physical examination, 12-lead ECG , signal averaged ECG, 24-hour Holter monitoring, maximal exercise testing, two-dimensional echocardiography with quantitative wall motion analysis, and more detailed imaging by cardiac magnetic resonance imaging (MRI) with delayed enhancement analysis. Invasive tests are also useful for diagnostic purposes: RV and LV cineangiography, electrophysiological

testing, and endomyocardial biopsies for histopathological and immunohistochemical analysis. [9-11]

Population studies, autopsy registries, and genetic testing targeted to older patients will help to better characterize this disease and its clinical presentation in the elderly .

Among the elderly population, ARVD/C can have atypical clinical presentation , Symptoms at presentation most commonly include SCD ,chest pain, palpitations, and syncope, but may also include symptoms of heart failure. [12]

The ECG shows abnormalities in 90% of symptomatic patients with ARVD/C. [13]

In 30% of cases, an epsilon wave can be found in the anterior precordial leads. Epsilon waves are potentials of small amplitude following the QRS complex that reflect delayed activation of RV myocytes . [14]

we noted a high incidence (42%) of spontaneous clinical atrial fibrillation( AF) and atrial flutter (aFL) in patients with ARVC/D treated for ventricular tachycardia (VT). These atrial arrhythmias are more common in patients with moderate to severe tricuspid regurgitation and enlarged RV volume and tend to be more common in older patients, suggesting that as RV disease develops progressive dilatation, atrial arrhythmias should be anticipated. [15]

Onset of atrial arrhythmias relative to development of ventricular arrhythmias is significant in 2 ways. First, as indicated by the work by Brembilla-Perrot et al [16]atrial arrhythmias in selected patients may occur sooner in the course of disease and suggest the possibility of direct right atrial involvement with the pathologic process. These early atrial arrhythmias may delay proper diagnosis of ARVC/D.

Second, occurrence of atrial arrhythmias with or after onset of ventricular arrhythmia in the setting of significant RV dilatation and tricuspid valve dysfunction suggests that in most patients the underlying pathogenesis of atrial vulnerability is secondary to the RV disease process.

MRI examinations have a high yield in patients in whom an ARVC/D is suspected since most of their findings represent major diagnostic criteria for this condition. [17]

Cardiac MRI has become the preferred imaging technique because it combines the evaluation of structural and functional ventricular abnormalities with noninvasive tissue characterization with the use of late gadolinium enhancement,which provides information about the presence and amount of fibrofatty myocardial scarring. [18]

The diagnosis of ARVC/D should be considered in individuals of all ages who present with a clinical syndrome consistent with ARVC/D and supportive evidence on ECG . [2]

To standardize the clinical diagnosis of ARVC/D, in 1994 an international task force (TFC ) proposed guidelines in the form of a qualitative scoring system with major and minor criteria. [19]

In 2010, TFC revised the guidelines to improve diagnostic sensitivity, mostly for the clinical screening of family members, by providing quantitative criteria for diagnosing right ventricular abnormalities and adding molecular genetic criteria .

The TFC include six different categories: 1) global and/or regional dysfunction and structural RV alterations, 2) tissue characterisation, 3) depolarisation abnormalities, 4) repolarisation abnormalities, 5) arrhythmias, and 6) family history and genetics. [20]A definite diagnosis is made if a patient has 4 points, with a major criterion equal to 2 points and a minor criterion equal to 1 point. A borderline diagnosis is made with 3 points and a possible diagnosis with 2 points.

In the case of our patient, the diagnosis of ARVC/D was retained in front of the presence of :

1 major criteria:

Regional RV akinesia and dyskinesia with ratio of RV end-diastolic volume to bodysurface area $\geq$110 ml /m² and RV ejection fraction $\leq$40% .

2 minor criteria:

- Inverted T waves in leads V1, V2, V3, and V4 (in the presence of complete right bundle-branch block).
- >500 ventricular extrasystoles per 24 hr on Holter monitoring .

However, the diagnosis remains problematic because of the low specificity of electrocardiographic abnormalities, multiple causes of right ventricular arrhythmias, difficulties in the use of imaging to assess right ventricular structure and function, and the sometimes puzzling results of genetic testing. [4]

The differential diagnosis for ARVD/C should include cardiac sarcoidosis, idiopathic right ventricular outflow tract ventricular tachycardia, and VT arising from the aortic root. For patients that present with symptoms of RV systolic heart failure, RV infarct or pulmonary hypertension should be considered. Dilated cardiomyopathy should also be kept in mind when there is evidence of biventricular heart failure, as well as biventricular arrhythmogenic cardiomyopathy when there is significant ventricular ectopy . [21]

Current therapeutic approaches to ARVD/C are palliative and partially alleviate symptoms and the risk of sudden cardiac death but do not prevent the development or progression of the disease process. A definitive curative treatment will require a deeper knowledge of the biologic mechanisms and environmental factors involved in the pathogenesis of ARVD/C. [4]

Few studies have been done to establish the efficacy and safety of pharmacologic therapy in ARVD/C ; beta-blocker therapy is recommended in ARVD/C patients with recurrent VT, appropriate ICD therapies, or inappropriate ICD interventions resulting from sinus tachycardia, supraventricular tachycardia, or atrial fibrillation/flutter with high ventricular rate (class I). [22]

For antiarrhythmic drugs , the goal is to improve quality of life by preventing symptomatic ventricular arrhythmias and ICD discharges.there are no randomized control trials to compare the different antiarrhythmic drugs available, but amiodarone and sotalol are commonly used. [7]

Therapy with oral anticoagulants is reserved for patients with atrial fibrillation or thromboembolic complications.

The role of catheter ablation in patients with ARVD/C is not curative but improves quality of life by decreasing the frequency of episodes of VT.The current recommendations for catheter ablation are for patients with incessant VT or frequent appropriate ICD therapies despite maximal pharmacological therapy.

An epicardial approach is for those who fail one or more attempts of endocardial VT ablation (class I) . [22]

Papaioannou et al. present an unusual case of late presentation of ARVD/C in a 72-year-old white Caucasian man. His post-cardioversion electrocardiogram, cardiac echocardiogram, coronary angiogram, magnetic resonance imaging and electrophysiological study confirmed the diagnosis of ARVD/C . The patient was treated with an

implantable cardioverter defibrillator and discharged on sotalol. [23]

One of the most important tools in preventing disease progression is restriction of exercise .[7]

The ARVD/C treatment consensus statement, published by Corrado et al. concluded that patients with a definite diagnosis of ARVD/C should not participate in competitive or endurance sports (class I) with the possible exception of recreational low-intensity sports (class IIa). Restriction from competitive sports may be considered in ARVD/C family members that are healthy gene carriers (class IIa) and those that have an unknown genotype. [22]

Cardiac transplantation is reserved for patients that have intractable arrhythmias and/or heart failure. Patients who received a transplant had a more prolonged course of the disease and a relatively early onset. [22]

Retrospective analysis of clinical and pathological studies identified several risk factors for sudden death or appropriate ICD therapy, such as previously aborted SCD, syncope, young age, severe RV dysfunction, and LV involvement. Patients without VT had the best prognosis . [24-25]

This case report demonstrates that ARVD/C may have a late presentation and this diagnosis should be considered as a one of principal cause of malignant dysrhythmias of right ventricular origin among the elderly.

**No conflict of interest**

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