*Ulster Med J* 2019;**88(1):**15-16

*Case Report*

# Fifty Years of Ventricular Tachycardia in a Single Patient

Stephen Adams, Alastair Gray, Niall Herity

Accepted: 22nd August 2018
Provenance: externally peer reviewed

**Key Words:** Arrhythmogenic Right Ventricular Cardiomyopathy, Dysplasia, Ventricular, Tachycardia

**Abstract** We report a patient who first presented during childhood in the early 1960's with several episodes of ventricular tachycardia (VT) and we describe her management which reflected the best medical knowledge at the time. She then presented more than 50 years later, again with VT, at which time a definitive diagnosis of the underlying cause was made. Her case illustrates the evolution in the understanding and management of VT over the past 50 years. This in turn reflects the clinical and technological advances in the management of cardiovascular disease over time.

## CASE PRESENTATION

A 7-year old patient presented with symptoms of palpitations in April 1961. The medical notes at the time described symptoms coming on while dancing or during physical exertion at school. There was no loss of consciousness. An electrocardiogram (ECG) showed ventricular tachycardia (VT) (Figure 1). An ECG during sinus rhythm was reported to show right ventricular hypertrophy with strain (Figure 2). The conclusion at the time was that the episodes of VT were triggered by sympathetic over-excitement and she was commenced on oral quinidine and propranolol. In July 1969 a chest X-ray report stated that '…prominence has developed in the left heart border…could be consistent with myopathy'. In 1983, an echocardiogram demonstrated normal biventricular structure and function.

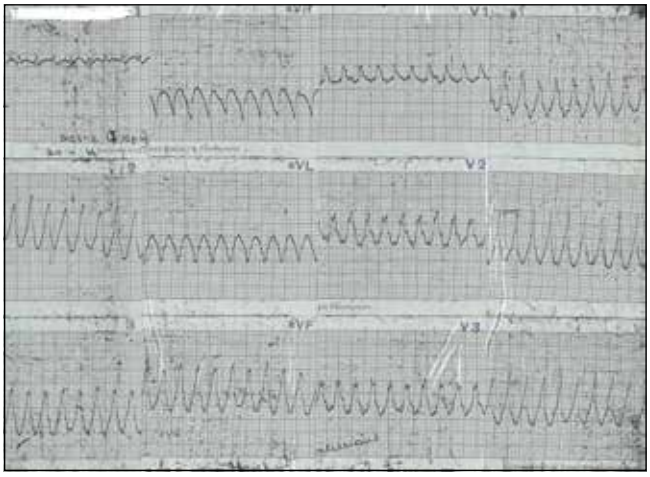

*Fig 1*. Electrocardiogram showing ventricular tachycardia

She remained under regular cardiology review and over a 10-year period, presented 15 times with symptomatic VT. Three episodes required DC cardioversion for haemodynamic instability; intravenous lignocaine was used on one occasion and spontaneous reversion to normal sinus rhythm occurred on other occasions.

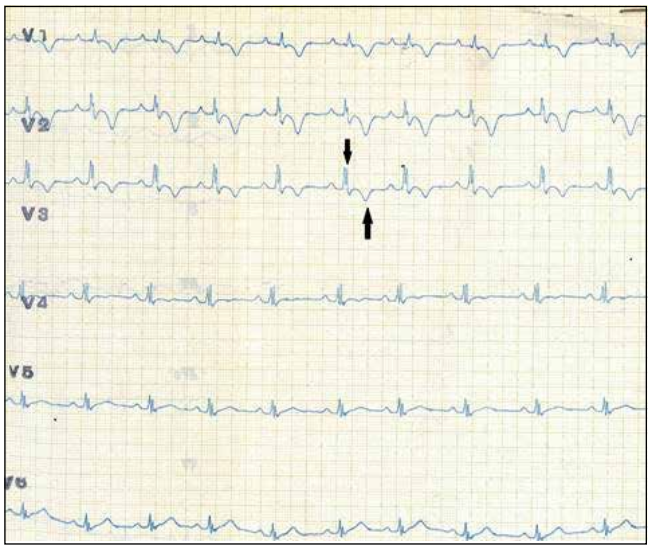


*Fig 2*. Electrocardiogram reported as showing right ventricular hypertrophy with strain pattern (arrows).

During the 1970's she came off her medication but remained asymptomatic until 2018, when she presented to the emergency department in VT, 57 years after her first admission (Figure 3). Following DC cardioversion for haemodynamic compromise, a 12-lead ECG showed right bundle branch block, anterior T-wave inversion, and an epsilon wave (Figure 4), features typical of arrhythmogenic right ventricular cardiomyopathy (ARVC).

An echocardiogram confirmed severe right ventricular dysfunction and aneurysmal dilatation and cardiac magnetic resonance imaging confirmed classic features of ARVC (Fig. 5).

A cardioverter defibrillator was implanted and she was referred to the Inherited Cardiac Conditions clinic for follow up and genetic screening. She has remained well since

Cardiology Department, Belfast HSC Trust
Correspondence to: Dr Stephen Adams
E-mail: stephenadams2301@gmail.com

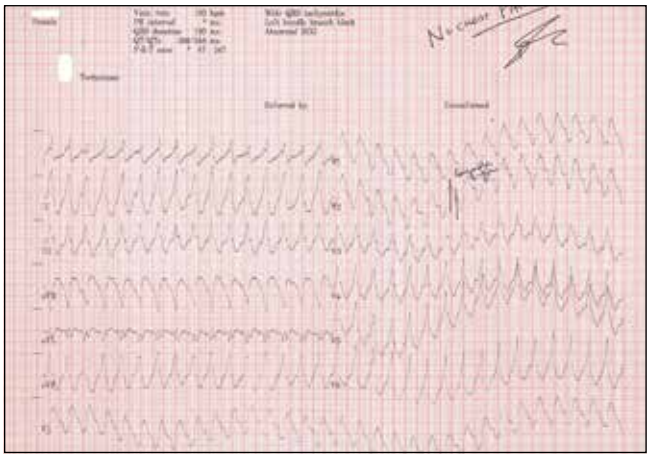

*Fig 3*. Electrocardiogram form 2018 showing ventricular tachycardia.

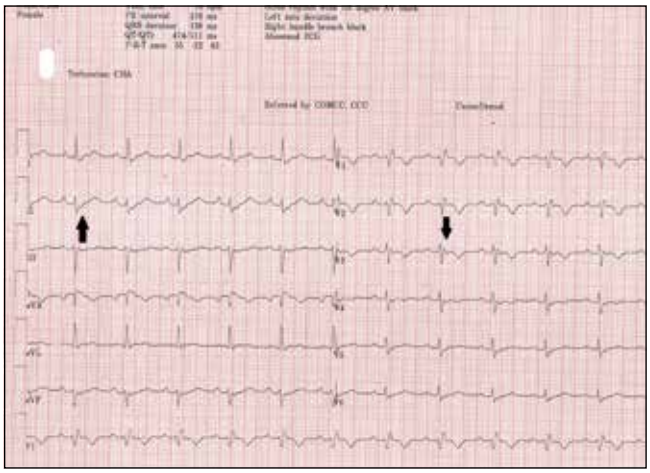

*Fig 4*. 12-lead electrocardiogram showing right bundle branch block, anterior T-wave inversion, and an epsilon wave (arrows).

## DISCUSSION

Arrhythmogenic right ventricular cardiomyopathy (ARVC) (also known as arrhythmogenic right ventricular dysplasia [ARVD]) is an autosomal dominant inherited cardiac disorder. It demonstrates incomplete penetrance with variable expression and is thought to be the result of a desmosomal abnormality arising from mutations in genes encoding desmoplakin and plakoglobin. [1] Characterised by fibro-fatty replacement of the ventricular myocardium, it predominantly affects the right ventricle but can also involve the left ventricle (LV) and rarely LV involvement can occur exclusively. [2,3]

Clinical presentation arises due to ventricular dysfunction and arrhythmia. Although symptoms may be non-specific they frequently include dyspnoea and palpitations. Arrhythmia or sudden cardiac death may be the first presenting feature. Diagnosis is challenging and may require several years of follow up and serial imaging investigations. A diagnostic task force was established in 1994 outlining diagnostic criteria which were updated in 2010. [4]

When this patient first presented with VT in 1966, ARVC had not been described formally and did not appear in cardiology textbooks. Of interest, in 1736, there had been descriptions of an Italian family who experienced right ventricular failure and cardiac death in four successive generations. It is reasonable to assume that the underlying cause was ARVC. The first modern description was in 1982 involving 24 adults with VT of left bundle branch block morphology. [5] In 1984 additional ECG findings including the epsilon wave were first recognised. [6]

This patient's clinical journey is a metaphor for the remarkable evolution in our understanding of VT and ARVC over the past 50 years. In particular it exemplifies how advances in cardiac imaging, electrophysiology and implantable device therapy, genetic analysis and the global and immediate availability of scientific information has transformed the diagnosis, management and prognosis of patients with complex cardiovascular diseases.

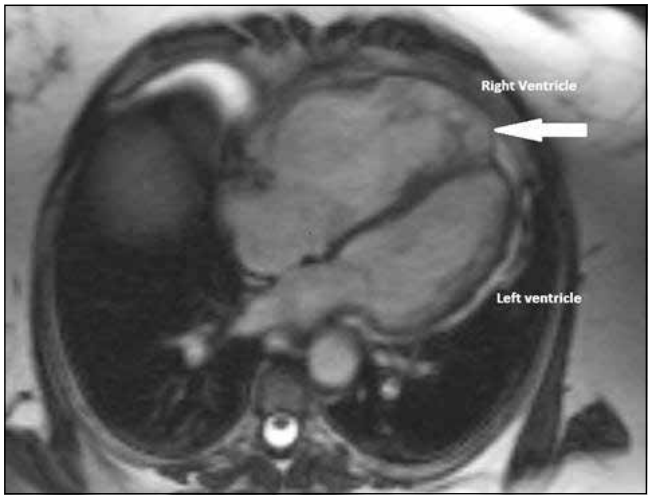


*Fig 5*. Cardiac magnetic resonance imaging showing a severely dilated right ventricle with multiple areas of wall thinning. There is aneurysm formation within the apical region (arrow). In contrast, the left ventricle is undilated.

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