Heart, Lung and Circulation (2016) **25,** 275–281
1443-9506/04/$36.00
http://dx.doi.org/10.1016/j.hlc.2015.08.019

ORIGINAL ARTICLE

High Arrhythmic Burden but Low Mortality during Long-term Follow-up in Arrhythmogenic Right Ventricular Cardiomyopathy

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Received 5 June 2015; accepted 27 August 2015; online published-ahead-of-print 28 September 2015

Background Arrhythmogenic right ventricular cardiomyopathy (ARVC) is associated with a high incidence of ventricular tachyarrhythmia and sudden death. The mainstay of management is the implantable cardioverter defibrillator (ICD). A small number of patient cohorts have generated a large number of reports.

Methods Prospective registry data supplemented with clinical and ICD records of 30 patients with ARVC fulfilling the 2010 modified Task Force Criteria. This cohort has not been reported on previously.

Results Median age at diagnosis: 46yrs (range 21-68); 20 (80%) male; six (19%) Maori. Duration of follow-up: 7.4yrs (range 1.7-23). Implantable cardioverter defibrillator implantation in 26; three (12%) for resuscitated sudden cardiac death; 17 (65%) for symptomatic ventricular tachyarrhythmia; three (12%) for syncope; and three (12%) for family history of sudden death attributable to ARVC. Two patients died during follow-up, one had an ICD, though died of a carcinoma. Thirteen (50%) experienced appropriate ICD therapy with median time to therapy 12 months, and four (15%) experienced inappropriate shock therapy. Male gender was an independent predictor of appropriate ICD therapy (HR 1.6, 95% CI 1.5-2.7, P=0.01).

Conclusions The long-term prognosis of patients with ARVC is favourable although high proportions receive appropriate ICD therapy. Male gender is an independent predictor of appropriate ICD therapy.

Keywords Arrhythmogenic right ventricular cardiomyopathy/dysplasia • Cardiomyopathy • Death • Sudden • Implantable cardioverter defibrillators • Electrophysiology

Introduction

Arrhythmogenic right ventricular cardiomyopathy/dysplasia (ARVC) is an inheritable heart muscle disease with a natural history that is associated with ventricular tachyarrhythmias and sudden cardiac death (SCD). Histologically it is characterised by fibro fatty substitution of the right ventricular (RV) myocardium, and in addition to ventricular tachyarrhythmia it results in mechanical RV dysfunction [1–4].

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Observational studies have demonstrated the efficacy of the implantable cardioverter defibrillator (ICD) in preventing SCD in patients with ARVC [5–10] and have resulted in guideline recommendations that these be implanted in those who have survived a cardiac arrest, and for the primary prevention of SCD in high risk individuals [11]. Additionally, several studies have reported that patients with ARVC who undergo ICD placement have a high probability of receiving an appropriate ICD therapy for treatment of a sustained ventricular arrhythmia [5,8,12–14]. These cohorts report on data that has median follow-up durations from 3.3 to 6.7 years, with little information on the occurrence of rapid ventricular tachycardia/fibrillation (VT/VF) and overall prognosis [5,7,14]. At present there is a paucity of prospective long-term data to provide definite guideline recommendations for primary prevention. Furthermore the subjects in these reports reside in Europe and North America and have been described on several occasions, in both single centre and multicentre reports. While these reports have been used to develop diagnostic criteria for ARVC and indications and outcome for the use of ICDs, these data may not be applicable in other regions of the world [3,5,7,8,12,15,16]. Local data from Australasia has not previously been reported, and it is important to compare and contrast these with that from Europe and North America, particularly with respect to VT/VF and overall prognosis.

Table 1 2010 Revised Taskforce Criteria for the diagnosis of arrhythmogenic right ventricular cardiomyopathy

	Major criteria	Minor Criteria
Global or regional dysfunction and structural alterations	**Echocardiography:** Regional RV akinesia, dyskinesia, or aneurysm, and 1 of (at end diastole): PLAX RVOT >32 mm, PSAX RVOT >36 mm, or fractional area change <33%. **Cardiac MRI:** Regional RV akinesia or dyskinesia or dyssynchronous RV contraction, and 1 of the following: RVEDV >110mL/m^2 (male) >100mL/m^2 (female), or RVEF <40%. **RV angiography:** Regional RV akinesia, dyskinesia, or aneurysm.	**Echocardiography:** Regional RV akinesia or dyskinesia, and 1 of (at end diastole): PLAX RVOT 29-32 mm, PSAX RVOT 32-36 mm, or fractional area change 33-40%. **Cardiac MRI:** Regional RV akinesia or dyskinesia or dyssynchronous RV contraction, and 1 of the following: RVEDV 100-110mL/m^2 (male) 90-100mL/m^2 (female), or RVEF 40-45%.
Tissue characterisation of the wall	Residual myocytes <60% by morphometric analysis, with fibrous replacement of the RV free wall myocardium in >1 sample, with or without fatty replacement of tissue on endomyocardial biopsy.	Residual myocytes 60-75% by morphometric analysis, with fibrous replacement of the RV free wall myocardium in >1 sample, with or without fatty replacement of tissue on endomyocardial biopsy.
Repolarisation abnormalities	Inverted T waves in right precordial leads (V_{1-3}) or beyond in those >14 years of age in the absence of a complete RBBB.	Inverted T waves in leads V_{1-2} in those aged >14 years (in the absence of a complete RBBB) or in V_{4-6}. Inverted T waves in leads V_{1-4} in those aged >14 years in the presence of a complete RBBB.
Depolarisation abnormalities	Epsilon wave in the right precordial leads V_{1-3}.	Late potentials by SAECG in >1 of 3 parameters in the absence of a QRS duration of >110ms on the standard ECG.
Arrhythmias	Ventricular tachycardia of left bundle-branch morphology with superior axis.	Ventricular tachycardia of left bundle-branch morphology with inferior axis. >500 ventricular extrasystoles per 24 hours (Holter).
Family history	ARVC confirmed in a first-degree relative. ARVC confirmed pathologically at autopsy or surgery in a first-degree relative. Identification of a pathogenic mutation categorised as associated or probably associated with ARVC in the patient under evaluation.	History of ARVC in a first-degree relative in whom it is not possible or practical to determine whether the family member meets current Task Force criteria. Premature sudden death (<35 years of age) due to suspected ARVC in a first-degree relative. ARVC confirmed in a second-degree relative.

A definite diagnosis of ARVC is made when 2 Major or 1 Major and 2 Minor or 4 Minor criteria are present [22].

ARVC – Arrhythmogenic right ventricular cardiomyopathy; MRI – Magnetic resonance imaging; PLAX – Parasternal long axis view; PSAX – Parasternal short axis view; RBBB – Right bundle branch block; RV – Right ventricle; RVEDV – Right ventricular end diastolic volume; RVEF – Right ventricular ejection fraction; RVOT – Right ventricular outflow tract; SAECG – Signal averaged ECG.

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