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Right ventricular dilated cardiomyopathy

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SUMMARY Fourteen patients with predominantly right sided dilated cardiomyopathy were studied, of whom five died suddenly. The condition is characterised by male preponderance, syncope, ventricular tachycardia, which typically has a left bundle branch block pattern on the surface electrocardiogram, and right heart failure.

The diagnosis should be considered in patients presenting with otherwise unexplained ventricular tachycardia or syncope; the diagnosis may be readily missed because of the nonspecific nature or absence of signs.

Dilated cardiomyopathy is a heart muscle disease of unknown cause,[1] which is recognised by ventricular dilatation and a low ejection fraction. In most patients the left ventricle is the more severely affected, and pathological abnormalities usually also predominate in the left heart.[2] We report fourteen cases of severe right ventricular dilatation of unknown cause, in which there was preservation of left ventricular function but which probably represent one end of the spectrum of dilated cardiomyopathy. Their clinical presentation was either with right heart failure or with arrhythmias and syncope, which brought the patient to medical attention when other symptoms were still absent.

Case reports

CLINICAL DATA

Fourteen cases are reported in which severe right ventricular dysfunction was the hallmark of the disease (Table 1). Five patients were female and nine were male, the mean age being 28 (range 9–62) years at initial presentation. Initial symptoms reflected arrhythmia in 10 (70%) patients; four patients had heart failure from the outset and three later. Ventricular tachycardia caused syncope in six patients, whereas supraventricular arrhythmia resulted in palpitation in four. Eight patients had no clinical signs of right ventricular failure.

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To date, 12 patients have been observed for a mean of 4·1 years (range 6 months to 14 years). The condition of one patient (case 2) improved initially, but she then died suddenly, and one (case 5) was a Madagascan seaman who was not seen again. Eleven of the 12 patients available for follow up died between six months and 12 years after the onset of their symptoms. Five patients died suddenly, but only two were in heart failure at that time. Two of the patients were siblings whose presentations and clinical findings were almost identical: both presented with syncope due to ventricular tachycardia and died suddenly two and three years later.

All patients had either a third or fourth heart sound as well as right ventricular impulse. The jugular venous pressure was raised only in the seven patients who developed right ventricular failure, and one patient (case 1) developed progressive tricuspid regurgitation during the 12 years of follow up.

ELECTROCARDIOGRAPHIC FINDINGS

In nine patients the electrocardiograms suggested a right ventricular abnormality (Table 2, Fig.1). Right bundle branch block was present in two patients, right ventricular hypertrophy in two, an abnormal mean frontal axis greater than +120° in two, and T wave inversion in leads II, III, aVF, and V1–V3 in three.

Arrhythmias

Arrhythmias were important in the clinical course of the disease in 12 patients. Supraventricular arrhyth-

Table 1 *Clinical data on 14 patients with right ventricular dilated cardiomyopathy*

Case No	*Age at presentation (yr)*	*Sex*	*Presenting symptoms*	*Onset of heart failure (yr)*	*Progress*	*Follow-up (yr)*	*Died*	*Comments*
1	37	F	Palpitation	2	Severe right heart failure	12	Yes	Developed primary biliary cirrhosis
2	16	F	Oedema, ascites	0	Initial improvement. Sudden death	6 m	Yes	
3	62	F	Palpitation	1	Recurrent right heart failure	8	Yes	
4	49	F	Palpitation	1-6 m	Sick sinus syndrome. Pacemaker. Cerebrovascular episode	2		Family history
5	49	M	Oedema	*	No follow up	—		
6	28	M	Chest pain, palpitation	†	Recurrent palpitation	3		
7	12	F	Ascites, oedema	*	Recurrent right heart failure	1		
8	16	M	Syncope	†	No further symptoms	1		
9	14	M	Syncope	†	Further syncope before sudden death	2	Yes	
10	14	M	Syncope	†	No further syncope. Sudden death	3	Yes	Brother of case 9
11	21	M	Syncope	†	Recurrent palpitation	6 m		
12	9	M	Syncope	†	Control with disopyramide	10		
13	20	M	Dizzy spells	†	Ventricular tachycardia controlled with amiodarone	1		
14	21	M	Syncope	†	Sudden death	6 m	Yes	

*No evidence of clinical right ventricular failure.
†Heart failure at time of presentation.

Table 2 *Electrocardiographic findings*

Case No	*Cardiac rhythm*	*QRST electrocardiogram*
1	SVT	Left axis deviation
2	SR, AF, SVT	Low voltage QRST, widespread repolarisation abnormalities
3	AF	RBBB
4	AF, A flutter, SVT, slow junctional rhythm	Normal QRS, widespread T abnormalities
5	SR	RBBB
6	SR, A flutter, VT	Right axis deviation
7	SR, multifocal VES	Low voltage QRS, intraventricular conduction delay
8	SR, VT, VF	Right axis deviation, T inversion V1–4
9	Recurrent VT	RVH right axis deviation
10	Recurrent VT, multifocal VES	Right axis, RVH
11	SR, VES, VT	T inversion (leads II, III, aVF, V1–4)
12	SR, VT, VF	T inversion (leads II, III, aVF, V1–4)
13	SR, recurrent VT	Low voltage, Rs V1, T inversion V1–4
14	SR, VT	T inversion (leads II, III, aVF, V1–4)

SVT, Supraventricular tachycardia; VES, ventricular extrasystoles; SR, sinus rhythm; VT, ventricular tachycardia; AF, atrial fibrillation; RBBB, right bundle branch block; RVH, right ventricular hypertrophy.

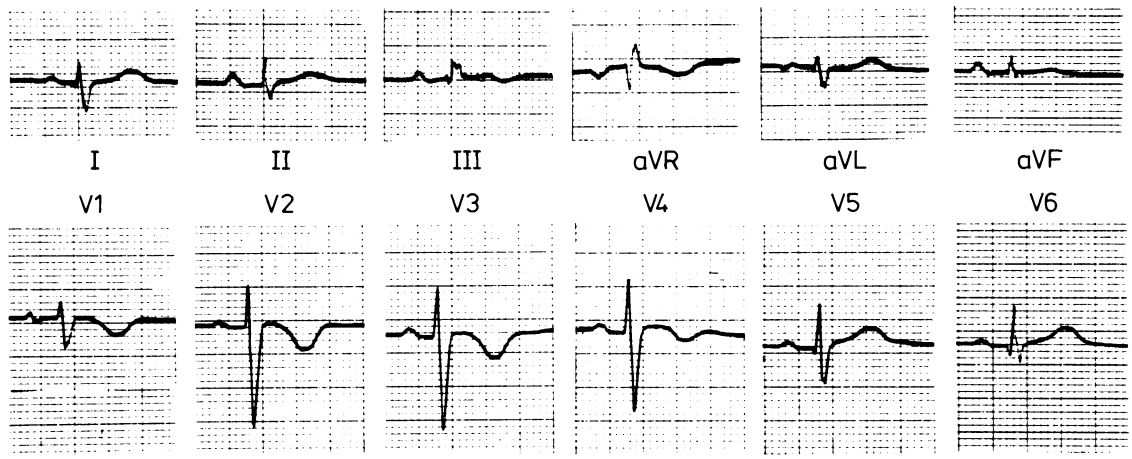

Fig. 1 *Typical electrocardiogram from a patient with right ventricular dilated cardiomyopathy.*

mias occurred in five patients (paroxysmal tachycardia in two, atrial fibrillation in three, and atrial flutter in two). Two patients had evidence of sinoatrial disease with changing supraventricular arrhythmias, one of whom required a permanent pacemaker because of severe heart failure associated with a slow junctional rhythm. In three patients (cases 1, 3, and 4) arrhythmias preceded the onset of heart failure by up to two years. Ventricular arrhythmias occurred in eight patients (ventricular tachycardia (four) and frequent ventricular premature beats (four)). In five of the patients with ventricular arrhythmias, syncope was the presenting symptom. Despite apparent suppression of the arrhythmias the two siblings (cases 9 and 10) died suddenly. Four of the patients with ventricular tachycardia had frequent multifocal ventricular extrasystoles that were only partially suppressed by quinidine, mexiletine, or propranolol. Amiodarone was successful in suppressing ventricular arrhythmias in three patients seen at a later date.

In two patients (cases 12 and 13) intracardiac recordings were obtained. Case 12 was a 19 year old man who had had episodes of palpitation associated with faintness and sweating since the age of 9 years. The resting electrocardiogram showed T wave inversion in leads II, III, aVF, and V1–V4. Both the echocardiogram and angiogram showed extreme right ventricular dilatation but a normal left ventricle. Electrophysiological investigation provoked a paroxysmal re-entry atrioventricular tachycardia which could be induced by incremental ventricular and atrial overdrive pacing. The tachycardia was terminated by either ventricular or atrial overdrive pacing, His bundle extrastimuli, or intravenous verapamil. A short spontaneous episode of ventricular tachycardia with left bundle branch block configuration could not be reproduced by ventricular extrastimuli. Several days later the ventricular tachycardia recurred spontaneously. It was shown to originate in the right ventricular outflow tract, could be provoked by programmed electrical stimulation, and could be stopped by ventricular overdrive pacing. During programmed ventricular extrastimuli a morphologically different tachycardia was initiated which was shown to originate low in the right ventricle. The ventricular extrasystoles were suppressed by disopyramide and to date the patient remains asymptomatic.

Case 13 was a 20 year old student of athletic habits who had had episodes of rapid regular palpitation associated with dizziness and chest discomfort precipitated by exertion. The resting electrocardiogram showed multifocal ventricular extrasystoles and short runs of ventricular tachycardia as well as T wave inversion in leads II, III, aVF, and V1–V4. Echocardiography and ventricular angiography showed a dilated poorly contracting right ventricle and a normal left ventricle. The intracardiac pressures and the cardiac output were normal An electrophysiological study showed ventricular tachycardia originating in the right ventricular outflow tract that could be initiated and terminated with ventricular extrastimuli. On one occasion multifocal ventricular tachycardia developed and led to ventricular fibrillation, which was successfully cardioverted. The study was repeated (on two occasions) while the patient was taking amiodarone. On these occasions the arrhythmia could still be provoked but did not degenerate and reverted spontaneously. He remained well and free from arrhythmia six months later.

ECHOCARDIOGRAPHIC DATA

In nine patients M mode echocardiograms were recorded, all of which showed a dilated right ventricle, paradoxical interventricular septal motion suggesting volume overload of the right ventricle, and a normal left ventricular chamber (Table 3, Fig. 2).

ANGIOGRAPHIC DATA

Thirteen patients underwent right ventricular contrast angiography (Table 3). This showed extreme dilatation of the chamber, which appeared to compress the small left atrium and ventricle posteriorly against the spine. The right ventricle contracted poorly, and there was neither localised dyskinesia nor a discrete aneurysm. The tricuspid valve was normally located in all patients. In five patients there was severe tricuspid regurgitation into a dilated right atrium. In two patients (cases 6 and 13) the left ventricle was slightly dilated.

HAEMODYNAMIC DATA

The right atrial and right ventricular diastolic pressures were raised in seven of 13 patients (Table 3). In two patients, the pulmonary arterial systolic pressure was above 40 mm Hg. Only in one patient (case 2) was the left ventricular end diastolic pressure substantially raised (25 mm Hg). The other patients had left ventricular pressures which were either at or below 16 mm Hg.

NECROPSY DATA

There was pronounced cardiac enlargement and hypertrophy in the three hearts examined at necropsy (average weight 440 g). The two brothers (cases 9 and 10) had similar findings with right ventricular dilatation and grossly abnormal left ventricles. The myocardium appeared pale with areas of endocardial fibrosis conspicuous in the right ventricle. Histological examination showed numerous foci of fibrous replacement involving both ventricles. In case 10 there was extensive muscle cell necrosis and lymphocytic infiltration which was not present in the

Table 3 *Echocardiographic, angiographic, and haemodynamic data*

Case No	*Echocardiography*	*Angiography*			*Pressures (mm Hg)*			*Cardiac index*
		RA	*RV*	*LV*	*RA*	*RV*	*LV*	
1	—	+	+	N	15	42/13	124/7	
2	Dilated RV	++	+++	N	24	28/24	120/25	
3	Dilated RV, IVS paradoxical	+	++	N	18	30/8	130/14	
4	Dilated RV	+	++	I		48/12	100/16	2·1
5		+++	++	N	14	40/20	10(PCWP)	
6			++	I				
7	Dilated RV, IVS paradoxical		++	N	16	35/18	100/14	
8	Dilated RV, IVS paradoxical	+	++	N	2	26/2	100/10	
9			+++	N	14	26/13	110/14	2·7
10	Dilated RV, normal LV							
11	Dilated RV, normal LV		++	N	4	25/6	120/3	3·0
12	Dilated RV, normal LV		++	N	4	20/5	110/7	2·5
13	Grossly dilated RV, normal LV, IVS paradoxical		++	I	7	24/8	110/15	1·7
14			+	N	3	22/4	120/7	

+, ++, and +++, degree of dilatation; RA, right atrium; RV, right ventricle; LV, left ventricle; N, normal function; I, mild impairment of function; PCWP, pulmonary capillary wedge pressure; IVS, interventricular septum.

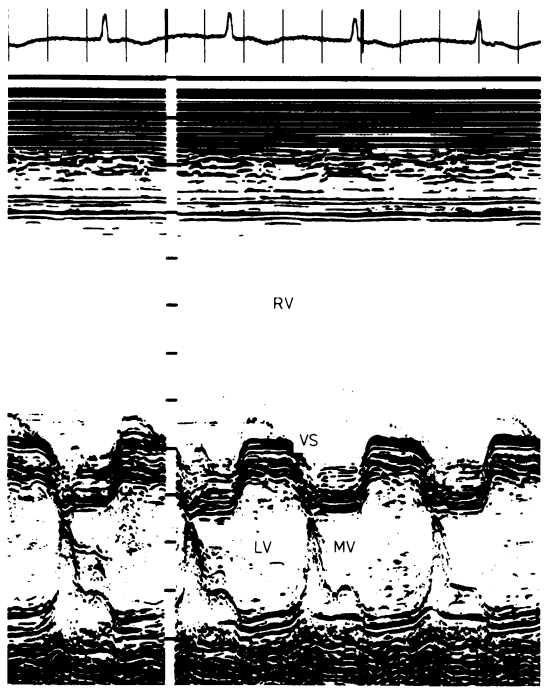

Fig. 2 *M mode echocardiogram showing a dilated right ventricle with normal left ventricular dimensions. RV, right ventricular cavity; LV, left ventricular cavity; VS, ventricular septum; MV, mitral valve.*

heart of case 9.

One patient (case 1) died 11 years after she presented with signs of right heart failure. At necropsy the right atrium and ventricle were extremely dilated. Although angiography eight years previously had shown a normal left ventricle, at the time of necropsy the left chamber was moderately dilated. Histological examination showed hypertrophied myocardial cells, an extensive acute inflammatory cellular infiltrate, but no muscle fibre necrosis.

Discussion

All fourteen patients reported in this study had severe right ventricular dilatation with relatively good left ventricular function. In most patients with dilated cardiomyopathy the clinical and pathological abnormalities are more pronounced in the left ventricle than in the right.[2] It is likely that the patients in the present study represented one of the spectrum of clinical and pathological features of dilated cardiomyopathy. Selective involvement of the right ventricle has been noted, albeit rarely, by several authors.[3-5] Right heart failure is most often a consequence of left ventricular dysfunction; however, when there is a diffuse myocardial disease right ventricular failure might in certain circumstances develop either before or after left heart failure. We believe that patients with right ventricular dilated cardiomyopathy have diffuse myocardial disease, but for reasons as yet undetermined the right heart failed first.

A variety of acquired or congenital abnormalities of the right heart—including atrial septal defect, partial anomalous pulmonary venous drainage, isolated tricuspid chordal rupture, congenital pulmonary regurgitation, and Ebstein's anomaly—may cause isolated right ventricular failure. Many of these conditions can be readily excluded by clinical investigation. The use of cross sectional echocardiography has greatly facilitated the diagnosis of Ebstein's anomaly. Three other conditions that cause selective right ventricular dysfunction have been described. Classically,

Uhl's anomaly[6] results in cyanosis and heart failure in infancy[7]; the electrocardiogram usually shows a lack of right ventricular forces[8] and pathologically there is a complete absence of right ventricular myocardium. The condition has, however, been reported in adults, and ventricular tachycardia of right ventricular origin may be a complication.[8] The right ventricle has normal wall thickness, the trabeculated and apical portions are almost completely absent, and the myocardium is histologically normal. Arrhythmia may occur. Marcus *et al.* reported 24 patients with right ventricular dysplasia who presented with ventricular or supraventricular tachycardia, right heart failure, or asymptomatic cardiomegaly.[9] Angiographic evidence of right ventricular enlargement was usually, though not invariably, present. Right ventricular angiography appeared to show regional wall motion abnormalities, and at necropsy apparently discrete aneurysmal dilatation was noted in the pulmonary infundibulum, apex, and inferior wall of the right ventricle. Microscopical examination showed infiltration of the interstitial tissue and hypertrophy or degeneration of the remaining myofibrils. Almost certainly there is some overlap between these various conditions, but the semantics of the diagnostic label are irrelevant. The angiographic and histological data from the patients in the present study do not support a diagnosis of right ventricular dysplasia. One patient (case 2) had a very thin walled right ventricle. The low voltage QRS complexes and the pressure traces from the right atrium, ventricle, pulmonary artery, and left ventricle suggested pericardial tamponade. Angiography showed a huge right atrium and a non-contractile right ventricle. Although the angiographic appearances might suggest right ventricular aplasia, the sudden onset of severe right heart failure in a previously asymptomatic young adult makes Uhl's anomaly unlikely.

Arrhythmias were important in the clinical course of most patients with right ventricular dilated cardiomyopathy. The paucity of prospective data on the incidence of ventricular arrhythmia and sudden death in a population of patients with the usual form of dilated cardiomyopathy precludes a statement on the relative incidences of arrhythmia in the two conditions. Ventricular tachycardia was shown to originate in the right ventricles of the two patients who underwent electrophysiological studies. Degeneration of the arrhythmia to ventricular fibrillation in one of these patients indicates the potentially unstable nature of the arrhythmia in myopathic hearts. Of the 10 patients with right ventricular dilated cardiomyopathy who presented with arrhythmias, four subsequently died suddenly despite apparent control of their ventricular arrhythmias.

Ambulatory electrocardiographic monitoring and electrophysiological study are the best methods of confirming control of arrhythmias. The more recent cases of right ventricular dilated cardiomyopathy had arrhythmias that were shown to be effectively suppressed using these methods. With successful control of arrhythmias using drugs such as amiodarone the prognosis of this high risk group of patients may possibly be improved. Whether or not vasodilator drugs will prevent progression of the cardiomyopathy is unknown. In many cases symptomatic arrhythmias may lead to an earlier presentation of cardiomyopathy in fit people who would not otherwise have sought medical attention.

The possibility of right ventricular dilated cardiomyopathy should be considered in any young patient presenting with syncope or ventricular tachycardia. The assessment of right ventricular function has previously been difficult, but recently a detailed assessment of right heart function has been possible using radionuclide and cross sectional echocardiographic techniques.[10]

References

1 Report of the WHO/ISFC task force on the definition and classification of cardiomyopathies. *Br Heart J* 1980; **44:** 672–3.
2 Olsen EG. Pathology of primary cardiomyopathies. *Postgrad Med J* 1972; **48:** 732–7.
3 Hamby RI. Primary myocardial disease. *Medicine (Baltimore)* 1970; **49:** 55–78.
4 Kreulen TH, Gorlin R, Herman MV. Ventriculographic patterns and hemodynamics in primary myocardial disease. *Circulation* 1973; **47:** 299–308.
5 Hatle L, Stake G, Storstein O. Chronic myocardial disease. II. Haemodynamic findings related to long-term prognosis. *Acta Med Scand* 1976; **199:** 407–11.
6 Uhl HSM. A previously undescribed congenital malformation of the heart: almost total absence of the myocardium of the right ventricle. *Bull Johns Hopkins Hosp* 1952; **91:** 197–209.
7 Arcilla RA, Gasul BM. Congenital aplasia or marked hypoplasia of the myocardium of the right ventricle (Uhl's anomaly). *J Pediatr* 1961; **58:** 381–8.
8 Reeve R, MacDonald D. Partial absence of the right ventricular musculature: partial parchment heart. *Am J Cardiol* 1964; **14:** 415–9.
9 Marcus FI, Fontaine GH, Guiraudon G, *et al.* Right ventricular dysplasia: a report of 24 adult cases. *Circulation* 1982; **65:** 384–98.
10 Sugrue DD, Kamal S, Deanfield JE, *et al.* Assessment of right ventricular function and anatomy using peripheral vein infusion of Krypton 81m *Br J Radiol* 1983; **56:** 657–63.

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