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Two cases of bi-ventricular dysplasia associated with ventricular tachycardia and familial occurrence of sudden death

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Summary

Two strikingly similar patients with arrhythmogenic right ventricular dysplasia which severely impaired not only the right ventricle but also the left ventricle are described in association with familial occurrence of sudden death.

A 49-year-old man experienced syncope which was due to ventricular tachycardia. Electrocardiography revealed a first degree atrioventricular block, incomplete right bundle-branch block, T wave inversions in leads II, III, aVF and V_1 to V_5, and multiformal ventricular extrasystoles. Echocardiography and ventricular cineangiography showed not only the right ventricular dilatation with an aneurysm in the right ventricular apex, inflow and outflow tracts, but also mild dilatation of the left ventricle with left ventricular apical and posterior aneurysms. Radionuclide angiography also disclosed dysfunction of both ventricles, especially during exercise. His family history revealed that 3 members of his family died of sudden deaths.

A 56-year-old woman experienced syncope secondary to ventricular tachycardia, with left bundle-branch block. Electrocardiography showed complete right bundle-branch block, left axis

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deviation, and T wave inversions in leads V_1 to V_4. Echocardiography and ventricular cineangiography revealed not only marked right ventricular dilatation with the "triangle of dysplasia", but also a left ventricular aneurysm in the apex and posterior portion. Her elder brother died of a sudden death, and electrocardiograms of 2 members of her family showed ventricular extrasystoles and T wave inversions.

These 2 cases may well be termed "familial bi-ventricular dysplasia".

Key words

Arrhythmogenic right ventricular dysplasia Bi-ventricular dysplasia Sudden death Uhl's anomaly Right ventricular dilated cardiomyopathy Familial genesis

Introduction

Ventricular dysplasia has been reported to be mainly associated with lesions of the right ventricle but rarely with minor lesions of the left ventricle[1~3]. Despite earlier reports of prevalence of this disease in members of the same family[1,4,5], there is no definite evidence that it is hereditary. We studied 2 patients with strikingly similar arrhythmogenic right ventricular dysplasia (ARVD) who had severe dysfunction in both ventricles and had family history of multiformal ventricular tachycardia and sudden deaths. This disease may well be termed "familial bi-ventricular dysplasia".

Here we present a report of these 2 cases and a brief review of the literature.

Case reports

Case 1

A 49-year-old man, 163 cm tall and weighing 55 kg, a school caretaker, was admitted to our emergency room because of syncope. He had been aware of a pulse deficit since he was 20 years of age. During a medical check-up in the spring of 1986, abnormalities were detected in his electrocardiogram. At a clinic, he was diagnosed as having premature ventricular contractions and was given mexiletine prescribed by his physician, but 2 months prior to his admission to this hospital, he discontinued it of his own accord. On June 7, 1987 he experienced syncope and was admitted to our hospital as an emergency case.

At a physical examination on admission, the patient was drowsy, with his blood pressure, 102/80, the pulse, 80 and irregular, and the temperature, 36.7°C. Anemia, jaundice or cyanosis was not noted. The lungs were clear. The fourth heart sound was audible without murmurs. No neurological abnormalities were

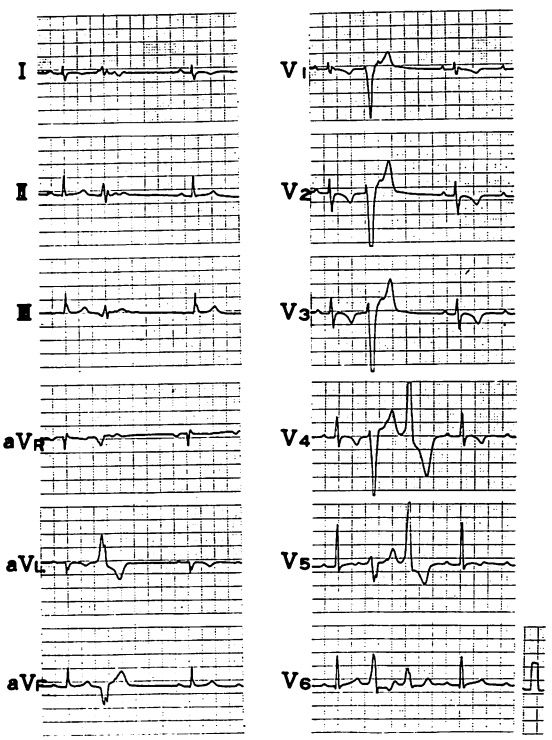

Fig. 1. Electrocardiogram on admission (Case 1).

First degree atrioventricular block, incomplete right bundle-branch block, T wave inversions in leads II, III, aVF and V_1 to V_5, and multiformal premature ventricular contractions are shown.

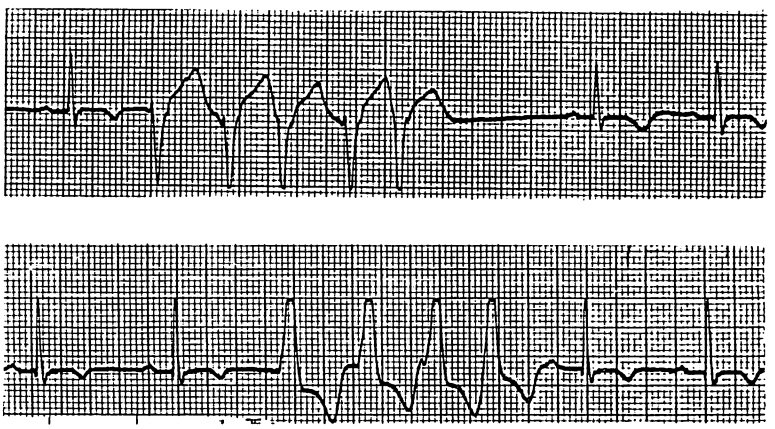

Fig. 2. Ambulatory monitoring showing multiformal ventricular tachycardia (Case 1).

observed.

Laboratory studies on admission disclosed elevated GOT, GPT and γ-GTP, presumably due to alcoholic liver injury. During his hospitalization, all these values returned to normal. Chest radiography showed mild cardiac enlargement without abnormalities in the lung fields. Electrocardiogram during sinus rhythm (**Fig. 1**) revealed first degree atrioventricular block, incomplete right bundle-branch block, T wave inversion in leads II, III, aVF and V_1 to V_5, and multiformal premature ventricular contractions. Ambulatory monitoring (**Fig. 2**) showed multiformal ventricular tachycardia. Echocardiogram (**Fig. 3**) revealed not only moderate dilatation of the right ventricle accompanied by akinesis with the thinning of the apical portion of the right ventricular wall and the inflow and outflow tracts, but also mild dilatation of the left ventricle with apical and posterior aneurysms accompanied by the thinning of its wall. Both the left and right atria were slightly dilated. Mitral valve prolapse was the only abnormalty observed in the 4 valves. Thallium-201 myocardial scintigraphy (**Fig. 4**) disclosed a persistent defect in the apex and postero-inferior left ventricular wall. Equilibrium radionuclide gated blood pool imaging at rest (**Fig. 4**) showed hypokinesis of the apico-anterior segment and akinesis of the postero-inferior segment. After ergometer exercise, the motion of the apico-anterior wall segment further deteriorated. The left ventricular ejection fraction decreased from 0.39 to 0.25; the right ventricular ejection fraction, from 0.25 to 0.17. Cardiac catheterization revealed slight elevation of the pulmonary and right ventricular pressures. Coronary arteries were free of stenoses or congenital anomalies. Left ventriculography (**Fig. 5**) showed an aneurysm in the apex and posterior segments with a calculated ejection fraction of 0.45. Right ventriculography (**Fig. 6**) showed dyskinesis in the inflow and outflow tracts, and diffusely severe hypokinesis in the infero-posterior wall with a calculated ejection fraction of 0.30.

The patient was maintained on bed rest. Immediately after admission, he received potassium supplements and intravenously-administered lidocaine, which relieved ventricular arrhythmia. The efficacies of procainamide, disopyramide, aprindine, mexiletine, propranolol, alprenolol and acebutolol were evaluated while the patient was in the steady state. Procainamide and aprindine proved to alleviate ventricular arrhythmia.

The pedigree of the patient is shown in **Fig. 7**. His elder sister died of a sudden death when she was 33 years of age. His niece died suddenly while swimming, at the age of 15 years, and his nephew died at the age of 14 during

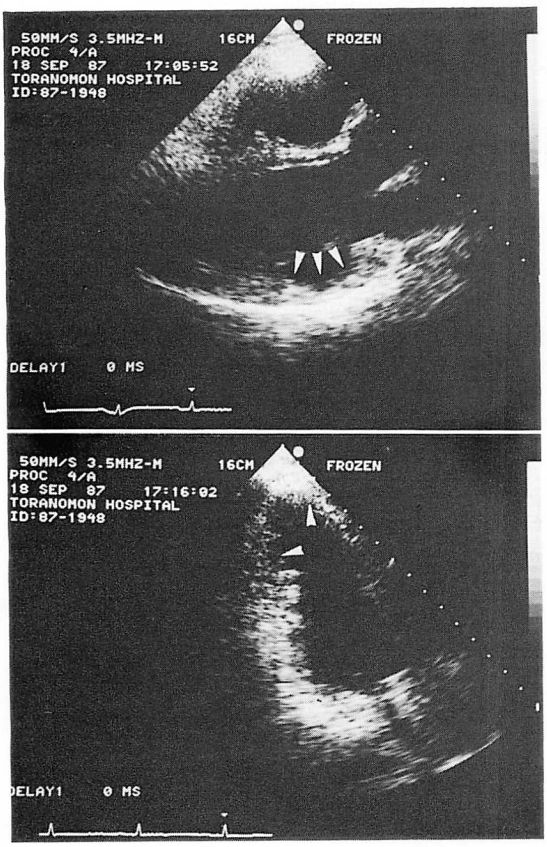

Fig. 3. Two-dimensional echocardiograms (Case 1).

Top: Parasternal long-axis view. White triangles indicate a posterior aneurysm of the left ventricle. Bottom: Apical left anterior oblique equivalent view. White triangles indicate an apical aneurysm of the left ventricle.

long-distance running. His younger sister has abnormalities on electrocardiography, which are similar to the patient's.

Case 2

A 56-year-old woman, 143 cm tall and weighing 51 kg, a diver, was admitted to our emergency room because of syncope. She had been in good health until April 3, 1988, when she was found lying unconscious after diving.

At a physical examination on admission, she was drowsy. The blood pressure and pulse were unobtainable. Anemia, jaundice, cyanosis or struma was not noted. Heart sound was faint but respiratory sound was normal. There were no neurological abnormalities.

Laboratory studies on admission revealed slightly elevated GOT, CPK and leukocyte count, presumably attributable to circulatory insufficiency secondary to ventricular tachycardia, and decreased serum potassium. These values reverted within normal limits soon after admission. Chest radiograph disclosed cardiac enlargement with a cardiothoracic ratio of 0.61. The lung fields were normal. Electrocardiogram on admission (**Fig. 8**) revealed ventricular tachycardia with left bundle-branch block at a rate of 207 beats/min, which reverted to normal after intravenous administration of 100 mg lidocaine. Electrocardiogram during normal sinus rhythm (**Fig. 8**) showed complete right bundle-branch block, left axis deviation, T wave inversions in leads V_1 to V_4 and ventricular extrasystoles. Two-dimensional echocardiogram (**Fig. 9**) showed marked right ventricular dilatation and akinesis with thinning of the wall, the apex, and the inflow and outflow tracts of the right ventricle. There were aneurysms in the left ventricle with thinning of the wall in the apex and the posterior segments. There were no abnormalities in the valves or the atria. Cardiac catheterization and coronary arteriography were normal. Right ventriculography (**Fig. 10**) exhibited marked enlargement in the akinetic regions and bulges in the apex and the inferior wall, and deep fissuring with a reduction of the right ventricular ejection fraction to 24%. Left ventriculography (**Fig. 11**) also showed aneurysms of the apex and of the inferior segment without left ventricular dilatation.

Ambulatory monitoring on admission showed unsustained, monomorphic ventricular tachycardia, which returned to normal sinus rhythm after intravenous infusion of 100 mg lidocaine. Subsequent treatment with mexiletine 300 mg/day and supplementary potassium prevented recurrence of the ventricular tachycardia.

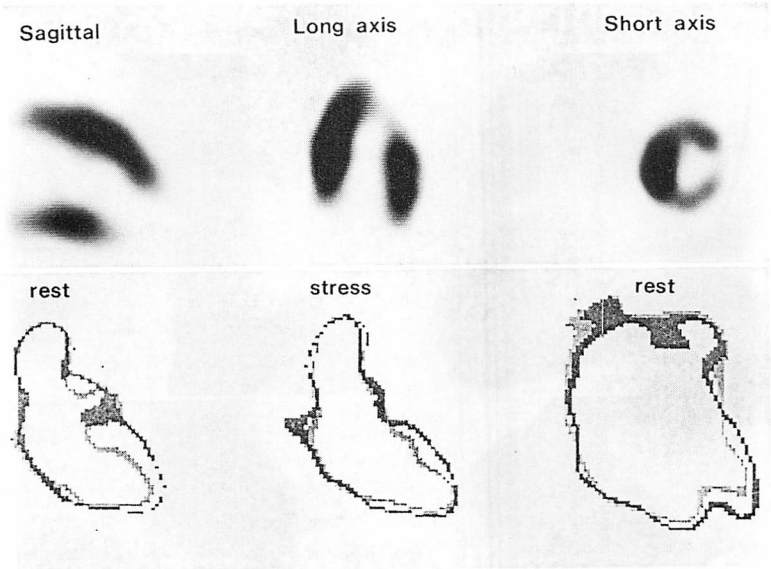

Fig. 4. Radionuclide studies (Case 1).
Top: Thallium-201 myocardial scintigram reveals a persistent defect in the apex and postero-inferior left ventricular wall. Bottom: Equilibrium radionuclide gated blood pool imaging at rest shows hypokinesis of the anterior segment and akinesis of the postero-inferior segment. After ergometer exercise, the wall motion of the anterior segment further deteriorated.

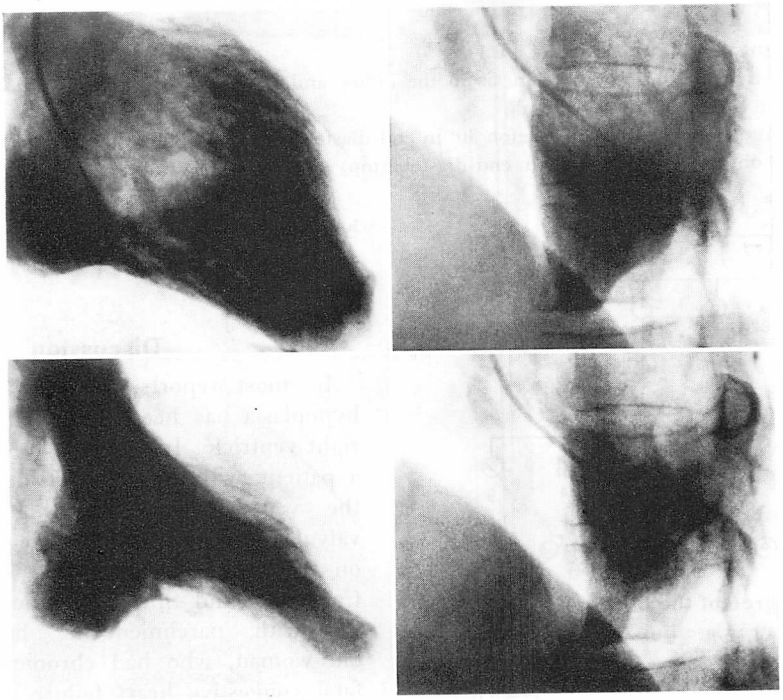

Fig. 5. Left ventriculograms (Case 1).
Left ventriculogram shows aneurysm in the apex and posterior segment.
Left: Right anterior oblique projection 30° in end-diastole (top) and end-systole (bottom). Right: Left anterior oblique projection 60° in end-diastole (top) and end-systole (bottom).

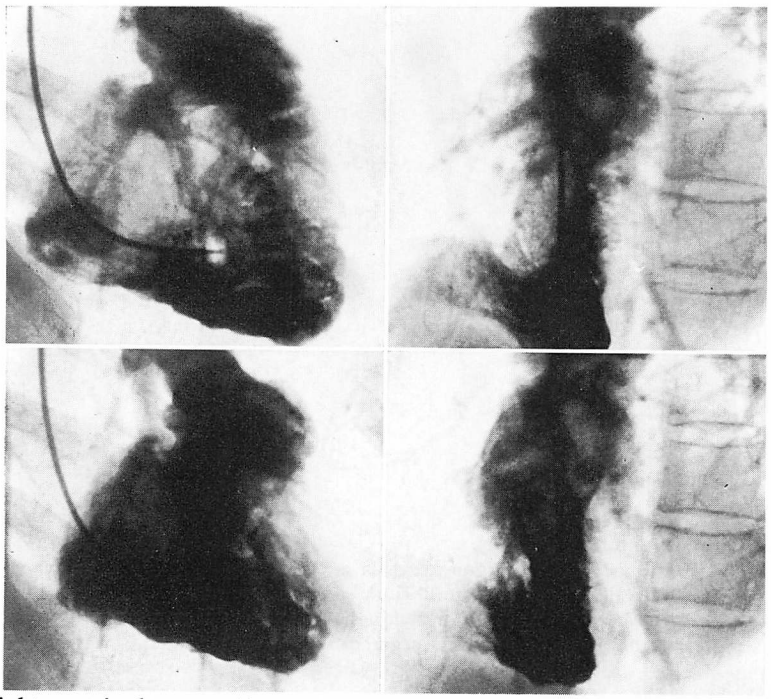

Fig. 6. Right ventriculograms (Case 1).
Right ventriculogram shows dyskinesis in the inflow and outflow tracts, and diffuse and severe hypokinesis in the infero-posterior wall.
Left: Right anterior oblique projection 30° in end-diastole (top) and end-systole (bottom). Right: Left anterior oblique projection 60° in end-diastole (top) and end-systole (bottom).

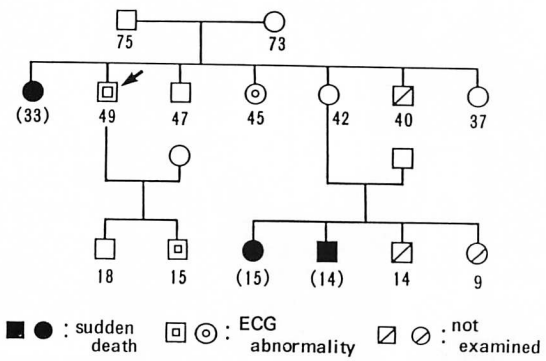

Fig. 7. Pedigree of the family (Case 1).
Arrow indicates Case 1.

The pedigree of the patient is presented in **Fig. 12**. Her elder brother died suddenly during his sleep at 43 years of age. Electrcardiograms of her elder sister and younger brother revealed frequent ventricular extrasystoles and extensive T wave inversions in the precordial leads.

Discussion

In most reports, ventricular dysplasia or hypoplasia has been described to involve the right ventricle. In 1905, Osler[6] first described a patient with "parchment-like" thinning of the ventricular wall without coronary or valvular disease. In 1950, Segall[7] elaborated on this briefly reported by Osler. In 1952, Castleman and Sprague[8] reported the second case with "parchment-like" heart; a 23-year-old woman, who had chronic and eventually fatal congestive heart failure. The third case was reported in 1952 by Uhl[9]. A 7-month-old girl had severe right-sided heart failure and marked thinning of most of the right ven-

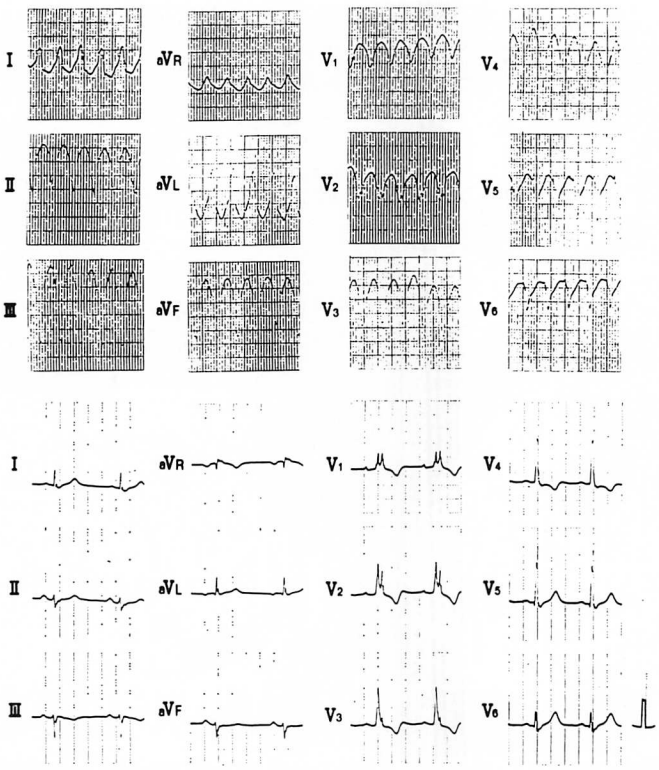

Fig. 8. Electrocardiograms (Case 2).

Top: The ECG on admission shows ventricular tachycardia with left bundle-branch block configuration at a rate of 207 beats/min. Bottom: The ECG with normal sinus rhythm shows complete right bundle-branch block, left axis deviation and T wave inversions in leads V_1 to V_4.

tricular wall. Since the publication of Uhl's report, "parchment-like" thinning of the right ventricular wall has been widely called "Uhl's anomaly". Waller et al[10], however, called this condition "congenital hypoplasia of ventricular myocardium".

In 1978, Frank and Fontaine et al[11] reported 4 cases of recurrent ventricular tachycardia associated with isolated right ventricular cardiomyopathy. Angiography revealed hypokinesis of the right ventricle with localized dyskinetic zones. They named this syndrome arrhythmogenic right ventricular dysplasia (ARVD). ARVD is histologically characterized by partial degeneration of the myocardial wall. Most muscle fibers are replaced by fatty and fibrous tissue[3]. The lesions are usually noted on the anterior surface of the pulmonary infundibulum, at the apex of the right ventricle, or at the inferior wall of the right ventricle, and these 3 sites of the lesion form a "triangle of dysplasia"[1]. According to Marcus et al[1], the male/female prevalence ratio was 2.7: 1 and that the mean age of the patients on admission was 39. They considered ARVD to be a stage of the developmental anomaly and that Uhl's anomaly in infancy and ARVD comprise one spectrum of the same disease entity.

ARVD used to be believed to involve the right ventricle exclusively[2,12]. However, left ventricular abnormalities have recently been described with right ventricular dysplasia[13]. Manyari et al[13] prospectively studied 6 patients with ARVD using radionuclide angiography.

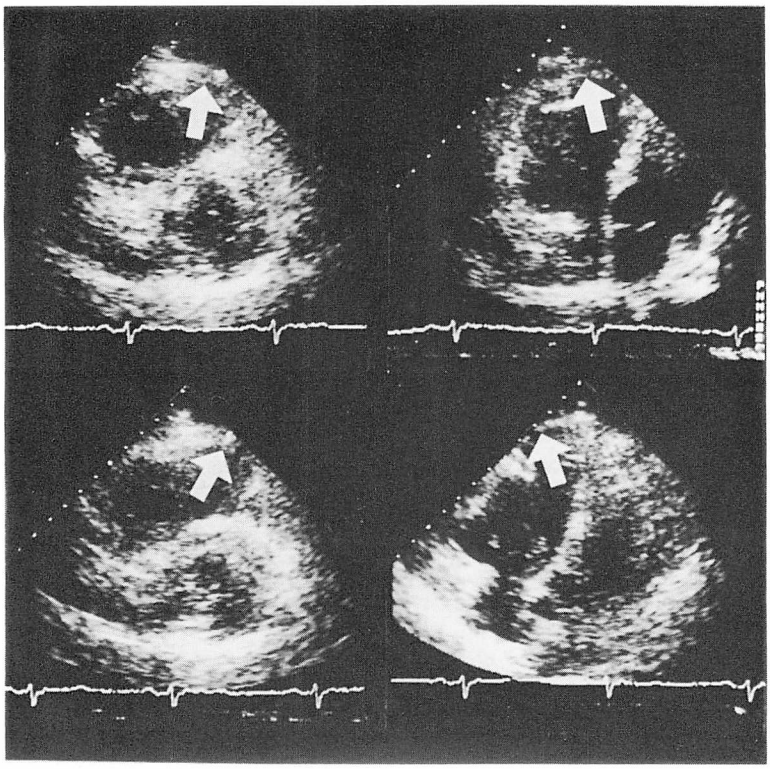

Fig. 9. Two-dimensional echocardiograms (Case 2).
White arrows indicate aneurysms.
Left: Parasternal short-axis view in end-diastole (top) and end-systole (bottom). Right: Apical four-chamber view in end-diastole (top) and end-systole (bottom).

Left ventricular ejection fractions at rest were lower in these patients than in control subjects with the difference being significant during peak exercise. Two had left ventricular septal hypokinesia at rest and all 6 patients exhibited left ventricular dysfunction during exercise. Webb et al[14] reported 4 cases of ARVD accompanied by left ventricular abnormalities on radionuclide angiography or catheterization. Our patient (Case 1) also had mild left ventricular dilatation and left ventricular dysfunction on left ventriculography. Moreover, the left ventricular ejection fraction was depressed at rest on radionuclide ventriculography and reduced to a lower value during peak exercise. The pathogenesis of the left ventricular involvement in ARVD has been associated with 1) dysfunction of the left ventricle accompanied by that of the right ventricle, 2) left ventricular myocardial damage in association with ischemia due to ventricular tachycardia and 3) myocardial changes similar to those in the right ventricle. Marcus et al[1] reported that of 24 adult patients with right ventricular dysplasia, one had left ventricular involvement. The morphology and microscopic findings of the left ventricle of this patient were the same as those of the patients with the right ventricular abnormalities. Although ARVD primarily involves the right ventricle, abnormalities in the left ventricle, another part of a spectrum of the disease, seemed to be later recognized which may have implications with regard to cardiac function and

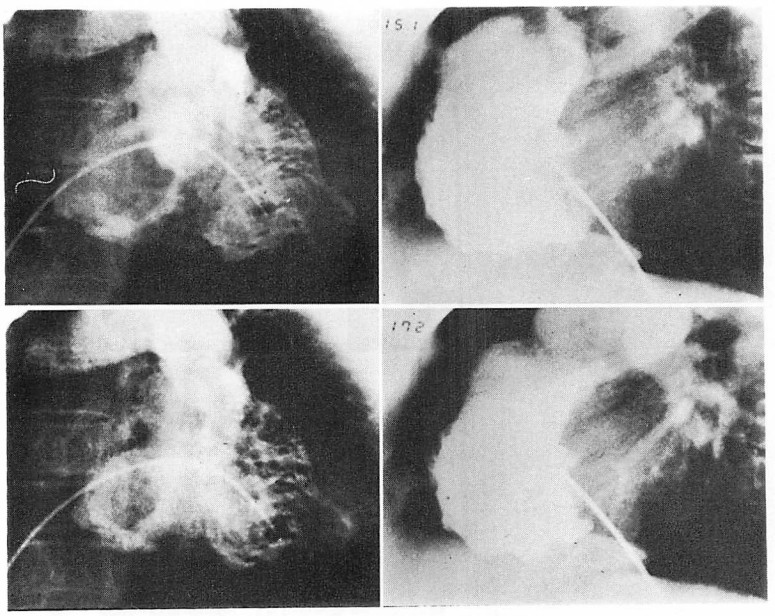

Fig. 10. Right ventriculograms (Case 2).

Right ventriculogram shows the marked enlargement, akinetic areas and bulges of the apex and inferior wall, and deep fissuring.

Left: Anteroposterior projection in end-diastole (top) and end-systole (bottom). Right: Lateral projection in end-diastole (top) and end-systole (bottom).

arrhythmogenesis[14]. The electrocardiographic findings of the present 2 cases consisted of extensively induced T wave inversions, left anterior hemiblock, an RSR pattern in V_1 and ventricular tachycardia with a left bundle-branch block configuration. The ventricular tachycardias in Case 2 were multiformal. Furthermore, an echocardiogram, radionuclide examination and ventricular cineangiogram revealed a "spotted" aneurysm of the left ventricle in both cases. If the left ventricular impairment of these cases had consisted of secondary changes associated with the right ventricular involvement, aneurysmal formation of the left ventricle in general would not have become "spotted" but "uniform". From this standpoint, the present 2 cases may as well be called "bi-ventricular dysplasia".

Right ventricular dilated cardiomyopathy was separately described by Murata et al[15], Fitchett et al[16] and Murakami et al[17]. This entity is characterized by marked right ventricular myocardial depletion, which is similar to the pathological features of ARVD. Most muscle fibers, however, were replaced by fibrous tissue and the right ventricle was diffusely dilated and impaired. Moreover, the left ventricle was mildly to moderately involved in right ventricular dilated cardiomyopathy. A report by Murakami et al[17] suggested a strong hereditable trait of this entity. Since the degeneration of the myocardium is thought to be the main process of the development of the right ventricular dilated cardiomyopathy, some of the right ventricular dilated cardiomyopathies may overlap those of ARVD.

Several cases of the familial occurrence of ARVD and Uhl's anomaly, including our 2 cases are presented in **Table 1**. These reports reveal a striking predominance of men, but do not seem to indicate an autosomal recessive genetic pattern. Rather, it is more likely to be

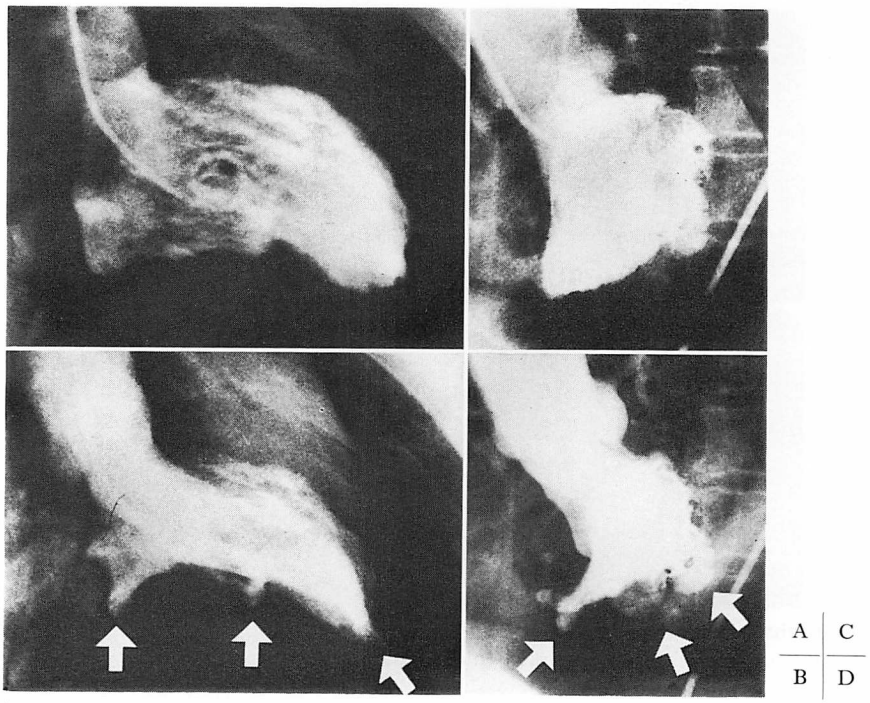

Fig. 11. Left ventriculograms (Case 2).
Arrows indicate aneurysms.
A and B: Right anterior oblique projection in end-diastole and end-systole. C and D: Left anterior oblique projection in end-diastole and end-systole.

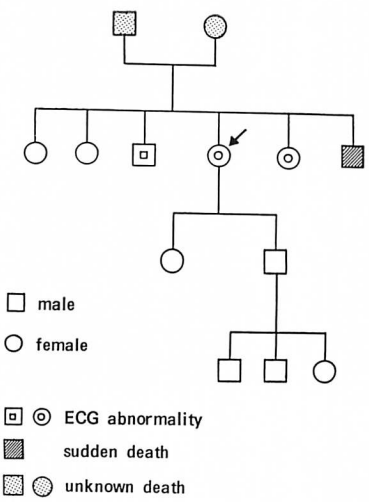

Fig. 12. Pedigree of the family (Case 2).
Arrow indicates Case 2.

a pattern of autosomal dominance with incomplete penetrance. Another possibility[5] might be familial exposure to a common toxic or infectious agent, perhaps with a genetic predisposition.

We recommend that systemic in-depth familial studies of all demonstrated cases of ARVD must be performed, and that follow-up studies should be done for all patients identified as having arrhythmias or isolated right ventricular morphologic abnormalities.

Table 1. Proven or suspected cases of familial ARVD

Author (Reference No.)	Year	No. of patients & sex	Right-sided VT	Sudden death	Surgery	Necropsy
Marcus (1)	1982	2 M	2		1	
Waynberger (18)	1974*	2 M	2			
Hoback (19)	1981	3 M	2	2		2
Guiraudon (20)	1983	4 M	1	3		
Guiraudon (20)	1983	6 M	1	5	1	
Diggelmann (21)	1984	3 M, 2 F	1	3	1	
Child (22)	1984	1 M, 2 F		2	1	
Affatato (23)	1985	5 M	2	1		
Nava (24)	1987	10 M, 6 F	2	5		2
Ruder (5)	1985	1 M, 2 F	2	1		1
Rakovec (4)	1986	1 M, 1 F	1	1		1
Nakanishi (25)	1986	7 M, 2 F	2	7		
Laurent (26)	1987	5 M, 1 F	4			
B-Lundqvist (27)	1987	2 M	2		1	
Hirooka (28)	1988	3 M, 1 F	3			
Present Case 1		3 M, 3 F	2	3		
Present Case 2		2 M, 2 F	1	1		

* Previously published as right ventricular dysplasia or disease and was later established as a clinical entity of arrhythmogenic right ventricular dysplasia (ARVD), including 3 families, one of which probably fulfills the criteria of dysplasia.

M=male; F=female; VT=ventricular tachycardia.

要　　約

家系内に突然死の集積を有し，心室頻拍を呈した両心室異形成症ともいうべき2症例

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**中野　赳

家系内に突然死の集積と類似する心電図異常を認め，心エコー図，心室造影，心核医学検査にて両心室の心尖部，流出路，流入路に壁の非薄化を伴う心室瘤様変化を有し，心室頻拍を呈した両心室異形成症と言うべき2症例を報告した．

症例 1：　49歳，男性．失神にて入院．心電図にて第1度房室ブロック，不完全右脚ブロック，II, III, aV_F, $V_1 \sim V_5$ の T 波逆転を認め，モニター心電図にて多源性心室頻拍を見た．心エコー図および心室造影にて右室心尖部，流入路，流出路に心室瘤様変化を認め，また左室にも心尖部と後壁に同様の変化を見た．心核医学検査にて両心室の機能低下があり，特に運動負荷時の強い機能低下を認めた．家系内に3人の突然死を認め，妹に同様の心電図異常を見た．

症例 2：　56歳，女性．失神にて入院．入院時心電図は左脚ブロック型の心室頻拍を呈し，リドカインの静注にて洞調律に復した．洞調律時の心電図は，完全右脚ブロック，左軸偏位，$V_1 \sim V_4$ の T 波逆転であった．心エコー図，心室造影にて右室の心尖部，流入路，流出路に心室瘤様変化を認めるだけでなく，左室にも同様の変化を心尖部，後壁に認めた．家系内に1人の突然死を認め，他の2人に同様の心電図変化を見た．

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