A 60-year-old man was hospitalized for close examination of hypertrophic cardiomyopathy (HCM). Atrial fibrillation has been noted by medical examination since 1985, and cardiomegaly since 1987, but the patient did not undergo close examination because of the absence of symptoms. In a medical examination performed by a local physician in December 1997, HCM was found by echocardiography. His father had died suddenly at the age of 54 years, and his eldest son was found to have HCM. The patient’s blood pressure was 138/84 mmHg, and his pulse was irregular (80 beats/min).

Cardiac sounds showed a systolic murmur of grade II in the apex cordis. Hematological and biochemical examination revealed elevation of total bilirubin (1.8) and direct bilirubin (0.6), which were spontaneously reduced during hospitalization, but no other abnormalities were noted. Cardiomegaly (cardiothoracic ratio, 62.5%) was observed by thoracic radiography (Fig 1). No abnormality in the lungs was observed. Atrial fibrillation and left ventricular hypertrophy were detected by electrocardiography (Fig 2). Echocardiography showed concentric hypertrophy with a 14-mm thick interventricular septum and a 13-mm thick left ventricular posterior wall (Fig 3A,B). The left ventricular end-diastolic diameter was 44 mm, the left ventricular end-systolic diameter was 28 mm, and the ejection fraction was 66.3%, showing good left ventricular systolic function. Dilation of the right ventricular cavity and paradoxical motion of the interventricular septum were observed by M-mode echocardiography (Fig 3B). The left atrium was also dilated (left atrial diameter: 38 mm). In the cross-section images of the 4 cavities, distinct dilation of the right ventricle and very mild tricuspid regurgitation were observed (Fig 3C). There was no distinct left-to-right shunt such as an atrial septal defect. By magnetic resonance imaging, the left ventricular end-systolic diameter was 29 mm, the left ventricular end-diastolic diameter was 41 mm, the interventricular septal wall thickness was 15 mm, and the left ventricular posterior wall thickness was 13 mm showing concentric wall hypertrophy (Fig 4). The left ventricular function was good. On the other hand, the right ventricular end-systolic diameter was 32 mm, and the right ventricular end-diastolic diameter was 45 mm, showing distinct dilation of the right ventricular cavity. No abnormalities in the inner pressure in both right and left sides of the heart were detected by cardiac catheterization (Table 1), and there was no gradient between aortic and left ventricular pressures.

**Key Words:** Dilated phase; Hypertrophic cardiomyopathy; Right ventricular dilatation

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**A Patient With Hypertrophic Cardiomyopathy Accompanied by Right Ventricular Dilation of Unknown Cause**

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Hypertrophic cardiomyopathy (HCM) is a disease characterized by an unknown cause of hypertrophy in the left or right ventricle. The dilated phase of HCM shows disease conditions resembling dilated cardiomyopathy, such as ventricular dilation, thin ventricular wall, and reduction of the ejection fraction. A patient presented with left ventricular concentric hypertrophy accompanied by right ventricular dilatation of unknown cause. Right ventricular endomyocardial biopsy specimens showed characteristic myocardial disarray. Therefore, there is the possibility that the patient had right and left ventricular HCM in the process toward the dilated phase, in which dilatation first occurred in the right ventricle. (*Jpn Circ J* 1999; 63: 137–140)

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*Fig 1. Thoracic radiography. The cardiothoracic ratio was 62.5%.*
Table 1 Results of Cardiac Catheterization

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<td>Right pulmonary arterial pressure (mmHg)</td>
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<td>Left ventricular pressure (mmHg)</td>
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<tr>
<td>Aortic pressure (mmHg)</td>
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<td>88</td>
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Fig. 2. Electrocardiography. Atrial fibrillation and left ventricular hypertrophy can be seen.

Fig. 3. Echocardiography. Parasternal long axis view (A), M-mode (B), and 4-chamber view (C). Left ventricular concentric hypertrophy and dilatation of right ventricular cavity can be seen. RA, right atrium; RV, right ventricle; LA, left atrium; LV, left ventricle.
The cardiac output, left ventricular diastolic and systolic volumes, and ejection fraction were normal (Table 2). Coronary angiogram showed no significant stenosis in the right or left coronary arteries. Distinct dilation of the right ventricle was observed by right ventriculography, and the ejection fraction was reduced to 42% (Fig 5). Tricuspid regurgitation was very mild (Sellors classification I). Histologically, right ventricular endomyocardial biopsy specimens revealed abnormal arrangement of subendocardial muscles and moderate myocyte hypertrophy (mean myocyte diameter: 21μm) (Fig 6).

### Discussion

HCM, which is characterized by asymmetric hypertrophy of the left or right ventricle including the interventricular septum, does not usually show dilation of the ventricular cavities. However, a dilated phase of HCM showing dilation of the ventricular cavities, thinning of the ventricular wall, and reduction of systolic function is sometimes observed. Our patient, with normal left ventricular systolic function despite its concentric hypertrophy that was accompanied by dilation of the right ventricular cavity and reduction of right ventricular systolic function, was considered to be a rare case.

In this patient, characteristic HCM findings such as asymmetric septal hypertrophy were not observed, but concentric hypertrophy of the left ventricular wall was noted without definite a cause such as aortic stenosis. His blood pressure value of 138/88 was close to the borderline.
period. Left bundle-branch block ventricular tachycardia has been reported at least once during the disease period, usually defined as a condition in which the patient has had ventricular arrhythmia during the disease period. ARVD is characterized by fatty and fibroadipose degeneration of cardiac muscles and shows marked dilation of the right ventricle without any loss of left ventricular function. ARVD accompanied by left ventricular failure has been reported, but the failure is due to local wall motion and slight reduction of diffuse wall motion in the left ventricle, which does not agree with the conditions found in the present patient. Right ventricular endomyocardial biopsy specimens in hypertensive cases. In our patient, such findings were not observed by endomyocardial biopsy.

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Uhl disease is a congenital heart disease first described by Uhl in 1952 as ‘almost total absence of the myocardium of the right ventricle.’ This disease is characterized by fibrous and fatty degeneration in the entire layers of cardiac muscles, and a parchment-like right ventricular wall is observed. In our patient, such findings were not observed by endomyocardial biopsy.

Right ventricular endomyocardial biopsy specimens from our patient showed myocardial fiber disarray, characteristic of HCM. These results suggested that the patient had right and left ventricular HCM in the process toward the dilated phase, in which dilatation first occurred in the right ventricle. In this condition, left ventricular dilation, which may cause heart failure, is likely to occur in the future; therefore, long-term follow-up observation is necessary. Further studies on patients with similar disease conditions are needed.

References