

## **Seventeen Year Follow-up of a Patient with Hypertrophic Cardiomyopathy Which Progressed to Dilated Cardiomyopathy**

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### **SUMMARY**

The progression of hypertrophic cardiomyopathy (HCM) to dilated cardiomyopathy (DCM) has been rarely reported. We report a patient with HCM who had been followed for 17 years and in whom DCM became evident.

### **Additional Indexing Words:**

Familial hypertrophic cardiomyopathy      Mitral regurgitation

**T**HE cardiomyopathies are defined as primary disease of cardiac muscle and divided into 3 major groups: hypertrophic, dilated and obliterative.<sup>1)</sup> In the hypertrophic type, massive hypertrophy, resistance to filling of the ventricles, small ventricular size and, often, obstruction to outflow from left ventricle are important factors. By contrast, dilated cardiomyopathy is manifested by a dilated heart without outflow tract obstruction and with only moderate hypertrophy associated with severe heart failure. However, the natural history of cardiomyopathies remained unknown. The patient described in this report had evidence of HCM in his early clinical course. During a 17 year follow-up, he developed progressive congestive heart failure with a dilated left ventricle. Thus, HCM developed into DCM.

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## CASE REPORT

The patient was a 52-year-old Japanese man. At 35 years of age (1965), cardiomegaly was observed on a chest roentgenogram taken at a routine medical check up. The electrocardiogram had showed left ventricular hypertrophy and marked strain-type ST and T changes (Fig. 2). Neither high blood pressure nor heart failure had been noted at that time. He had consumed 150 to 400 ml of Japanese Sake (14–15% alcohol) daily for about 20 years (1955–1977). He weighed 63 Kg, was in management and had no particular form of athletic exercise. He had no evidence of pulmonary disease including cor pulmonale. He was asymptomatic, but was admitted for clinical evaluation of cardiomegaly in 1977. A fourth heart sound was audible but there was no heart murmur. His blood pressure was 115/70 mmHg. The chest roentgenogram showed moderate cardiac enlargement (CTR=65), with left ventricular predominance. The ECG revealed an abnormal left axis deviation ( $-46^\circ$ ), left atrial overload, Q wave at leads I and  $aV_L$ , and left ventricular hypertrophy with a negative T wave in leads  $V_{5,6}$ . The vectorcardiogram revealed left ventricular and septal hypertrophy. The apex cardiogram showed a prominent 'a' wave. The M-mode echocardiogram (Fig. 1) also showed hypertrophy of both the interventricular septum (IVS) and the left ventricular posterior wall (LVPW). There was a B bump between the A-C intervals of the mitral valve and a dilated left atrium. Cardiac catheterization performed on June 7, 1977 revealed elevated left ventricular end-diastolic pressure (35 mmHg) but no pressure gradient in the left ven-

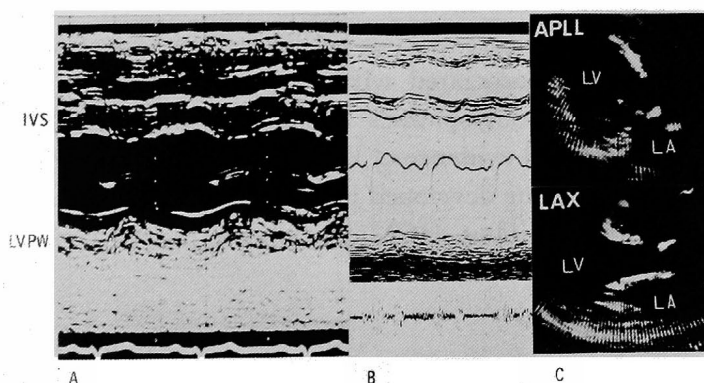


Fig. 1. M-mode left ventricular echocardiogram (A: 1977, B: 1982) and apical long-axis view (APLL) and long-axis (LAX) two-dimensional echocardiogram (C: 1982).

A: LA 50 mm, LVDd 45 mm, LVDs 35 mm, IVST 15 mm, LVPWT 12 mm,  
B: LA 55 mm, LVDd 65 mm, LVDs 57 mm, IVST 8 mm, LVPWT 8 mm.

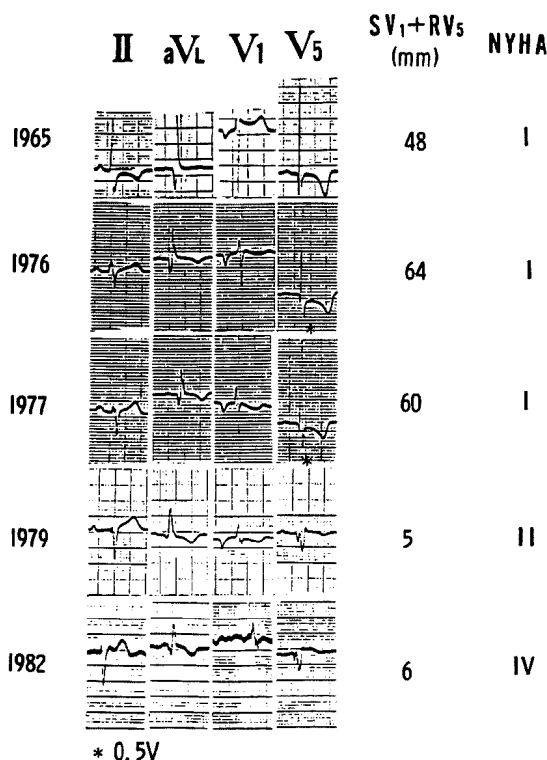


Fig. 2. Progressive increase and then decrease in the precordial voltage criteria of left ventricular hypertrophy. In 1979, I° A-V block and in 1982, atrial fibrillation were noted.

tricle. The left ventriculogram showed hypertrophy of the LVPW and diminished systolic function. Mitral regurgitation of grade I was noted. There were no abnormalities on the coronary angiogram. Diagnosis of hypertrophic nonobstructive cardiomyopathy was made. He had a family history of cardiomegaly (4 out of 13 members). From his brother's examination in our hospital, this is a familial hypertrophic cardiomyopathy. He was discharged and remained asymptomatic until 1979 when he first noticed mild dyspnea on exertion. The echocardiogram revealed an increased LV dimension and decreased LV function, compared to findings in 1977 (LVDd 45 mm, LVDs 35 mm, IVST 15 mm, LVPWT 12 mm). In 1982, the M-mode and two-dimensional echocardiograms showed an even more marked distention of the LV cavity (LVDd 65 mm, LVDs 57 mm) and a decrease in both IVST (8 mm) and LVPWT (8 mm). The ejection fraction was markedly decreased (33%). The CTR on the chest X-ray was 62%. During the 17 years of follow-up, the ECG showed an increase of 16 mm in the voltage of SV<sub>1</sub>+RV<sub>5</sub> for the first 11 years, but then decreased to 5 mm in

1979. A rapid decrease was observed between 1977–1979, but the patient remained in good condition at home (NYHA I). He was readmitted on December 1982 with a severe dyspnea and hypotension. Physical examination showed severe congestive heart failure and he died suddenly with a ventricular tachycardia on January 1983. Permission for autopsy was refused by the family. Beta-blocking agents and/or  $\text{Ca}^{++}$  antagonists were never prescribed for this patient.

### DISCUSSION

The clinical course of cardiomyopathy is remarkably variable and long-term follow-up without chemotherapy which has an untoward effect on prognosis has been rarely reported. McKenna et al reported the natural history of HCM,<sup>2)</sup> and noted that 4 out of 100 patients had a decreased left ventricular hypertrophy on the ECG. Two cases out of 4 had been treated for a long period with a high dose of propranolol. The other 96 patients showed progressive hypertrophy and a poor prognosis. Beder et al reported a child with HCM who was treated with propranolol and in whom congestive cardiomyopathy developed.<sup>3)</sup> These cases were all the obstructive type of cardiomyopathy. Conversion of obstructive cardiomyopathy to nonobstructive cardiomyopathy is usually observed when progressive clinical deterioration and severe congestive heart failure developed.<sup>4)</sup> Although no data on cardiac catheterization during early phases are available, this patient was free from clinical symptoms and a pathological heart murmur when he first came to medical attention at age 35, and continued to be asymptomatic for another 12 years. This leads us to think that this patient had nonobstructive cardiomyopathy from the beginning of the 17 year follow-up period. However, it is unknown whether our patient had “obstructive” hemodynamics before coming to medical attention. The alcohol intake may have modified the clinical course, but it is hard to interpret as alcoholic cardiomyopathy, because of the volume of alcohol intake and the definite family history of hypertrophic cardiomyopathy which is independent of alcohol intake. Since an autopsy was not performed, a silent transmural infarction, which is found in 15% of patients with hypertrophic cardiomyopathy,<sup>5)</sup> cannot be ruled out.

### ACKNOWLEDGMENT

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