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Recurrent Syncope Induced by Left Ventricular Outflow Tract Obstruction : Demonstration in a Patient With Hypertrophic Obstructive Cardiomyopathy

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Abstract

A 70-year-old man presented with repeated syncope induced by left ventricular outflow tract obstruction. He was referred to us because of repeated syncope with convulsion at rest.

During syncope, electrocardiography showed marked ST segment depression with negative T waves in leads I, II, aV_L, aV_F and V_2–V_6 but no arrhythmias. Echocardiography revealed asymmetric septal hypertrophy and complete obstruction of the left ventricular outflow tract due to systolic anterior movement of anterior mitral leaflet and concomitant severe mitral regurgitation. During the catheterization study, syncope with convulsion developed repeatedly without antecedent cause, and was associated with a decrease in systemic blood pressure. Simultaneous pressure monitoring of the left ventricle and femoral artery showed a significant pressure gradient (maximum 110 mmHg). During each episode, systemic blood pressure rose spontaneously with the recovery of consciousness over several minutes.

He received temporary atrioventricular sequential pacing and underwent successful mitral valve replacement. Four years later, he was doing well.

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Key Words
Cardiomyopathies (hypertrophic), Syncope, Death (sudden)

INTRODUCTION

Sudden cardiac death is recognized as an important characteristic of the natural history of hypertrophic cardiomyopathy. However, except for arrhythmias, the cause of death remains uncertain in the majority of patients who die suddenly[1-3]. We present a patient with hypertrophic obstructive cardiomyopathy (HOCM) who had recurrent syncopal attacks induced by hemodynamic deterioration secondary to left ventricular outflow tract (LVOFT) obstruction. During syncopal attacks, the transaortic pressure gradient gradually increased and he consequently lost consciousness. However, there was almost no pressure gradient while the patient was asymptomatic.

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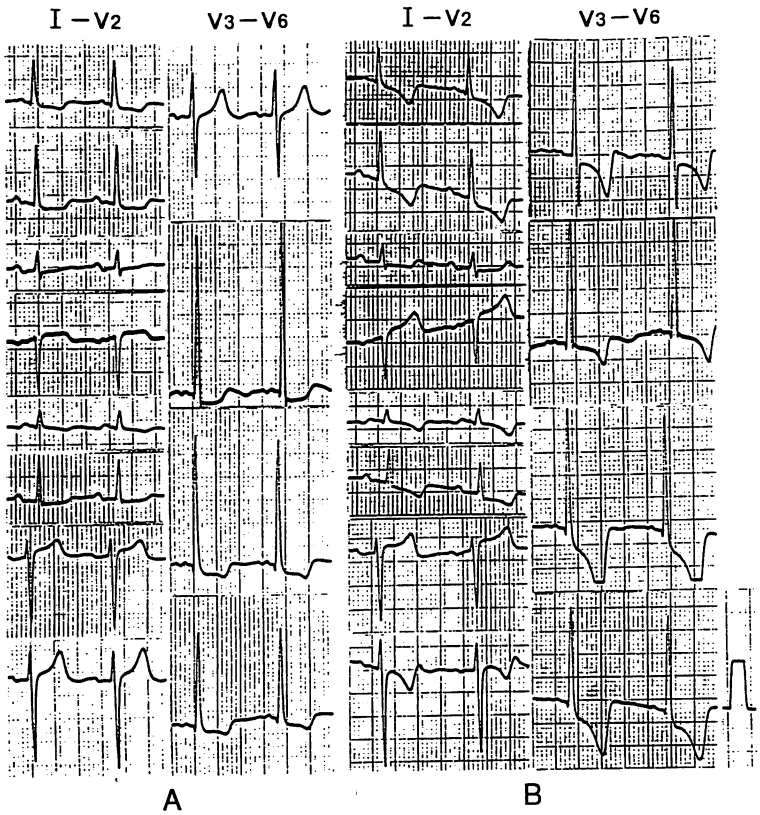

Fig. 1 Twelve-lead electrocardiograms before (*A*) and during syncopal attack (*B*)

A : The baseline electrocardiogram shows normal sinus rhythm, left ventricular hypertrophy, ST segment depression in leads I, II, aVL, aVF and V4–V6, and negative T waves in leads I, aVL and V5–V6.
B : The electrocardiogram during the attack shows marked ST segment depression with negative T waves in leads I, II, aVL, aVF and V2–V6.

Selected abbreviations and acronyms

HOCM = hypertrophic obstructive cardiomyopathy
LVOFT = left ventricular outflow tract

CASE REPORT

A 70-year-old man was referred to us on November 21, 1992 because of repeated syncope with convulsion. He had experienced syncope with increasing frequency for 3 years, but especially in the last 2 weeks. On admission, he was alert with systemic blood pressure of 120/72 mmHg. Cardiac auscultation revealed a II/VI systolic ejection murmur with the maximal point at the apex.

Electrocardiography (ECG) showed normal sinus rhythm, left ventricular hypertrophy, ST segment depression in leads I, II, aVL, aVF and V4–V6, and negative T waves in leads I, aVL and V5–V6 (**Fig. 1-A**). Echocardiography revealed asymmetric septal hypertrophy (left ventricular posterior wall thickness at end-diastole : 13.7 mm, interventricular septum thickness at end-diastole : 26.0 mm), systolic anterior motion of the anterior mitral leaflet and mild mitral regurgitation (**Figs. 2-A, B**). He had repeatedly developed syncope with convulsion, when the systemic blood pressure fell progressively below 40 mmHg. However, monitoring ECG showed normal sinus rhythm without arrhythmias. Cardiac auscultation revealed the development of a new III/VI holosystolic murmur at the apex and 12-lead ECG showed marked ST segment depression with negative T waves in leads I, II, aVL, aVF and V2–V6 (**Fig. 1-B**). About 30 sec after the episode, systemic blood pressure rose spontaneously with recovery of consciousness and there were no residual neurological abnormalities. During the episode, echocardiography revealed that the ejection flow velocity in LVOFT had gradually accelerated; subsequently, the anterior mitral leaflet moved to the interventricular septum and finally touched the septum during systole. This peculiar movement of the anterior mitral leaflet resulted in complete obstruction of the LVOFT and concomitant grade IV mitral regurgitation (**Figs. 2-C, D**).

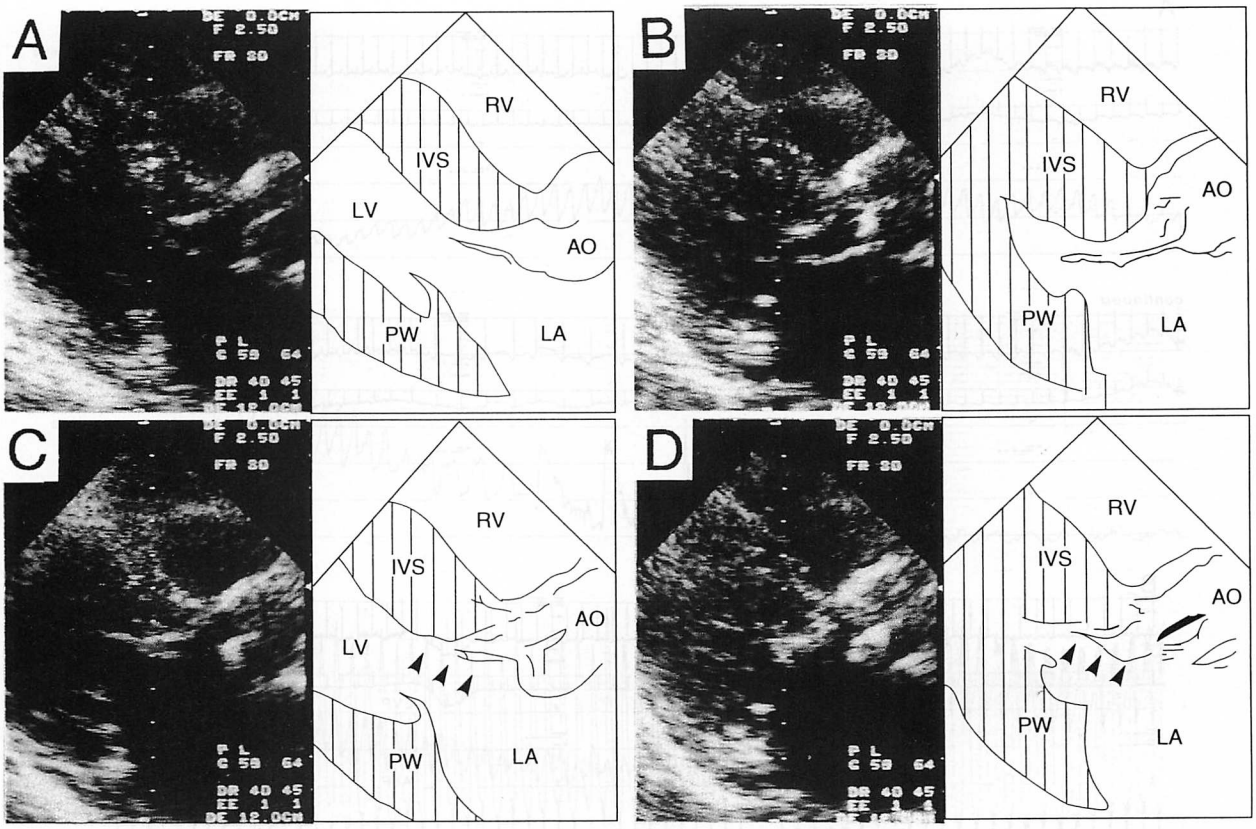

Fig. 2 Two-dimensional echocardiograms before (*A*, *B*) and during syncopal attack (*C*, *D*)

A, *B* : The baseline echocardiogram reveals asymmetric septal hypertrophy and systolic anterior motion of the anterior mitral valve.

C, *D* : During the attack, the anterior mitral leaflet became attached to the interventricular septum and transiently obstructed the left ventricular outflow tract.

A, *B*, *C* and *D* = parasternal long-axis view at end-diastole (*A*) and end-systole (*B*) at the baseline, and at end-diastole (*C*) and end-systole (*D*) during the attack (*C*, *D*). These echocardiograms are reproduced from videotape.

RV = right ventricle; LV = left ventricle; LA = left atrium; AO = ascending aorta; IVS = interventricular septum; PW = left ventricular posterior wall.

He underwent a cardiac catheterization study. During the catheterization study, syncope with convulsion developed repeatedly without antecedent cause, and was associated with a decrease in systemic blood pressure. Approximately 30 sec later, he recovered as previously with an increase in systemic blood pressure (**Fig. 3–A**). Simultaneous pressure monitoring of left ventricle and femoral artery showed a pressure gradient of more than 110 mmHg when he lost consciousness (**Fig. 3–C**). However, there was no pressure gradient observed while he was alert (**Fig. 3–B**). Coronary angiography revealed large arteries and no organic stenosis in any major branch.

Thus, we established a diagnosis of HOCM with recurrent syncope induced by decreased blood flow secondary to transient LVOFT narrowing. Propranolol (240 mg/day) and verapamil (120 mg/day) were administered, but there was no effect on the recurrence of attacks. Temporary atrioventricular sequential pacing was performed to prevent the reduction of blood flow through the LVOFT. There was no pressure gradient between the left ventricle and aorta in the catheterization study. Syncope did not recur after initiation of the pacing. He underwent mitral valve replacement 15 days later, and the postoperative course was uneventful. A cardiac catheterization study after mitral valve replacement showed the left ventricular-aortic pressure gradient was nearly absent at rest, but a slight pressure gradient of less than 40 mmHg was disclosed during the Valsalva maneuver, post-extrasystolic state, and after sublingual nitroglycerin administration. He has not experienced syncope for 4 years since dis-

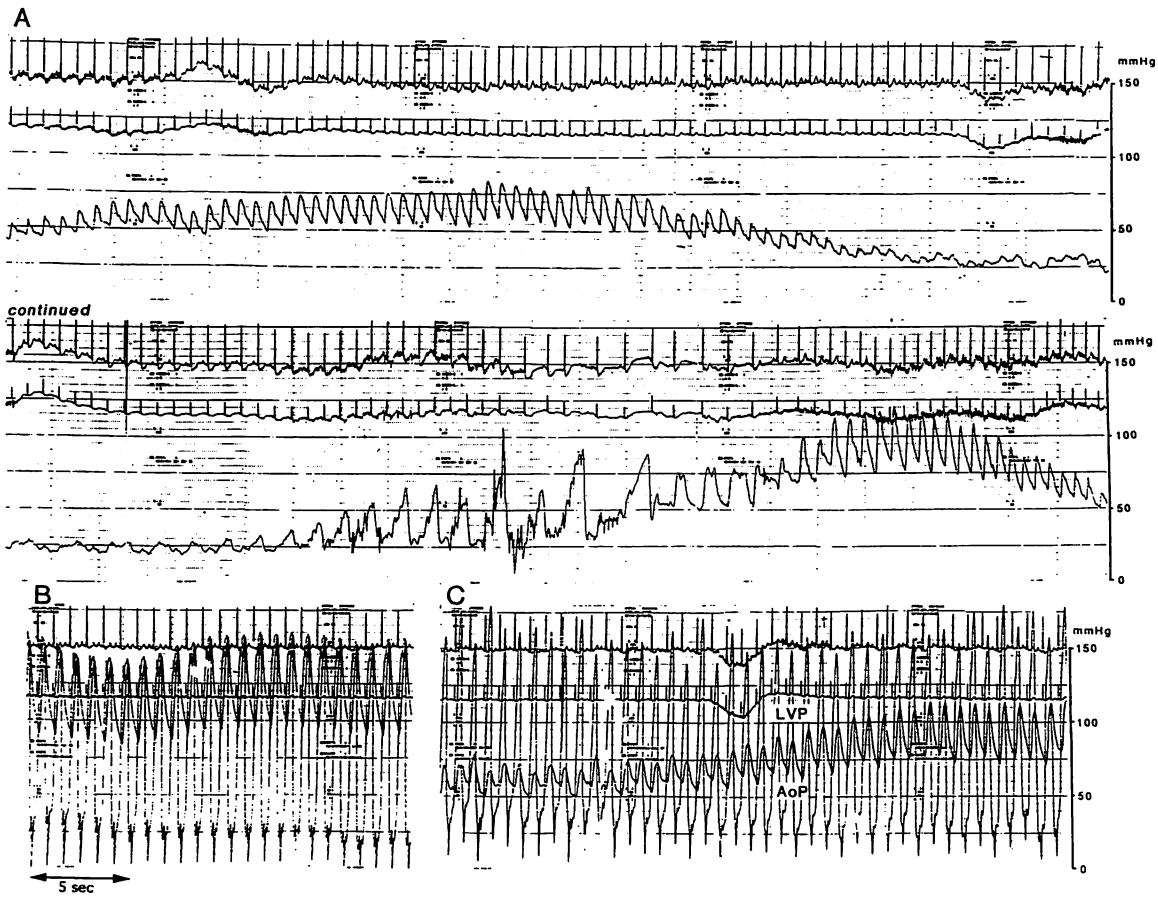

Fig. 3 Aortic pressure tracing during repeated syncopal attacks (*A*), and simultaneous left ventricular and aortic pressure tracings (*B*, *C*)

Systemic blood pressure recurrently decreased without preceding heart rate change. There was no pressure gradient between the left ventricle and femoral artery when he was asymptomatic (*B*), but a marked pressure gradient developed during the attack (*C*).
LVP=left ventricular pressure; AoP=right femoral arterial pressure.

charge.

DISCUSSION

This patient with HOCM had recurrent syncopal attacks induced by LVOFT obstruction. Systemic blood pressure falls as a result of decreased cardiac output due to LVOFT obstruction in HOCM[4]. However, no reports has described simultaneous pressure monitoring of left ventricular and systemic blood pressures during syncopal attack in HOCM.

Generally, LVOFT narrowing is activated due to increased left ventricular contractility or decreased pre- or afterload during exercise[4]. In the present patient, all attacks had developed at rest, not during exercise, and heart rate and blood pressure did not change before the attacks. These facts suggest that increased sympathetic tone and cathecholamine or activated vagal tone were not major causes of the attack. However, we could not clarify the precise etiology of LVOFT obstruction in this particular case. Activation of the ventricular baroreceptor reflex induced by decreased left ventricular volume and hypercontractility of the left ventricle might have caused the hemodynamic collapse. If inappropriate peripheral vasodilatation following stimulation of the left ventricular baroreceptors can cause abnormal systemic blood pressure response, such a mechanism may result in more serious events in HOCM[5].

The clinical course in the present patient demonstrates the sequence of LVOFT narrowing and provides many invaluable insights in the clinical profile of HOCM. First, ventricular arrhythmias secondary to global myocardial ischemia are commonly thought to be the usual mechanism of syncope or sudden cardiac death. However, no fatal arrhythmia

had developed during the syncopal episodes in this patient. This suggests that further research is necessary to elucidate the cause of sudden death both at rest or during sleep in HOCM. Second, atrioventricular sequential pacing effectively controlled the attacks before the mitral valve replacement, which implies that pacing could be an effective alternative approach in patients unresponsive to medical therapy[6]. We have no explanation for the mechanism of spontaneous recovery in the present patient. Some unknown compensatory mechanism may develop in response to the attacks.

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要　　約

左室流出路狭窄により失神発作を繰り返した
閉塞性肥大型心筋症の1例

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石川　正　　小岩屋　靖　　江藤　胤尚

症例は70歳，男性．繰り返す意識消失発作の精査のため入院した．

発作発生時の心電図に不整脈はなく，I，II，aV_L，aV_F，V_2–V_6誘導に陰性T波を伴うST低下を認めた．心エコー図上，非対称性中隔肥大および僧帽弁前尖収縮期前方運動による左室流出路狭窄と，高度の僧帽弁閉鎖不全を認めた．緊急心臓カテーテル検査中，痙攣を伴う失神発作が誘因なく繰り返し発生した．発作の出現時には大動脈圧が著明に低下し，左室–大動脈間に最高110 mmHgの圧較差が出現した．しかし，数分後には自然に血圧は上昇し，意識も回復した．以上より，閉塞性肥大型心筋症，および左室流出路狭窄に起因する意識消失発作と診断した．

失神発作の内科的コントロールは不能で，体外ペーシングにより小康を得た．Semielectiveに僧帽弁置換術を行い，以後4年間の経過は良好である．

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