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Sigmoid septum causing left ventricular outflow tract obstruction : A case report

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Summary

A 67-year-old man with a sigmoid septum causing the left ventricular outflow obstruction by inotropic stimulation was reported. This patient was admitted to the Hospital of the University of Tsukuba because of chest pain. Phonocardiography revealed a systolic ejection murmur which was intensified by amyl nitrite inhalation. A carotid pulse tracing showed a mid-systolic dip and a secondary slow wave during amyl nitrite inhalation. M-mode echocardiography demonstrated neither systolic anterior motion of the mitral valve (SAM) nor mid-systolic closure of the aortic valve at rest. Two-dimensional echocardiography revealed a basal interventricular septum markedly protruding into the left ventricle (sigmoid septum). The remainder of the septum and the left ventricular free wall were not hypertrophied, and no enlargement of the left ventricular cavity was observed. During exercise tests, blood pressure dropped significantly. Cardiac catheterization showed a pressure gradient within the left ventricle with isoproterenol infusion and post-extrasystolic potentiation.

These findings suggest that left ventricular outflow tract obstruction could occur in a patient with sigmoid septum by inotropic stimulation, producing a fall of blood pressure during exercise.

Key words

Sigmoid septum | Left ventricular outflow tract obstruction | Fall of blood pressure during exercise

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Introduction

A marked protrusion of the interventricular septum into the left ventricular cavity (" sigmoid septum ") was reported to be an exaggeration of the physiologic bending of the interventricular septum and to be brought about by physiologic decrease in cardiac output associated with aging and shrinking of the left ventricular cavity[1]. Though its pathophysiological significance has not been completely studied, it is generally thought that sigmoid septum does not cause the outflow tract obstruction of the left ventricle[1]. However, recent noninvasive study demonstrated that left ventricular outflow tract obstruction might occur in some patients with sigmoid septum[2]. The present report describes a patient with sigmoid septum showing a fall of blood pressure during exercise, in whom left heart catheterization study demonstrated a provoked pressure gradient across the left ventricular outflow tract.

CASE REPORT

History of Present Illness:

A 67-year-old man was admitted to the hospital for evaluation of an abnormal electrocardiogram and uncomfortable feeling of the chest in April, 1984. He had been under medical care elsewhere since 1980 for systemic hypertension; his blood pressure before treament was 190/90 mmHg. He had been suffering from general fatigue, easy fatigability and uncomfortable precordial feeling on exercise. He was diagnosed as ischemic heart disease and treated medically at another hospital, but there was no improvement in his complaints. Therefore, he was referred to our clinic for cardiac evaluation.

Past History:

Appendectomy at the age of 24.

Family History:

His mother and father died of heart disease, but the details were unknown.

On examination, the patient's height was 158 cm, and weight 49 kg. Blood pressure was 130/90 mmHg, and heart rate 60/min with regular rhythm. A grade 2/6 systolic ejection murmur was heard loudest at the apex. The abdomen was soft and flat, and no hepatomegaly was noticed. There was no peripheral edema in the extremities. Neurological examination revealed no significant abnormalities. Chest radiography showed no abnormalities. Cardiothoracic ratio was 43% (**Fig. 1**). Electrocardiogram showed regular sinus rhythm (heart rate=58), complete right bundle branch block and deep q waves in II, III, aVF, V_5 and V_6 (**Fig. 2**). These q waves were rgarded as insignificant, because the width was less than 0.04 sec. Two-dimensional echocardiograms showed marked thickening of the basal interventricular septum and its protrusion into the left ventricular cavity (sigmoid septum) (**Fig .3**). The angle formed by the aorta and the interventricular septum (aorto-septal angle) was about 90 degrees. No other portion of the left ventricular wall showed thickening, and the M-mode echocardiographic measurement disclosed that the thickness of the interventricular septum was 12 mm. Posterior wall thickness of the left ventricle was 12 mm, left ventricular end-diastolic dimension (EDD) was 54 mm, and end-

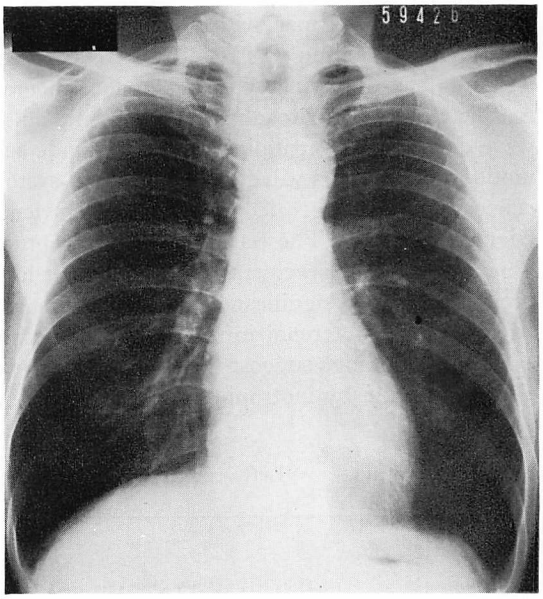


Fig. 1. Posteroanterior chest radiograph on admission.

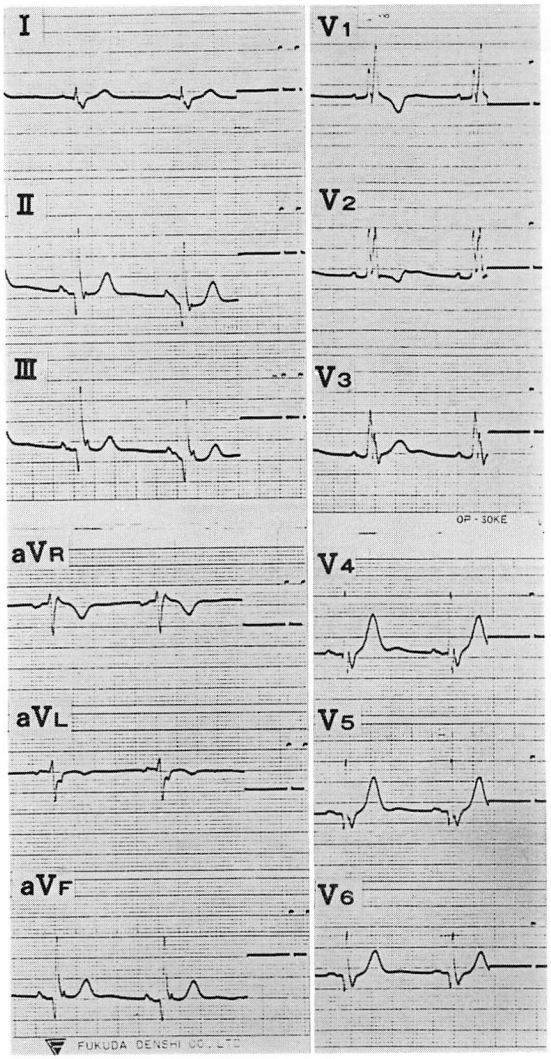
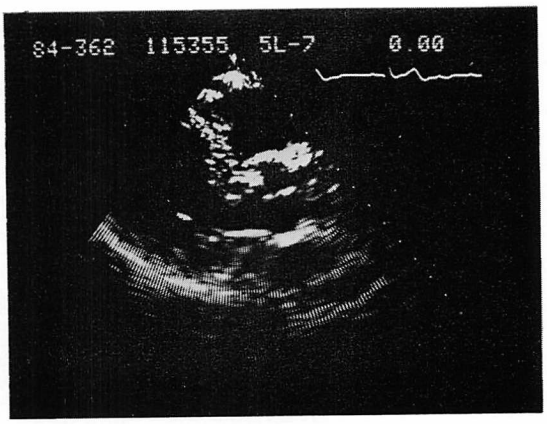


Fig. 2. Electrocardiogram on admission.

The electrocardiogram shows regular sinus rhythm (heart rate=58), complete right bundle branch block and deep q waves in II, III, aVF, V_5 and V_6.



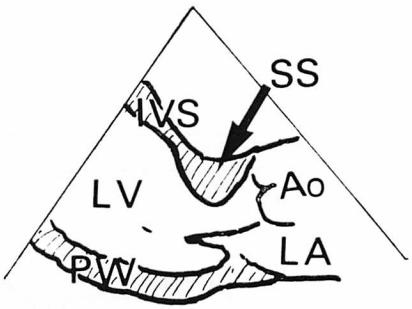


Fig. 3. Two-dimensional echocardiogram (long axis) **and schema.**

A marked protrusion of the upper portion of the interventricular septum (sigmoid septum: SS) (black arrow) is seen in the diastolic frame.

IVS=interventricular septum; SS=sigmoid septum; LV=left ventricle; PW=posterior wall of the left ventricle; Ao=aorta; LA=left atrium.

systolic dimension (ESD) was 30 mm. Fractional shortening [(EDD−ESD)/EDD×100] was 44%. These echocardiographic parameters were nearly within normal limits. Systolic anterior motion of the mitral valve (SAM) was not demonstrated by echocardiography (**Fig. 4 A**). Late systolic semi-closure of the aortic valve was revealed, though mid-systolic semi-closure was not (**Fig. 4 B**). Phonocardiograms demonstrated a systolic ejection murmur at the apex. After amyl nitrite inhalation, the systolic murmur was intensified and the carotid pulse recording showed a mid-systolic dip and a slow secondary wave (**Fig. 5**). Heart rate and blood pressure responses during Master's two-step, treadmill and ergometer exercise tests are shown in **Table 1.** Blood pressure fall was seen on exercise, especially during Master two-step test from 196/108 mmHg to 100/70 mmHg, though heart rate was increased by exercise.

Cardiac catheterization performed while the

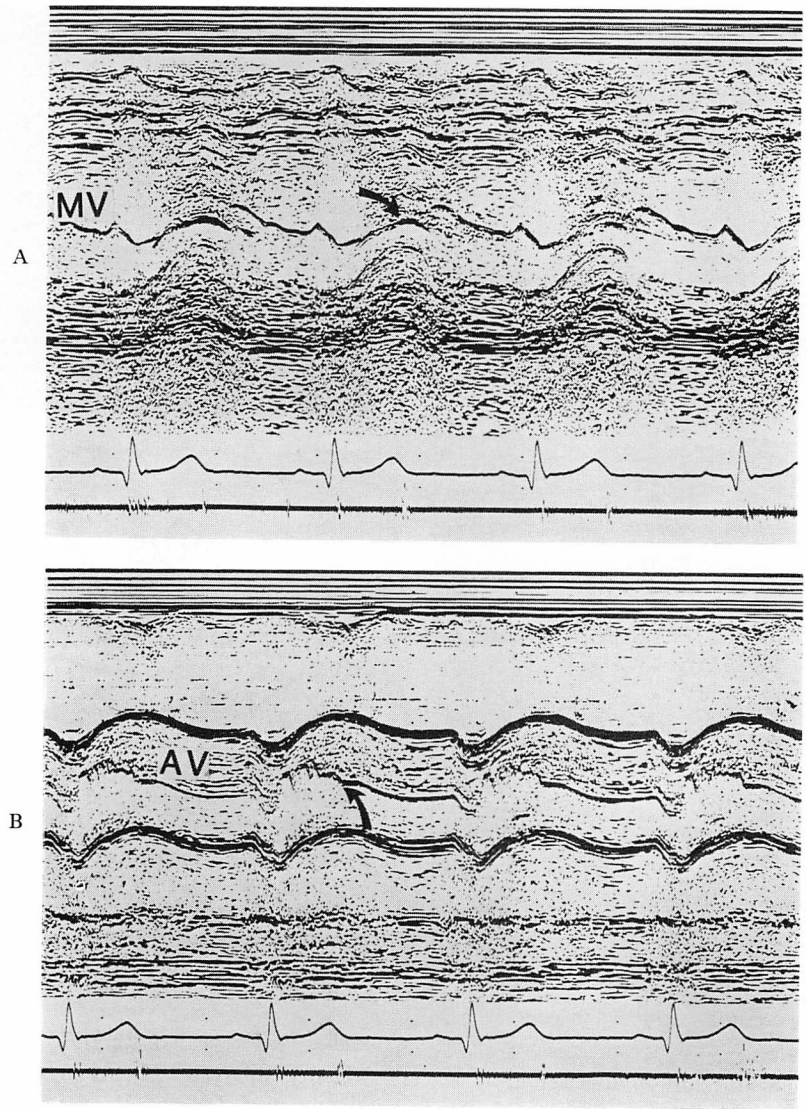


Fig. 4. M-mode echocardiograms.

A: Systolic anterior motion of the mitral valve is not clear at rest (black arrow).

B: Late systolic semi-closure of the aortic valve is shown, though mid-systolic semi-closure is not (black arrow).

patient was taking no cardioactive medication demonstrated no significant abnormal finding at rest (**Table 2**). However, with post-extrasystolic potentiation, pulse pressure (50 mmHg) was smaller than in the control beat (60 mmHg) and a pressure gradient (more than 60 mmHg) was found between the left ventricle (more than 160 mmHg) and the aorta (100 mmHg) (**Fig. 6**). After intravenous infusion of isoproterenol a pressure gradient was provoked between the left ventricular outflow tract (100 mmHg) and the left ventricular apex (130 mmHg) (**Fig. 7**).

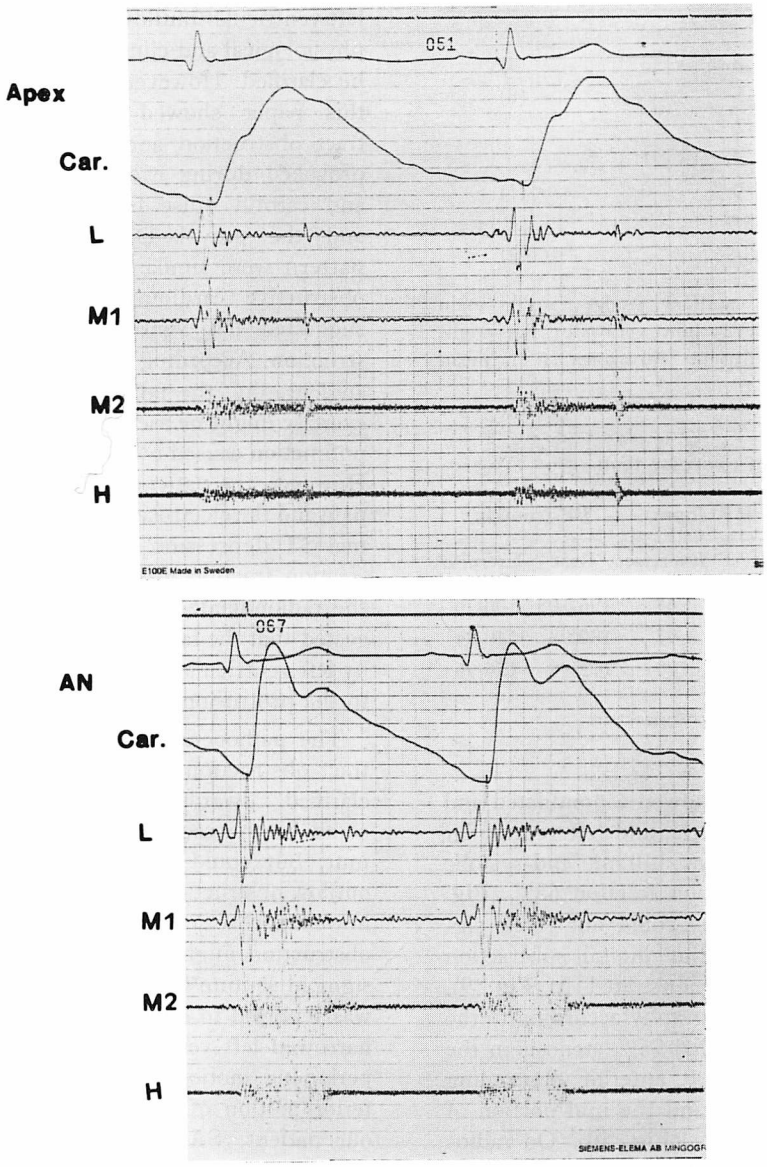


Fig. 5. Phonocardiograms and carotid pulse recordings before and after amyl nitrite inhalation.

Before amyl nitrite inhalation, the tracing shows a systolic ejection murmur at the apex and nearly normal carotid pulse. Following the inhalation, the systolic murmur is intensified and the carotid pulse tracing shows a mid-systolic dip and a slow secondary wave (spike and dome).

Table 1. Catheterization data

			(mmHg)
PCW	a 5	v 4	
	x 3	y 3	(4)
mPA	17/5		(8)
RV	20/2		
RA	a 2	v 1	
	x 0	y 0	(1)
LV	135/7		
Ao	128/76		(102)

PCW=pulmonary capillary wedge; mPA=main pulmonary artery; RV=right ventricle; RA=right atrium; LV=left ventricle; Ao=aorta. ( )=mean pressures.

Table 2. Results of exercise tests

	HR (bpm)		BP (mmHg)	
	Rest	Exercise	Rest	Exercise
Master (double)	55	65	196/108	100/70
Treadmill	80	97	180/98	140/84
Ergometer	62	100	172/98	156/96

HR=heart rate; BP=blood pressure.

Left ventriculography showed a nondilated and nonhypertrophied ventricle, and left ventricular end-diastolic volume was 140 ml, end-systolic volume 45 ml and ejection fraction 68% (**Fig. 8**). Left ventriculography could not reveal sigmoid septum clearly, but digital subtraction angiography clearly demonstrated it (**Fig. 9**). Selective coronary angiography according to the Judkins technique revealed 75% stenosis in the distal portion of the left anterior descending coronary artery (LAD) and the mid-portion of the right coronary artery (**Fig. 10**). On radionuclide angiograms, ejection fraction was 78% at rest which increased to 92% with exercise.

DISCUSSION

Sigmoid septum has been reported to be an exaggeration of the physiologic process of aging, and sclerotic and fibrous changes in the aorta and the interventricular septum were suggested as its causes[1,3]. The sigmoid septum has been believed not to cause obstruction to the left ventricular outflow tract[1], though its pathophysiological and clinical significance remains to be clarified. However, the patient described in this paper showed left ventricular outflow tract obstruction and a fall of blood pressure provoked during exercise. Phonocardiographic and carotid pulse findings after amyl nitrite and the provoked left ventricular pressure pattern were similar to those of hypertrophic obstructive cardiomyopathy (HOCM)[4,5] and suggested left ventricular outflow tract obstruction. According to the findings of coronary angiography, the fall of blood pressure during exercise might be ascribed to left ventricular hypofunction caused by exercise-induced ischemia. However, the patient did not complain of chest pain and the electrocardiogram showed no abnormal ST depression during exercise. Moreover, ejection fraction was increased by exercise on the radionuclide angiogram. These results suggested that the fall of blood pressure was caused by left ventricular outflow tract obstruction due to the protrusion of sigmoid septum.

The pathogenesis of left ventricular outflow tract obstruction in this patient has not been clarified. Edwards suggested that sigmoid septum could be the cause of functional systolic murmur, but could not result in left ventricular outflow obstruction[1]. Recent noninvasive study, however, revealed that left ventricular outflow obstruction might occur in some patients with sigmoid septum[2]. In that study the obstruction was reported to be ascribed to sigmoid septum, narrowed left ventricle and left ventricular hyperkinetic motion associated with systolic anterior motion of the mitral valve (SAM)[2]. In our patient, SAM was not recorded on the echocardiogram and left ventriculography did not show the narrowed left ventricle and hyperkinesis at rest. Unfortunately, the echocardiogram and left ventriculogram on provocation (isoproterenol infusion and post extrasystolic potentiation) were not obtained. However, left ventricular hyperkinesis and SAM might be induced by provocation, because radionuclide angiography

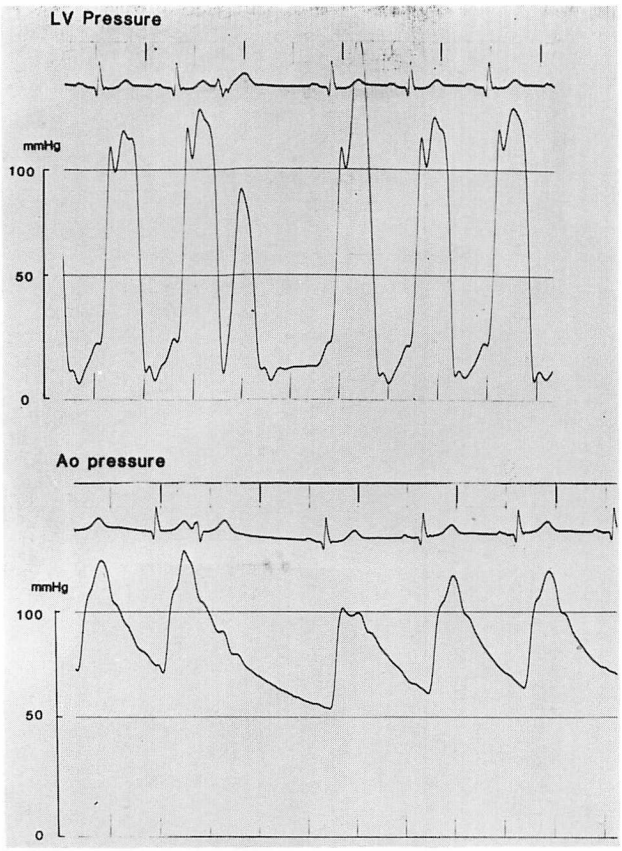


Fig. 6. Aortic and left ventricular pressure tracings.
With a post-extrasystolic potentiation, pulse pressure is decreased and a pressure gradient between the aorta and the left ventricle is noted.

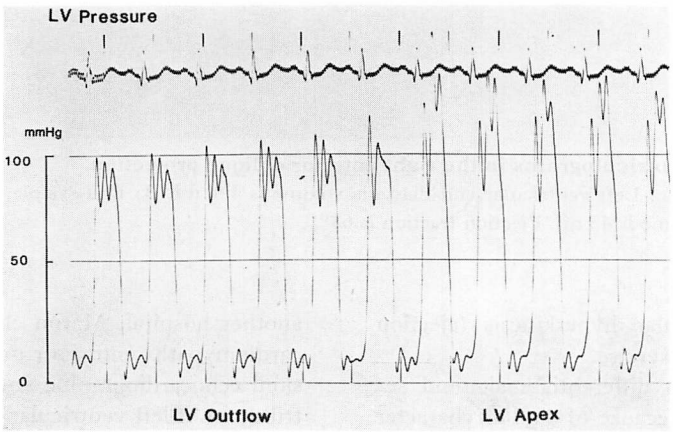


Fig. 7. Left ventricular pressure showing a developed pressure gradient.
After isoproterenol infusion, a pressure gradient (more than 60 mmHg) is provoked.

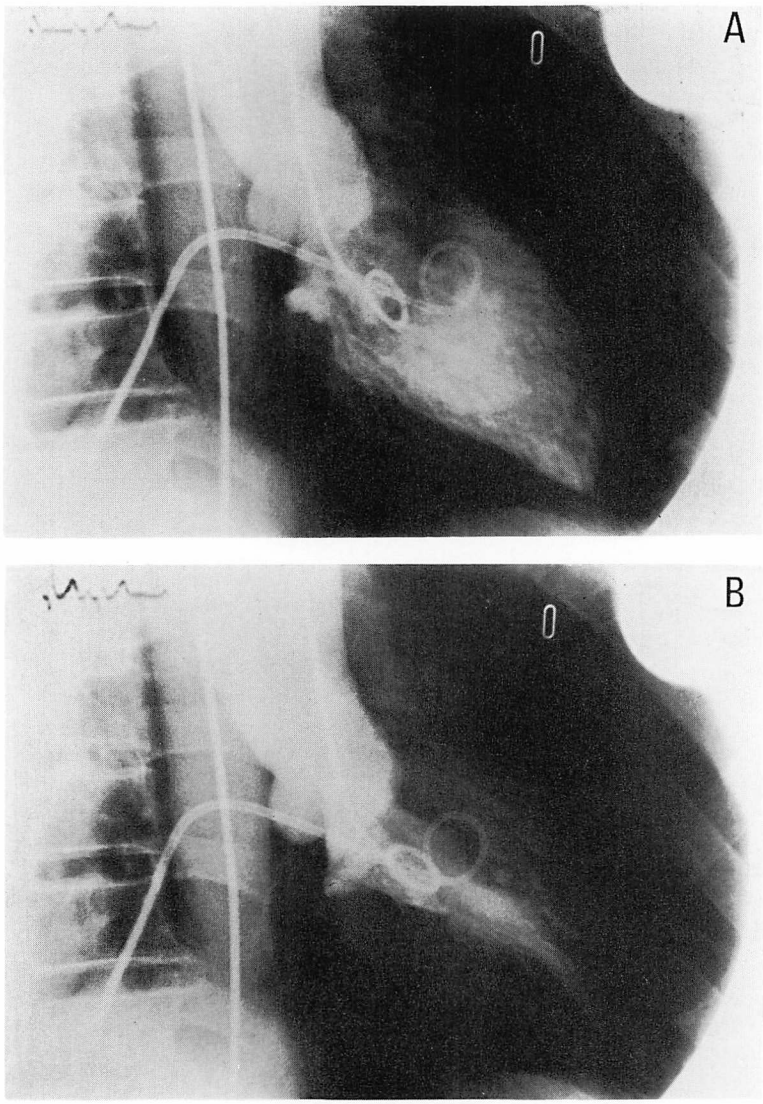


Fig. 8. Left ventriculograms in the right anterior oblique projection.
A: End-diastole. Left ventricular end-diastolic volume is 140 m*l*. B: End-systole. Left ventricular end-systolic volume is 45 m*l*. Ejection fraction is 68%.

showed left ventricular hyperkinesis (ejection fraction=92%) on exercise.

It is important to differentiate sigmoid setum from HOCM, because of similar character of the systolic murmur. Actually, our patient has been misdiagnosed as having HOCM at another hospital. Maron classified hypertrophic cardiomypathy into four groups by two-dimensionl echocardiographic study according to distribution of left ventricular hypertrophy[6] : long-axis echocardiogram of patients of group I showed ventricular septal hypertrophy confined

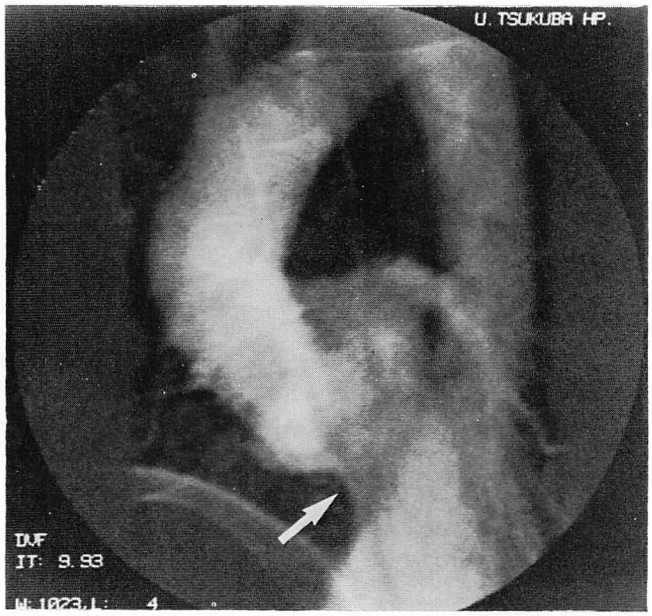


Fig. 9. Digital subtraction angiogram of the left ventricle in the left anterior oblique projection.

Sigmoid septum is clearly demonstrated (white arrow).

to the most cranial portion of the anterior septum producing the appearance of a prominent and localized bump, which resembled sigmoid septum, but the aorto-septal angle was not so acute as sigmoid septum. Outflow obstruction was revealed in two of 12 patients of group I. Therefore, there remained question about differentiation of the two diseases. Aorto-septal angle was reported to be decreased in patients with sigmoid septum compared with those with hypertrophic cardiomyopathy[3]. The angle of our patient was acute, which corresponded with the previous study[3]. Thus, aorto-septal angle may be expected to play a certain role to differentiate the two diseases.

Sudden death often occurs during exercise in patients with HOCM[4]. Sudden development of a pressure gradient is thought the cause of sudden death[4]. As our patient showed a similar pathophysiological phenomenon, sigmoid septum could play some causal roles in sudden death of the aged. Thus, sigmoid septum should not be thought of as physiologic phenomenon associated with aging and its clinical significance should be investigated as a new clinical entity.

左室流出路狭窄を有する S 字状中隔の 1 例

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松本龍馬，富沢巧治，野口祐一，
行定公彦，小川　剛，伊藤　巌

67 歳，男性における S 字状中隔例の非観血的ならびに心臓カテーテル検査の結果を報告した．患者は胸痛を主訴に筑波大学付属病院に入院した．心音図では亜硝酸アミルにて増強する収縮期雑音，頸動脈波では亜硝酸アミル吸入により出現する mid-systolic dip ならびに secondary slow wave を認めた．M モード心エコー図では SAM や大動脈弁の収縮中期半閉鎖を認めなかった．断層心エコー図では心基部心室中隔の左室内腔への

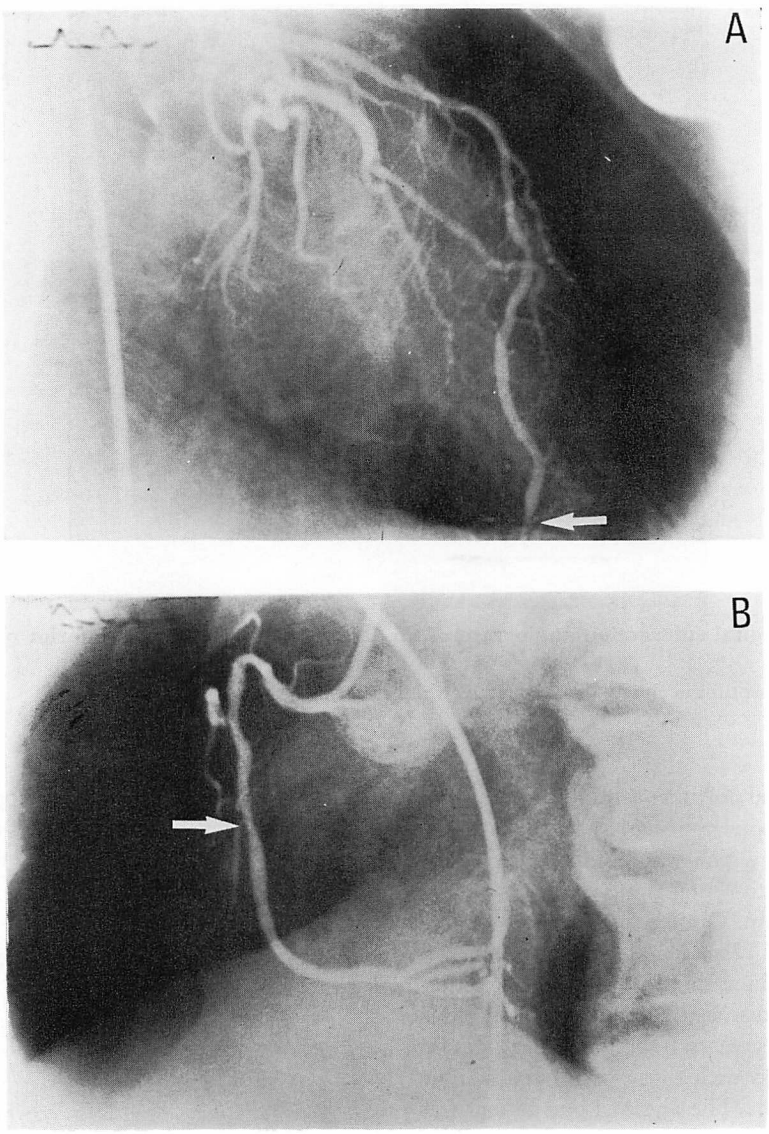

Fig. 10. Selective coronary angiograms.
A: Left coronary artery (LCA). B: Right coronary artery (RCA). Significant stenosis is seen in the distal portion of the LCA (white arrow) and the mid-portion of the RCA (white arrow).

著明な突出 (S 字状中隔) を認めたが，心基部以外の心室中隔ならびに左室自由壁に肥大を認めず，左室拡張もなかった．運動時血圧は著明に低下し，心臓カテーテル検査にてイソプロテレノールにて増強する左室内圧較差，および Brockenbrough 現象を認めた．

以上より S 字状中隔では，陽変力刺激により左室流出路狭窄が生じ，運動時に血圧が低下する症例が存在することが明らかとなった．

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