

**IDIOPATHIC HYPERTROPHIC SUBAORTIC STENOSIS**  
**A CASE REPORT CENTERING ON FINDINGS BY ULTRASOUND CARDIOGRAPHY**

特発性肥大性大動脈弁下狭窄症の1症例  
超音波心臓検査所見を中心として

CHOW HOW LIN, M.D. 林朝河  
HIROSHI FURONAKA, M.D. 風呂中弘  
KAZUNORI KODAMA, M.D. 児玉和紀  
THOMAS L. ROBERTSON, M.D.  
KAZUMI TAGUCHI, M.D. 田口一美



**RADIATION EFFECTS RESEARCH FOUNDATION**  
財団法人放射線影響研究所

A cooperative Japan - United States Research Organization  
日米共同研究機関

RERF TECHNICAL REPORT SERIES

放影研業績報告書集

The RERF Technical Reports provide the official bilingual statements required to meet the needs of Japanese and American staff members, consultants, and advisory groups. The Technical Report Series is in no way intended to supplant regular journal publication.

放影研業績報告書は、日米専門職員、顧問、諮問機関の要求に応えるための日英両語による公式報告記録である。業績報告書は決して通例の誌上発表論文に代わるものではない。

---

*The Radiation Effects Research Foundation (formerly ABCC) was established in April 1975 as a private nonprofit Japanese Foundation, supported equally by the Government of Japan through the Ministry of Health and Welfare, and the Government of the United States through the National Academy of Sciences under contract with the Department of Energy.*

放射線影響研究所(元ABCC)は、昭和50年4月1日に公益法人として発足した。その経費は日米両政府の平等分担とし、日本は厚生省の補助金、米国はエネルギー省との契約に基づく米国学士院の補助金とをもって充てる。



**IDIOPATHIC HYPERTROPHIC SUBAORTIC STENOSIS**  
**A CASE REPORT CENTERING ON FINDINGS BY ULTRASOUND CARDIOGRAPHY**

特発性肥大性大動脈弁下狭窄症の1症例

超音波心臓検査所見を中心として

CHOW HOW LIN, M.D. (林朝河); HIROSHI FURONAKA, M.D. (風呂中弘);  
 KAZUNORI KODAMA, M.D. (児玉和紀); THOMAS L. ROBERTSON, M.D.;  
 KAZUMI TAGUCHI, M.D. (田口一美)\*

*Department of Medicine*

臨床部

**SUMMARY**

A case (male aged 16) presenting distinctive echo findings on ultrasound cardiography is described with mention of findings following surgical treatment. Apexcardiography showed a characteristic "a" wave and the carotid pulse tracing showed a markedly bisferious ejection wave. On cardiac auscultation and phonocardiography, systolic ejection murmurs were found which were loudest at the apex, where S<sub>3</sub> and S<sub>4</sub> were also audible.

On ultrasound cardiography characteristic findings of idiopathic hypertrophic subaortic stenosis were found; these included systolic deviation of the mitral valve, causing stenosis of the left ventricular outflow tract, early systolic closure of the aortic valve, and a decreased rate of diastolic descent of the mitral valve which reflected hypofunction of the left ventricular myocardium and hypertrophy of the ventricular septum.

Postoperatively, the following were found: improvement of subjective symptoms, and on ultrasound cardiography less systolic anterior deviation of the mitral valve during systole, less thickening of the ventricular septum, and a faster rate of diastolic descent of the mitral valve.

In this case, ultrasound cardiography was found to be the most reliable and easy method for

**要 約**

超音波心臓記録検査(UCG)で特徴的なエコー所見を呈した特発性肥大性大動脈弁下狭窄症の1例(16歳の男性)について記述し、さらに外科的療法による所見にも言及した。心尖拍動図では、特徴的な"a"波を認め、頸動脈波では顕著な二峰性の駆出波を認めた。心臓聴診および心音図では、心尖部を最強点とする収縮期駆出性雑音と同部におけるⅢ、Ⅳ音を認めた。

UCGでは、特発性肥大性大動脈弁下狭窄症の特徴的な所見が認められた。すなわち、左室流出路の狭窄の原因である僧帽弁の収縮期偏位、大動脈弁の早期収縮期閉鎖、ならびに左室心筋機能低下および心室中隔の肥大を反映する僧帽弁前尖拡張期後退速度(DDR)の低下等が特徴的な所見として認められた。

術後所見として次が認められた: すなわち、自覚症の改善、ならびにUCGでは、僧帽弁の収縮期前偏位の減少、心室中隔の肥厚の減少、および僧帽弁前尖拡張期後退速度の増加である。

本症例においては、UCGが最も確実かつ容易な臨床診断法であり、また術後の状態の判断にきわめて有

\*Department of Surgery, Hiroshima University School of Medicine

広島大学医学部外科学第一講座

clinical diagnosis and very useful as a noninvasive method for evaluating the postoperative course.

## INTRODUCTION

Idiopathic hypertrophic subaortic stenosis (IHSS) is a disease which as a result of asymmetric hypertrophy of either the ventricular septum or the left ventricular wall, is characterized by narrowing of the left ventricle and stenosis of left ventricular outflow tract which is aggravated by cardiac contraction.<sup>1</sup> Heretofore, left ventricular catheterization and angiocardiology have been considered necessary for definitive clinical diagnosis of this disease.<sup>2,3</sup> With the availability in recent years of noninvasive cardiac examination methods, especially ultrasound cardiography (UCG) for detection of distinctive ecographic images,<sup>4</sup> clinical diagnosis of this disease has been simplified. Recently the authors encountered a case of this disease which was diagnosed by UCG and confirmed at the time of surgical treatment. The therapeutic outcome was good.

## CASE REPORT (MEZ ████████)

Male, aged 16, senior high school student. Chief complaint was dyspnea on exertion.

**Family History.** Mother died at age 30 because of atrial septal defect. Otherwise unremarkable.

**Past History.** Unremarkable.

**History of Present Illness.** The subject was the product of a normal gestation and delivery, and no heart disease had ever been noted. At a periodic physical examination at school in the Spring of 1974, a cardiac murmur was described for the first time. Because an electrocardiographic (ECG) examination was abnormal, the patient visited RERF in Hiroshima for detailed physical examination. The only subjective symptom he complained of was dyspnea on exertion. He had never lost consciousness nor had chest pains.

**Physical Examination.** Nourishment and physical build were medium (height 170.8 cm, weight 50 kg). No ocular or palpebral abnormality was present. No abnormality such as engorgement of jugular veins, cyanosis, or deformity of the thorax was noted. The area of cardiac dullness was normal. On cardiac auscultation, moderate

な非観血的検査法であることが認められた。

## 緒言

特発性肥大性大動脈弁下狭窄症、すなわち Idiopathic hypertrophic subaortic stenosis (以下 IHSS と略記) は心室中隔あるいは左心室壁の非対称性肥大により、左室内腔の狭小化および左室流出路の狭窄が発生し、心収縮によってさらに狭窄の度が増強する状態を示す疾患である。<sup>1</sup> 本症に対する確定臨床診断には従来より、左心系カテーテル検査および心血管造影法が必要であるとされてきた。<sup>2,3</sup> 近年普及してきた非観血的心臓検査法、特に超音波検査法、すなわち Ultrasound cardiography (以下 UCG と略記) によっても特徴的なエコー波型<sup>4</sup> を認めることにより、臨床診断が容易かつ可能となってきた。最近著者らは、UCG によって本症を診断し、外科的治療によって診断を確認し、また治療効果も十分であった。

## 症例 (MEZ ████████)

男性、16歳、高校生。主訴：運動時の呼吸困難。

**家族歴：** 母親が30歳で心房中隔欠損症により死亡しているほか、特記すべき事項を認めない。

**既往歴：** 特記すべき事項を認めない。

**現病歴：** 満期安産・生来健康で、心疾患を指摘されたことはなかったが、昭和49年の春に実施された学校の定期検診により、初めて心雑音を指摘された。心電図所見にも異常が認められたので、精密検査の目的で放影研(広島)で検診を受けた。自覚的に運動時の呼吸困難を訴えるのみで、失神ならびに胸痛などの発作は経験していない。

**全身検査：** 栄養、体格は中等度(身長170.8cm, 体重50kg)。眼球および眼瞼に異常はない。頸静脈の怒張、チアノーゼ、胸郭の変形なども認められない。心濁音界は正常である。心の聴診において、I音の中等

TABLE 1 CLINICAL FINDINGS AT INITIAL EXAMINATION

表 1 初診時臨床検査成績

Urinalysis . . . . .	No remarkable findings	<b>Biochemistry Tests</b>	
Stool . . . . .	No remarkable findings	GOT	27 Unit
Blood Sedimentation Rate . . . . .	2 mm/1 hr	GPT	12 Unit
	3 mm/2 hrs	LDH	118 Unit
<b>Cell Counts</b>		Alkaline phosphatase	10 Unit
Erythrocytes	562×10 <sup>4</sup>	LAP	180 Unit
Hemoglobin	14.8 g/100ml	TTT	1 Unit
Hematocrit	40%	ZnTT	3 Unit
Leukocytes	4000	BUN	15 mg/100ml
Differential	No remarkable findings	Creatinine	1.9 mg/100ml
<b>Biochemistry Tests</b>		<b>Serum Electrolytes</b>	
Total Serum Proteins	7.0 g/100ml	Na	140 mEq/L
<b>Serum Protein Fractions</b>		K	4.7 mEq/L
Albumin	56.5%	Cl	99 mEq/L
α <sub>1</sub> -globulin	2.3%	Ca	1.2 mEq/L
α <sub>2</sub> -globulin	11.3%	<b>Serological Tests</b>	
β-globulin	13.0%	ASLO	<200 Unit
γ-globulin	16.9%	CRP	Negative
Total Bilirubin	0.8 g/100ml	Wasserman's reaction	Negative
Direct Bilirubin	0.2 g/100ml		

accentuation of S<sub>1</sub> and respiratory splitting of S<sub>2</sub> were found, and S<sub>3</sub> and S<sub>4</sub> were audible at the apex. A Levine 4/6 ejection systolic murmur was most intense at the apex, and it did not radiate to the neck. Further, an early diastolic murmur of short duration was audible from the 4th intercostal space at the left sternal edge toward the apex. No abnormality of the lungs was noted either on auscultation or palpation. No abdominal abnormality was found. The extremities showed no edema or clubbing. No abnormalities of the tendon reflexes were noted.

**Clinical Findings at Initial Examination.** No abnormalities were found in the urine and stool at initial examination. Hematological examinations showed no specific abnormality (Table 1).

**Chest X-Ray.** There was no enlargement or deformity of the cardiac shadow nor enlargement of the aorta, and the lung fields appeared normal (Figure 1).

**ECG Findings.** The 12-lead ECG examination (Figure 2) revealed normal sinus rhythm but marked deviation of axis to the right. The PQ, QRS, and QT intervals were all within normal

度の亢進およびⅡ音の呼吸性分裂を認め、心尖部ではⅢ、Ⅳ音が聴取された。また心尖部に最強点を有する Levine 4/6度の駆出性収縮期雑音を聴くが頸部には放散していない。さらに第4肋間胸骨左縁から心尖部に向かって短い早期拡張期雑音を聴取した。肺では聴、打診ともに異常なく、腹部においても異常を認めない。四肢に浮腫、爪の変形なく、腱反射にも異常を認めなかった。

**初診時臨床検査成績：** 初診時における尿、糞便の検査に異常を認めない。血液検査成績は表1に示すごとく、特定の異常を認めなかった。

**胸部X線像：** 胸部X線撮影は図1に示すごとく、心陰影の拡大、変形ならびに大動脈の拡大はなく、また肺野にも異常の所見を認めない。

**心電図所見：** 12誘導の心電図は図2に示すごとく、正常の洞調律を示すが、顕著な右軸偏位を示している。PQ, QRS, QT間隔はいずれも正常の範囲内で

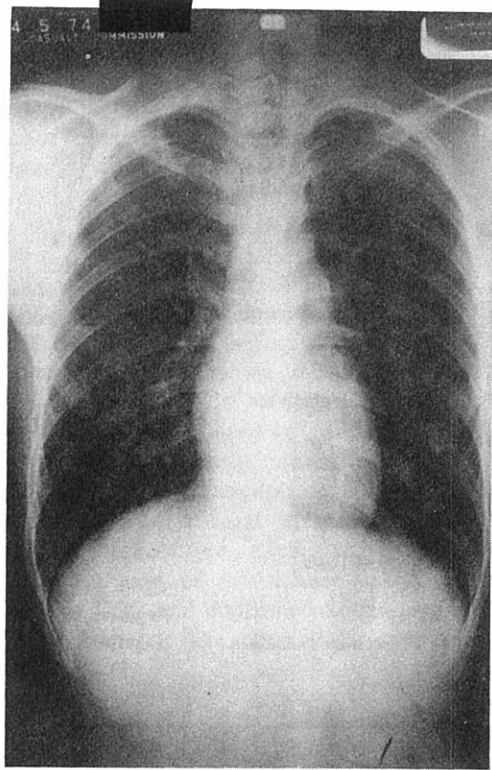


Figure 1 Frontal chest X-ray  
図 1 胸部X線像(正面)

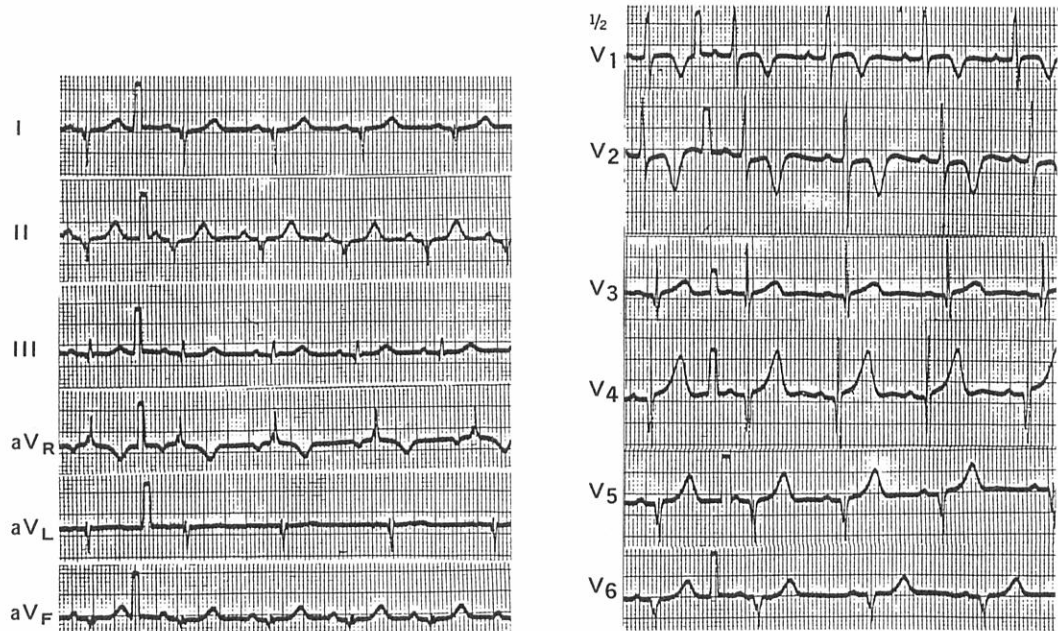


Figure 2 Preoperative electrocardiogram  
図 2 術前心電図

range, but a QS pattern was noted in leads I, II, V<sub>5</sub> and V<sub>6</sub>. Furthermore, there were findings suggestive of right ventricular hypertrophy with the R/S in V<sub>1</sub> in excess of 1 and deeply negative T waves in V<sub>1</sub> and V<sub>2</sub>.

**Cardiographic Tests.** The phonocardiographic findings were consistent with the findings at auscultation; S<sub>3</sub> and S<sub>4</sub> were recorded at the apex. A midsystolic ejection murmur and early diastolic murmur of short duration were also noted. The apex cardiogram showed elevation of the "a" wave and late systolic expansion, while the carotid pulse tracing showed a steep rise of ejection waves with bisferious or slow secondary waves during the ejection period (Figure 3).

**UCG Findings.** When the transducer was angled posteriorly from the 4th intercostal space at the left edge of the sternum, an M-mode scan showed the echo waves reaching the left ventricular posterior wall via the chest wall, right ventricle, ventricular septum, and anterior mitral cusp (Figures 4 and 5). The IV septal wall thickness was 2.0 cm, showing a marked thickening. Study of anterior mitral cusp movement revealed that the cusp moved close to or came into contact with the ventricular septum at the same time that the peak of ejection sound was reached on the cardiophonogram, forming a systolic hump and presenting a picture of narrowing of the left ventricular outflow tract. The LV posterior thickness was 1.0 cm, and marked posterior motion of the LV posterior wall was noted during early systole. The diastolic descent rate (DDR) had dropped markedly to 30 mm/sec. Further, when the transducer was placed on the left edge of the 3rd intercostal sternum and angled very slightly upwards, echoes of the aortic lumen and aortic valve were recorded as shown in Figure 6. The dimension of the aortic lumen was 2.8 cm, which is within normal range, but on phonocardiography at the time the aortic valve was open, a picture of transient semiocclusion of the aortic valve which was consistent with the peak of the ejection murmur was noted.

Based on the above distinctive findings on these noninvasive examinations, the case was diagnosed as IHSS. On the assumption that surgical treatment was necessary, left ventricular catheterization, a traumatic examination, and angiocardiology were made at the First Department of Surgery, Hiroshima University School of Medicine.

あるが、I、IIおよびV<sub>5</sub>、V<sub>6</sub>の各誘導にQS型の波型を認める。またV<sub>1</sub>におけるR/Sは1を超えており、V<sub>1</sub>、V<sub>2</sub>には深い陰性T波を認め、右室肥大を示唆する所見が認められた。

**心機図：**心音図所見は聴診所見と一致し、心尖部における記録ではⅢ音ならびにⅣ音を認める。また駆出性収縮中期雑音と短い早期拡張期雑音とを認める。心尖拍動図では"a"波の増高と収縮後期の膨隆(late systolic expansion)を認め、頸動脈波においては駆出波の急峻な立ち上がりと二峰性の波型、すなわち駆出期にslow secondary waveを認めた(図3)。

**UCGの結果：**探触子を胸骨左縁第4肋間より後方に向けることにより、図4、5に示すごとく、M-mode scanでは、胸壁、右室、心室中隔、僧帽弁前尖を経て、左室後壁に達するエコー波が認められた。心室中隔の厚さ(IV septal wall thickness)は2.0cmを示し、著明な肥厚が認められた。僧帽弁前尖の動きを見ると、心音図における駆出音のpeakの時期に一致して前尖は心室中隔に近づくか、または接するようになり、systolic humpを形成して左室の流出路が狭小化する像を認めた。左室自由壁の厚さ(LV posterior thickness)は1.0cmを示し、自由壁の早期収縮期における著明なposterior motionが認められた。拡張期僧帽弁前尖後退速度(DDR)は30mm/secと著明に低下していた。さらに探触子を第3肋間胸骨左縁に置き、ごくわずかに内上方に向けると、図6に示すごとく大動脈腔ならびに大動脈弁のエコーが記録された。大動脈腔のdimensionは2.8cmを示し正常範囲内であったが、大動脈弁の開放時に心音図の駆出性雑音のpeakに一致して、大動脈弁が一過性に半閉鎖状態となる像を認めた。

以上のように非観血的検査法による特徴的な所見に基づき、IHSSと診断し、外科的治療を前提として観血的検査法である左心カテーテル検査および心血管造影法が広島大学医学部第1外科において実施された。

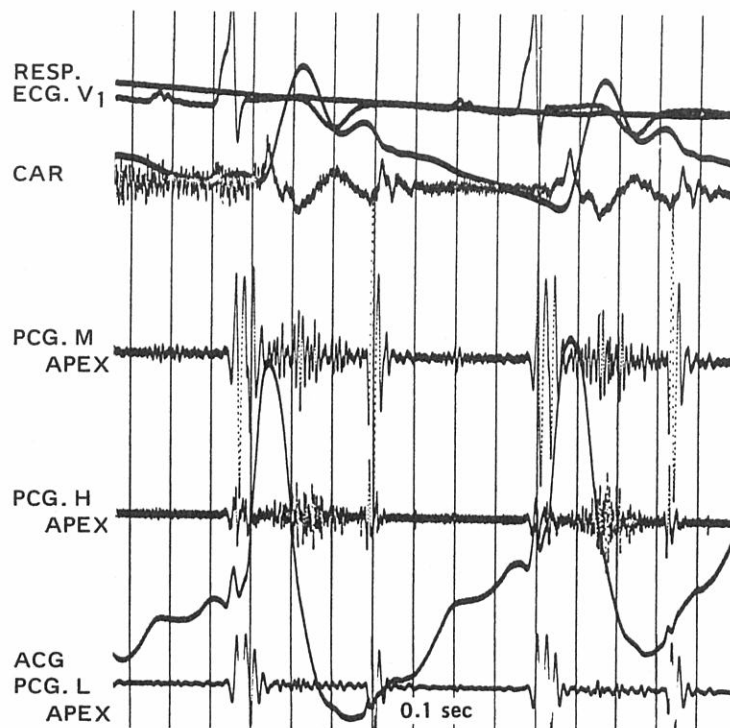


Figure 3 Cardiac function tests

图 3 心機能検査図

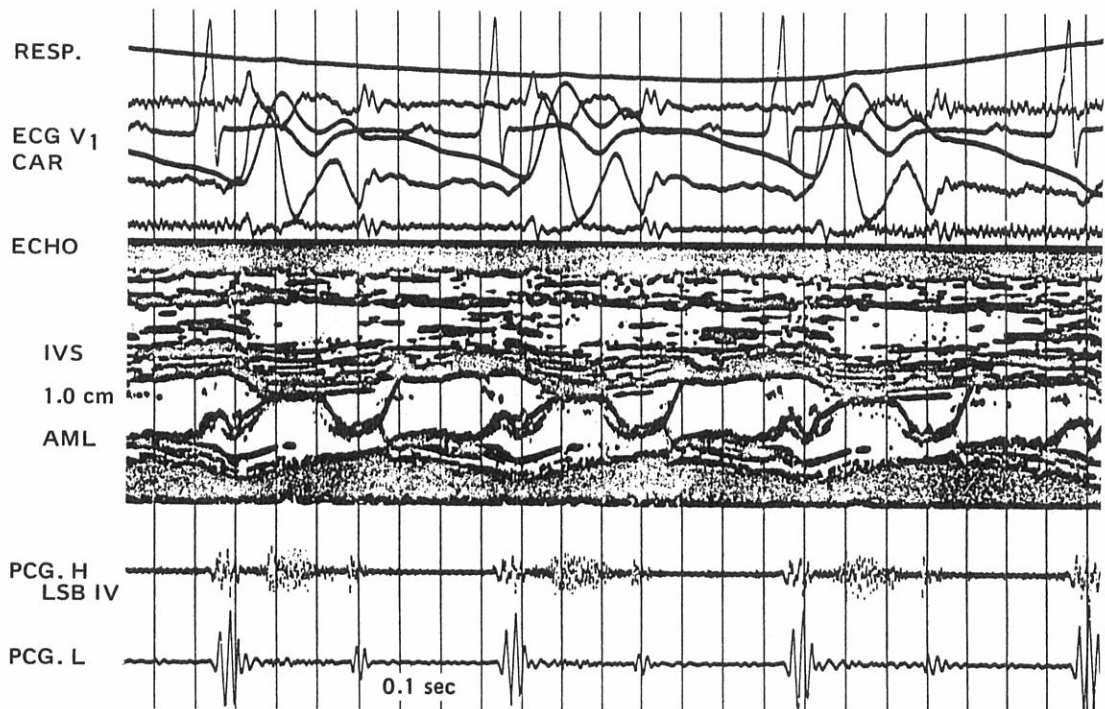


Figure 4 Ultrasound cardiogram

图 4 超音波心臟記錄圖



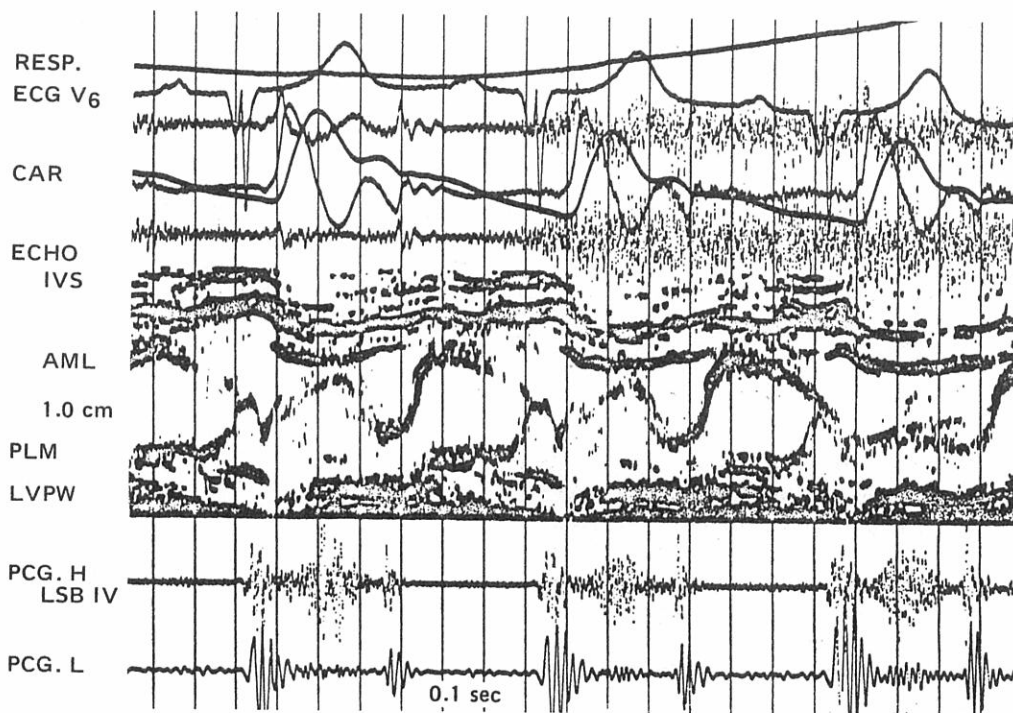


Figure 5 Ultrasound cardiogram

图 5 超音波心臟記錄圖



Figure 6 Ultrasound cardiogram

图 6 超音波心臟記錄圖

TABLE 2 ENDOCARDIAL &amp; VASCULAR PRESSURE BY PREOPERATIVE LEFT VENTRICULAR CATHETER EXAMINATION

表2 術前左心カテーテル検査による心・血管内圧

Site	Isoproterenol loading	
	Before	After
Left ventricle	136/00 mmHg*	172/00 mmHg
Left ventricular outflow tract	96/00	150/00
Aorta	90/56	104/64

\*Systolic / Diastolic

**Left Ventricular Catheter Findings.** The results of the left ventricular catheterization are shown in Table 2. The left ventricular systolic pressure was 136 mmHg, left ventricular diastolic pressure 0 mmHg, systolic pressure of the left ventricular outflow tract 96 mmHg, aortic systolic pressure 90 mmHg, and aortic diastolic pressure 56 mmHg. After a load of isoproterenol, the left ventricular systolic pressure became 172 mmHg, systolic pressure of left ventricular outflow tract 150 mmHg, aortic systolic pressure 104 mmHg, and aortic diastolic pressure 64 mmHg. All showed high values after loading, and the difference between left ventricular and aortic pressures after loading increased from 46 mmHg to 68 mmHg, and an increased systolic murmur was noted at the same time.

**Angiographic Findings of the Left Ventricle.** Frontal and lateral angiograms of the left ventricle are shown in Figures 7 and 8. The lateral picture showed narrowing of the left ventricular outflow tract into the shape of an inverted cone during systole. The frontal picture also showed stenosis accompanying thickening of the ventricular septum.

**Postoperative Course and Examination Findings.** Under the clinical diagnosis of IHSS based on the findings described above, excision of the hypertrophic myocardium was performed via the aorta. The postoperative course was very satisfactory and the patient was able to return to normal everyday life. Left ventricular angiography performed 45 days after the operation resulted in findings as shown in Figures 9 and 10. The shadow of the shape of an inverted cone present before the operation could no longer be seen.

左心カテーテル所見： 左心カテーテル検査成績は表2に示すごとく、左室収縮期圧は136mmHg、左室拡張期圧は0mmHg、左室流出路収縮期圧は96mmHg、大動脈収縮期圧は90mmHg、拡張期圧は56mmHgであった。これに対し、Isoproterenol 負荷後には左室収縮期圧172mmHg、左室流出路収縮期圧150mmHg、大動脈収縮期圧104mmHg、拡張期圧64mmHgとなり、いずれも負荷後に高値を示し、さらに負荷後の左室と大動脈との圧較差は46mmHgから68mmHgに増加しており、同時に収縮期雑音の増強が認められた。

左室腔内造影所見： 左室腔内造影所見の正面像は図7に、側面像は図8に示した通りである。側面像では収縮期に左室流出路は逆円錐状を呈して狭窄の像を示した。正面像においても同様に心室中隔の肥厚に伴う狭窄像が認められた。

術後経過ならびに検査成績： 上述のごとき所見によりIHSSの臨床診断の下に、経大動脈的に肥厚心筋切除術が実施された。術後の経過は極めて良好で正常の日常生活に復帰することが可能になっている。術後45日目に実施された左室造影像では図9および10に示すごとく、術前に認められた逆円錐形の陰影を認め得なくなった。

*Figure 7 Frontal angiocardigram of the left ventricle during systole*

图7 左室造影像(收缩期正面)

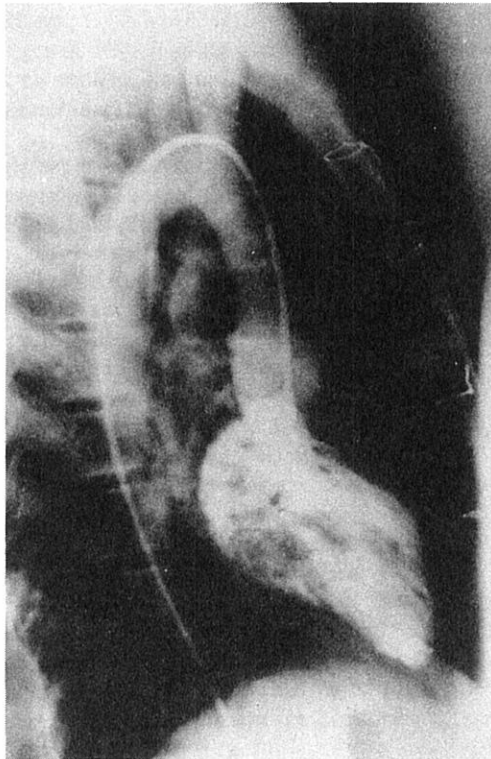
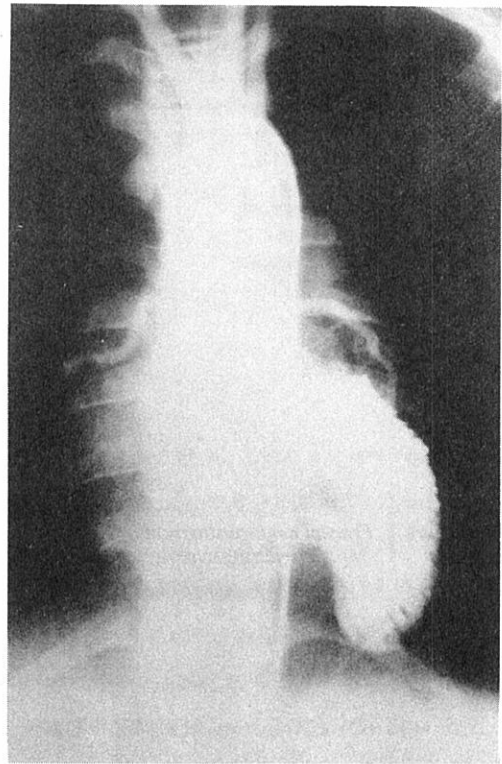


*Figure 8 Lateral angiocardigram of the left ventricle during systole*

图8 左室造影像(收缩期侧面)

*Figure 9 Postoperative frontal left ventricular angiogram*

図9 術後左室造影像(正面)



*Figure 10 Postoperative lateral left ventricular angiogram*

図10 術後左室造影像(側面)

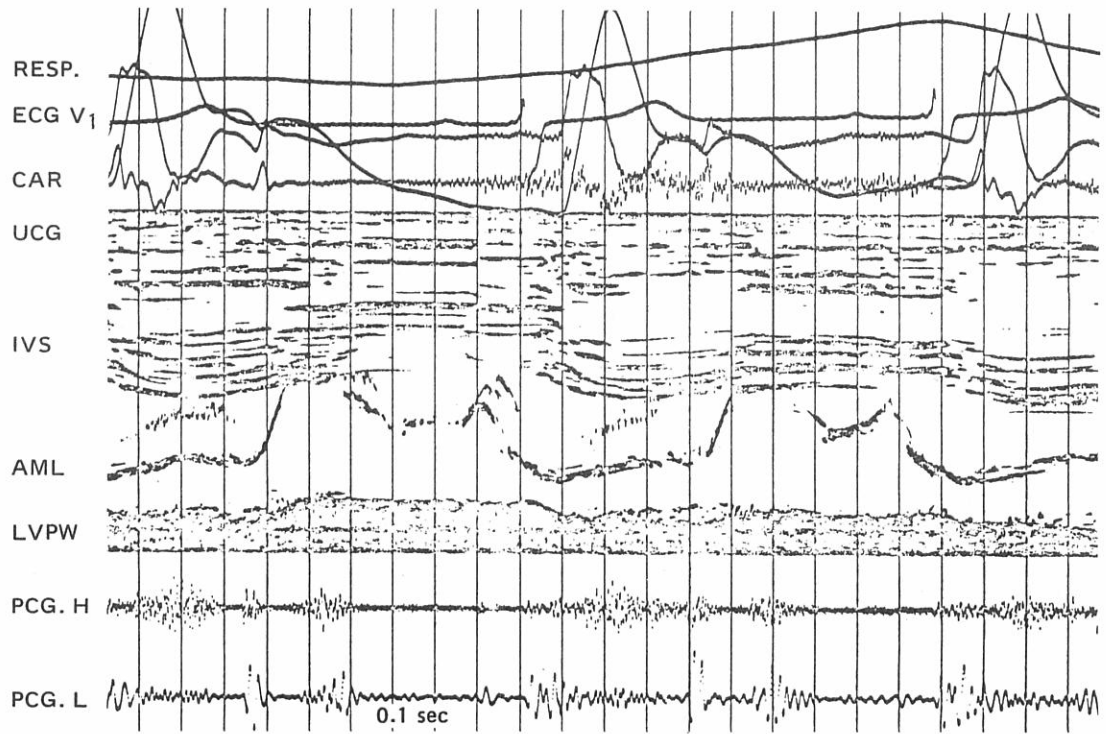


Figure 11 Postoperative ultrasound cardiogram

图11 術後超音波心臟記錄圖

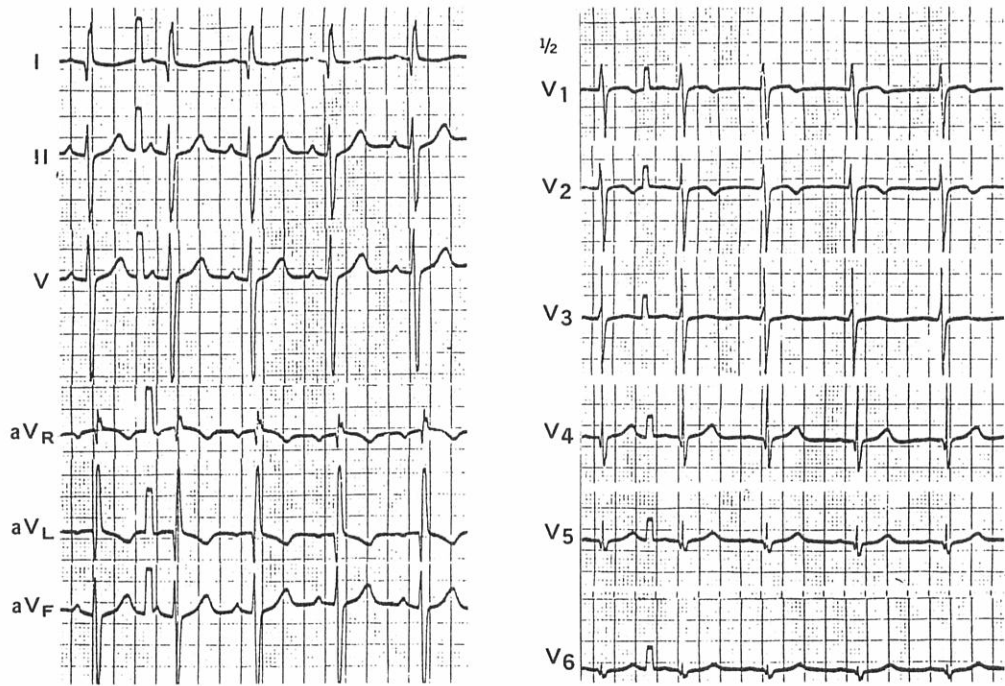


Figure 12 Postoperative electrocardiogram

图12 術後心電圖

TABLE 3 ENDOCARDIAL &amp; VASCULAR PRESSURE BY POSTOPERATIVE LEFT VENTRICULAR CATHETER EXAMINATION

表3 術後左心カテーテル検査による心・血管内圧

Site	Isoproterenol loading	
	Before	After
Left ventricle	128/08 mmHg*	180/-20 mmHg
Left ventricular outflow tract	128/08	160/-20
Aorta	112/60	132/64

\*Systolic / Diastolic

In the left ventricular catheter test, the left ventricular outflow vessel pressure was 128/8 mmHg and the aortic pressure was 112/60 mmHg (Table 3). With an isoproterenol load, the left ventricular outflow tract pressure became 160/-20 mmHg and the aortic pressure, 132/64 mmHg, showing a marked decrease in the difference in pressure after loading compared with before the operation. The UCG findings (Figure 11) indicated improvement of the DDR of the anterior mitral cusp to 62.5 mm/sec, a slight decrease of the systolic hump, and enlargement of the left ventricular outflow tract. Further, the transient semiclosure of the aortic valve seen before the operation had completely disappeared. On ECG the axis deviation seen before the operation had become normal, and the broad Q wave of II,  $aV_f$  and  $V_3$  had disappeared (Figure 12).

#### DISCUSSION

This is a report of a 16-year-old male suspected of having IHSS on conservative clinical examination who, after having the diagnosis confirmed by invasive procedures, was successfully treated by surgical operation. This is a disease which has recently attracted attention because it presents various clinical pictures as a result of special circulatory kinetics, but is now possible to diagnose clinically by conservative examinations. IHSS, with particular reference to the use of UCG, is discussed below.

According to the classification of Goodwin and Oakley<sup>5</sup> this disease can be considered to belong to the obstructive type which shows narrowing of the left ventricular outflow tract in the hypertrophic type of idiopathic cardiomyopathy. In recent years this condition is also called asymmetric septal hypertrophy (ASH).<sup>6</sup>

左心カテーテル検査においては、左室流出路の圧は128/8 mmHg、大動脈圧は112/60 mmHgであった(表3)。また Isoproterenol 負荷後では左室流出路の圧は160/-20 mmHg、大動脈圧は132/64 mmHgとなり、術前に比べて負荷後の圧較差は著明に減少していた。一方 UCG においては図11に示すごとく僧帽弁前尖のDDRも62.5 mm/secに改善し、systolic hump のわずかな減少と左室流出路の拡大が認められた。さらに術前にみられた大動脈弁の一過性半閉鎖の所見は完全に消失していた。また心電図では術前に比べて軸偏位が正常化し、II,  $aV_f$ ,  $V_3$  の幅広いQ波が消失していた(図12)。

#### 考 察

非観血的な臨床検査によってIHSSが疑われ、観血的な検査によってその臨床診断が下され、外科的療法によって診断を確認すると共に、改善させることに成功したIHSSの16歳の男性例について記載した。本疾患は特殊な血行動態により多彩な臨床像を呈することで、最近注目されてきた疾患であるが、非観血的な検査法を用いて臨床的に診断することが可能になってきた。以下本症、特にUCG利用を中心として若干の考察を加えたいと思う。

本症例はGoodwin およびOakley<sup>5</sup> の分類によれば、特発性心筋症の肥大型における左室流出路の狭窄を示す閉塞型(obstructive type)に属するものと考えられる。一方この状態は近年、非対称性中隔肥大、すなわち asymmetric septal hypertrophy (ASH)と

Mentioned as subjective symptoms are dyspnea on exertion, zonesthesia in the anterior chest area, palpitation, tachycardia, vertigo, and syncope. The apex beat is easily palpated on physical examination and mid-systolic ejection murmur reaching the peak at the apex and  $S_3$  and  $S_4$  are audible on auscultation. It is considered characteristic of the systolic murmur that it diminishes on intravenous injection of phenylephrine and increases with inhalation of amyl nitrite.<sup>7</sup> Noted in the cardiogram are marked "a" wave, systolic dip during early-to-mid-systole and late systolic expansion. The carotid wave rises sharply and presents bisferious pulse due to slow secondary wave which is caused by narrowing of the left ventricular outflow tract.

The ECG often shows left ventricular hypertrophy and abnormal Q waves<sup>8-10</sup> so that it is readily mistaken for myocardial infarction, but as this is caused by hypertrophy of the ventricular septum it is believed to be related to narrowing of the left ventricular outflow tract. On the other hand, as observed in this case, the findings of right axis deviation and right ventricular hypertrophy are sometimes found.<sup>1</sup> Tajik and Guiliam<sup>11</sup> mention as distinctive UCG findings of this disease: 1) abnormal forward movement (i.e., systolic anterior motion or systolic hump of the anterior mitral cusp), 2) IV septal hypertrophy or asymmetric septal hypertrophy, 3) decreased DDR, 4) systolic deformity of aortic valve, 5) narrowing of the left ventricle, and 6) diastolic septum contact.

Since the discovery of systolic anterior motion (SAM) as an abnormal finding in IHSS for the first time by Shah et al,<sup>12</sup> it has been considered a distinctive finding for the diagnosis of IHSS because it reflects stenosis of the left ventricular outflow tract. This abnormal SAM appears suddenly in mid-diastole and appears characteristically as a posterior motion from this period to late systole or isochoric diastole. The mechanism of its development is explained as follows: 1) The mitral valve is pulled toward the septum due to contraction of the papillary muscles, 2) the mitral valve is sucked forward due to narrowing of the left ventricular outflow tract and the so-called venturi effect is seen, and 3) the mitral valve is drawn from behind due to the valve being abnormally positioned in relation to the blood flow in the left ventricular outflow tract.<sup>13,14</sup>

も称されている。<sup>6</sup> 自覚症状としては運動時の呼吸困難, 前胸部絞扼感, 動悸, 頻脈, めまいおよび失神発作などが挙げられている。理学的検査においては心尖拍動がよく触知され, 聴診では心尖部に最強点を有する収縮中期駆出性雑音とⅢ, Ⅳ音が聴取される。この収縮期雑音は phenylephrine の静脈内注射で減弱し, amyl nitrite の吸入で増強することが特徴的である<sup>7</sup>とされている。心尖拍動図においては著明な"a"波, 収縮の前期から中期にかけ陥凹 (systolic dip), 収縮後期の膨隆 (late systolic expansion) を認める。頸動脈波は急峻な立ち上がり と二峰性脈 (bisferiens pulse) を示すが, これは左室流出路の狭窄による slow secondary wave によるものである。

心電図はしばしば左室肥大と異常Q波を示すので<sup>8-10</sup> 心筋硬塞と混同されやすいが, これは心室中隔の肥大によるもので, 左室流出路の狭窄と関係があるものとみられる。一方, 本症例においてみられたごとく, 右軸偏位, 右心肥大の所見を示す場合もみられている。<sup>1</sup> Tajik および Guiliam<sup>11</sup> は本疾患におけるUCGの特徴的所見として 1) 僧帽弁前尖の異常前方運動, すなわち systolic anterior motion (SAM) または systolic hump, 2) 心室中隔肥厚または非対称性肥大 (IV septal hypertrophy or asymmetric septal hypertrophy), 3) 僧帽弁前尖拡張期後退速度の低下 (decreased DDR), 4) 大動脈弁収縮期変形, 5) 左室内腔の狭小化および 6) diastolic septum contact を挙げている。

SAMはIHSSにおける異常所見として初めて Shahら<sup>12</sup> によって発見されて以来, 左室流出路の狭窄を反映するものとしてIHSSの診断上特徴的な所見となっている。この異常なSAMは収縮中期に急に出現し, 収縮後期あるいは等容弛緩期にかけて特異的に posterior motion として現れる。その発生機序として次のように説明されている。すなわち, 1) 乳頭筋の収縮により僧帽弁が中隔に向かって牽引される, 2) 左室流出路狭小のため僧帽弁が前方に吸引されて, いわゆる Venturi 効果がみられる, 3) 僧帽弁が左室流出路の血流に対して異常な位置にあるため, 後方から引かれるとされている。<sup>13,14</sup>

Recently, however, many investigators hold that this motion is not related to contraction of the papillary muscles but is due to a hemodynamic force caused by contraction of the left ventricle in a condition where there is abnormal thickening of the septum, and Nimura<sup>15</sup> reported that the cause, in essence, is not in the stenosis of the outflow tract itself but in the process of thickening and abnormal contraction of the posterior wall of the left ventricle. SAM is also noted in atrial septal defect, but differing from IHSS, it usually is noted only during late systole.<sup>16</sup> Further, in arteriosclerotic heart diseases and diseases of hyperdynamic status which present left ventricular hypertrophy, echo-waves easily mistaken for SAM of IHSS are obtained,<sup>17</sup> but these are readily distinguishable from anterior mitral cusp echoes because they have no continuity, clearness, nor reproducibility.<sup>16</sup>

It is reported also that when SAM is absent in IHSS, it can be induced by amyl nitrite or isoproterenol.<sup>18</sup> On the other hand, Henry et al<sup>19</sup> stress asymmetric septal hypertrophy as an abnormal UCG finding in IHSS, mentioning in particular that the ratio of septal thickness to left ventricular posterior wall thickness (IS/PW) was 1.68 where the diagnostic criterion for hypertrophic myocardiosis is a ratio of 1.3 or over. The thickness of the septal echo is naturally increased as an UCG finding of IHSS but this finding is sometimes also noted in hypertrophic myocardiosis and in arteriosclerotic heart disease in which no outflow tract stenosis is noted.<sup>20</sup>

Organic change of the mitral valve has been given as the cause for decreased DDR of the anterior cusp of mitral valve,<sup>21</sup> but this is considered to be affected by many factors such as decreased amount and rate of blood flow into the left ventricle and decreased distensibility of the left ventricle.<sup>16</sup> Decreased DDR is an important finding for mitral stenosis but this is noted also in primary myocardial diseases, retention of pericardial fluid and grave aortic insufficiency. That is, decreased DDR is believed to be caused by elevation of the left ventricular blood pressure in the terminal stage of diastole, decreased repletion of the left ventricle, and decreased compliance of the left ventricular myocardium.<sup>22</sup> The DDR which is normally 70-150 mm/sec had decreased markedly to 30 mm/sec in this case.

There are reports to the effect that the change

最近この motion は乳頭筋の収縮とは関係なく、中隔の異常な肥厚における左室収縮による hydrodynamic force のためと主張する研究者が多く、仁村<sup>15</sup>はこれを流出路の狭窄そのものによるのではなく、その本質は左室後壁の肥厚、異常収縮過程にあると報告している。SAMは心房中隔欠損症においてもみられるが、IHSSと異なり収縮後期のみ認められるのが通常である。<sup>16</sup> また左室肥大を示す動脈硬化性心疾患や hyperdynamic status にある疾患ではIHSSのSAMと紛らわしいエコー波が得られるが、<sup>17</sup> 僧帽弁前尖エコーとの連続性、明確さ、再現性はなく容易に鑑別できる。<sup>16</sup>

IHSSではSAMを欠如する場合には amyl nitrite あるいは isoproterenol によって誘発されるという記載もみられる。<sup>18</sup> 一方 Henry ら<sup>19</sup>はIHSSのUCGにおける異常所見として、asymmetric septal hypertrophy (ASH)を強調し、特に中隔と左室後壁の厚みの比 (IS/PW)が1.3以上になることを肥大型心筋症の診断基準とし、IHSSでは1.68であったと記載している。IHSSのUCG所見として中隔エコーの厚さが増加することは当然であるが、この所見は流出路の狭窄がみられない肥大型心筋症または動脈硬化性心疾患においてもみられることがある。<sup>20</sup>

僧帽弁前尖のDDRの低下は従来より僧帽弁の器質的変化が原因であるとされているが、<sup>21</sup> 左室流入量、流入速度の低下、左室の distensibility の低下などの多くの因子に影響されることも考えられている。<sup>16</sup> DDRの低下は僧帽弁狭窄に対する重要な所見であるが、原発性心筋疾患、心嚢液貯溜、重症大動脈弁閉鎖不全症などにおいても認められる。すなわちこのDDRの低下は左室拡張終末期圧の上昇、左室内充満度の減少、左室心筋コンプライアンスの低下によるものと考えられる。<sup>22</sup> DDRの正常値は70-150 mm/sec であるが、本症例においては30 mm/sec で著明に低下していた。

大動脈弁の収縮期にみられる動きの変形は、等容収



of motion of the aortic valve during systole occurs due to restriction of the downward movement and transient expansion of the valve toward the aorta, caused by changes in the mode of ventricular contraction leading to isochoric systole. Tanaka et al<sup>23</sup> attribute this mainly to decrease in amount of ejected blood due to stenosis of left ventricular outflow tract, but the true developmental mechanism seems to be yet unknown. In our case also, the echo waves at the time of open aortic valve showed transient semioclusion of the aortic valve consistent with the peak of the ejection murmur on phonocardiography. Diastolic septal contact which has been emphasized as being a characteristic finding of IHSS,<sup>4</sup> is also to be found in atrial septal defect<sup>24</sup> and arteriosclerotic cardiac diseases. Furthermore, due to physiologically decreased circulation rate and narrowing of the left ventricle with aging, the ventricular septum is forced to curve and the point of the greatest curvature comes to be positioned under the mitral valve. This contact is also seen occasionally among persons of advanced age.<sup>16</sup>

As postoperative changes on UCG, Bolton et al<sup>25</sup> reported finding markedly decreased SAM, improvement of stenosis of left ventricular outflow tract and decreased thickness of the ventricular septum. Our case also showed, postoperatively, decreased SAM, improvement of decreased DDR, slight decrease in thickness of ventricular septum, and disappearance of deformity of the aortic valve during systole. The patient was able to return to normal life with improvement of subjective symptoms.

As above, it was found in the present case that of the many clinical-laboratory tests, UCG was not only the most reliable and easy method for clinical diagnosis but also because of its conservatism and repeatability, a very useful method for evaluating the postoperative course.

縮期に至るまでの心室収縮様式の変化により、弁輪の下降が制限されると共に大動脈側への一過性膨隆によって起こるという記載がみられる。田中ら<sup>23</sup>は左室流出路の狭窄による駆出血流量の減少が主因であるとしているが、真の発生機序はいまだ不明のようである。本報告の症例においても大動脈弁開放時におけるエコー波をみると、心音図における駆出性雑音の peak に一致して、大動脈弁の一過性の半閉鎖状態が認められた。diastolic septal contact は IHSS の特徴的な所見として強調されてきたが、<sup>4</sup> その他に心房中隔欠損症<sup>24</sup> および動脈硬化性心疾患においても認められる。また加齢に伴う生理的な心拍出量の減少と左室腔の狭小化により、心室中隔の弯曲が強られるため僧帽弁下に中隔の最大屈曲点が位置するようになるので、この contact は高齢者においても認められることがある。<sup>16</sup>

術後の UCG 像の変化として Bolton ら<sup>25</sup> は SAM の著明な減少、左室流出路の狭窄の改善、心室中隔の厚さの減少などがみられると記載している。本報告の症例においても SAM の減少、DDR 低下の改善、心室中隔の厚さの軽度減少および大動脈弁収縮期変形の消失が術後に認められ、自覚症も改善して通常の生活に復帰することが可能であることが観察された。

以上本症例においては多くの臨床検査のうち UCG が最も確実かつ容易な臨床診断法であるばかりでなく、非観血的検査であるので繰り返し検査を実施することにより、術後経過の判断にも極めて有用であることが認められた。

## REFERENCES

## 参考文献

1. BRAUNWALD E: Idiopathic hypertrophic subaortic stenosis. *Circulation* 30 Suppl 4:1-223, 1964
2. BRAUNWALD E, MORROW AG, CORNELL WP, AYGEM MM, HILBISH TF: Idiopathic hypertrophic subaortic stenosis; clinical, hemodynamic and angiographic manifestation. *Am J Med* 29:924-45, 1960
3. 鷹津 正, 北浦 泰: 特発性肥大性大動脈弁下狭窄症(血行動態と診断). *日本臨牀* 31: 2550-62, 1973.  
(TAKATSU T, KITaura Y: Idiopathic hypertrophic subaortic stenosis (hemodynamic and diagnosis). *Nippon Rinsho-Jpn J Clin Med*)
4. MOREYRA E, KLEIN JJ, SHIMADA H, SEGAL BL: Idiopathic hypertrophic subaortic stenosis diagnosed by reflected ultrasound. *Am J Cardiol* 23:32-37, 1969
5. GOODWIN JF, OAKLEY CM: The cardiomyopathies. *Br Heart J* 34:545-52, 1972
6. HENRY WL, CLARK CE, EPSTEIN SE: Asymmetric septal hypertrophy (ASH); the unifying link in the IHSS disease spectrum. Observations regarding the pathogenesis, pathophysiology, and course. *Circulation* 47:827-32, 1973
7. NELLEN M, BECK W, VOGELPOEL L, SHIRO V: Auscultatory phenomena in hypertrophic obstructive cardiomyopathy. Ciba Foundation Study Group. No. 37, Ed by Wolstenholme GEW & O'Conner M, London, J and A Churchill, 1971. p77
8. PRESCOFF R, QUINN JS, LITMANN D: Electrocardiography changes in hypertrophic subaortic stenosis due to ventricular septal hypertrophy. *Am J Cardiol* 14:599-607, 1964
9. HOLLISTER RM, GOODWIN JF: The electrocardiogram in cardiomyopathy. *Br Heart J* 25:357-74, 1963
10. WIGLE ED, BARON RH: Electrocardiogram in muscular subaortic stenosis. *Circulation* 34:585-94, 1966
11. TAJIK AJ, GUILIAM GT: Echocardiographic observations in idiopathic hypertrophic subaortic stenosis. *Mayo Clin Proc* 49:89-97, 1974
12. SHAH PM, GRAMIAK R, KRAMER DH: Ultrasound localization of left ventricular outflow obstruction in hypertrophic obstructive cardiomyopathy. *Circulation* 40:3-11, 1969
13. HENRY WL, CLARK CE, GRIFFITH JM, EPSTEIN SE: Mechanism of left ventricular outflow obstruction in patient with obstructive asymmetric septal hypertrophy (idiopathic hypertrophic subaortic stenosis). *Am J Cardiol* 35:337-45, 1975
14. 鷹津 正, 堀 健次郎, 井上啓造, 北浦 泰, 陳 憲雄, 田中孝生, 園谷 昇, 諏訪道博, 塘 二郎: 特発性心筋症の諸問題. *臨床科学* 10: 1273-91, 1975.  
(TAKATSU T, HORI K, INOUE K, KITaura Y, CHIN N, TANAKA T, SONOYA N, SUWA M, TSUTSUMI J: The problems of idiopathic cardiomyopathy. *Rinsho Kagaku-J Clin Sci*)
15. 仁村泰治: 超音波像による特発性心筋症へのアプローチ(会). *Jap Circul J* 38(Suppl): 90, 1974.  
(NIMURA Y: Ultrasound in the approach of idiopathic cardiomyopathy. *Nippon Junkankigakushi-Jpn Circ J* (Abstract))
16. 吉川純一, 田中久米夫, 田中忠次郎, 古川宏太郎, 川合清毅, 竹内一秀, 大脇 嶺, 加藤 洋: UCG による特発性肥大性大動脈弁下狭窄症(IHSS)の診断. *Heart* 6: 328-37, 1974.  
(YOSHIKAWA J, TANAKA K, TANAKA C, FURUKAWA K, KAWAI K, TAKEUCHI K, OOWAKI R, KATO H: Echocardiographic diagnosis of idiopathic hypertrophic subaortic stenosis. *Heart*)

17. POPP RL, HARRISON DC: Ultrasound in the diagnosis and evaluation of therapy of idiopathic hypertrophic subaortic stenosis. *Circulation* 40:905-14, 1969
18. SHAH PM, GRAMIAK R, ADELMAN AG, WIGLE ED: Role of echocardiography in diagnostic and hemodynamic assessment of hypertrophic subaortic stenosis. *Circulation* 44:891-98, 1971
19. HENRY WL, CLARK CE, EPSTEIN SE: Asymmetric septal hypertrophy - Echocardiographic identification of the pathognomonic anatomic abnormality of IHSS. *Circulation* 47:225-33, 1973
20. ABBASI AS, MACALPIN RN, EBER LM, PEARCE ML: Echocardiographic diagnosis of idiopathic hypertrophic cardiomyopathy without outflow obstruction. *Circulation* 46:897-904, 1972
21. JOYNER CR JR, REID JM, BOND JP: Reflected ultrasound in the assessment of mitral valve disease. *Circulation* 27:503-11, 1963
22. STEWART S, MASON DT, BRAUNWALD E: Impaired rate of left ventricular stenosis and valvular aortic stenosis. *Circulation* 37:8-14, 1968
23. 田中元直, 香坂茂美, 岡 捨己, 寺沢良夫, 海野金次郎, 仁田佳子, 柏木 誠, 海老名敏明: 大動脈弁動態とその診断的意義. 第21回日本超音波医学会講演集. 1972. p 101  
(TANAKA M, KOSAKA S, OKA S, TERAZAWA Y, KAINO K, NITA Y, KASHIWAGI M, EBINA T: The diagnosis of aortic valve motion. Proceedings of the 21st Meeting of Japanese Ultrasound Medicine Society, 1972. p101)
24. DIAMOND MA, DILLON JC, HAINE CL, CHANG L, FEIGENBAUM H: Echocardiographic features of atrial septal defect. *Circulation* 43:129-35, 1971
25. BOLTON MR, KING JF, POLUMBO RA, MASON D, PUGH DM, REIS RL: The effect of operation on the echocardiographic features of idiopathic hypertrophic subaortic stenosis. *Circulation* 50:897-900, 1974