

Phonocardiography and intracardiac phonocardiography in hypertrophic cardiomyopathy

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Summary: The salient phonocardiographic features of patients having hypertrophic cardiomyopathy (HCM) with or without left ventricular outflow tract (LVOT) gradients are reviewed. Intracardiac sound and pressure recordings from high fidelity catheter-tipped micromanometers have documented that the precordial murmur is the summation of both the systolic ejection murmur (SEM) arising from the LVOT, as well as the mitral regurgitant murmur recorded from the left atrium. The intensity of the precordial murmur varies directly with the LVOT gradient, which in turn is determined primarily by the contractility and loading conditions of the left ventricle.

Reversed splitting of the second heart sound (S_2) with paradoxical respiratory movement is a common finding in HCM, and when present, almost always denotes a significant LVOT gradient. It is due to marked lengthening of the left ventricular ejection time secondary to prolongation of the contraction and relaxation phases of left ventricular systole. The presence of a fourth heart sound (S_4) is the rule in HCM when normal sinus rhythm is present, and is a reflection of a forceful left atrial contraction into a hypertrophied noncompliant left ventricle. A third heart sound (S_3) is also common in HCM, and often the initial vibrations occur before the 0 point of the apexcardiogram (ACG) and continue giving the auscultatory impression of a diastolic rumble. When associated with a loud S_1 , which is frequently present, the clinical presentation may mimic mitral stenosis. This is particularly true when the patient has chronic atrial fibrillation. Careful attention to evidence of marked left ventricular hypertrophy as well as the typical echocardiographic findings of HCM preclude this diagnosis.

In conclusion, phonocardiography is a simple non-invasive technique which almost always makes the definitive diagnosis of HCM.

Phonocardiography is the graphic representation of the bedside cardiovascular examination. It includes not only the recording of the auscultatory phenomena of the precordium, but also the carotid, venous, and cardiac apical pulsations simultaneously with the electrocardiogram (Shaver, 1981). When coupled with the M-mode echocardiogram, the diagnostic capabilities of this technique are further enhanced by correlating the sound and pressure tracings of the cardiac cycle with dynamic valvular and ventricular wall motion. Probably no disease lends itself so well to evaluation by these techniques as hypertrophic cardiomyopathy (HCM), because of its unique physical and anatomical findings. In this report, the salient features of patients with HCM will be described as recorded by phonocardiography. Furthermore, new insights into the mechanisms of the auscultatory findings in this entity will be provided by utilizing intracardiac recordings of sound and pressure

obtained from high fidelity catheter-tipped micromanometers.

The phonocardiogram of a 54 year old patient with HCM is shown in Figure 1, and is typical of patients with this disease having a significant left ventricular outflow tract gradient. At cardiac catheterization, a resting peak gradient of 60–80 mmHg was found with a left ventricular peak pressure of 180 mmHg, and a left ventricular end-diastolic pressure of 16 mmHg. The carotid pulse has the typical rapid rise followed by a late systolic plateau, and the left ventricular ejection time is prolonged. The apexcardiogram (ACG) demonstrates the classic triple pulsation consisting of the presystolic a wave, an early systolic outward pulsation, and a late systolic bulge. A loud fourth heart sound (S_4) occurs during the rapid upstroke of the a wave, and a softer third heart sound (S_3) occurs during the rapid filling wave (RFW) of the ACG. A grade V systolic ejection murmur begins during early left ventricular ejection shortly after the development of the left ventricular-aortic pressure gradient, and peaks during mid-systole at the time of the peak gradient.

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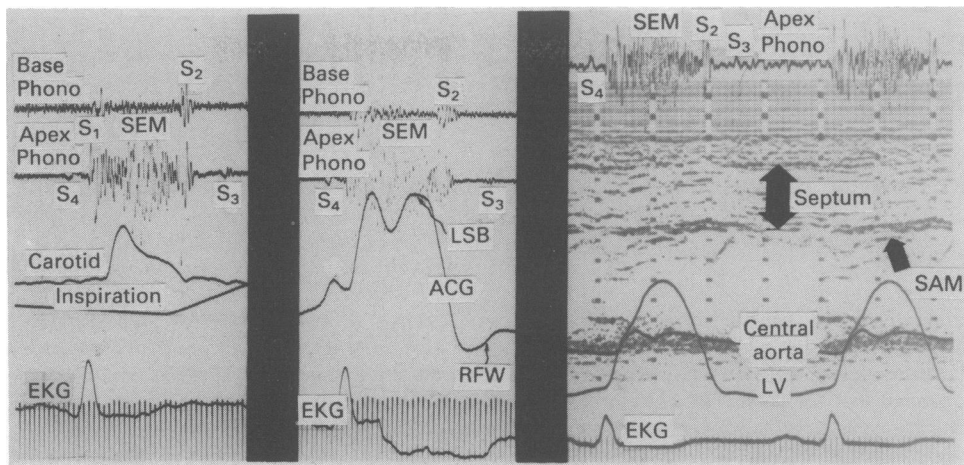


Figure 1 Simultaneous base and apex phonocardiograms are recorded with the carotid pulse and apexcardiogram (ACG) in the left and centre panels respectively, in a 54 year old male with HCM. The carotid pulse rises rapidly, has a late systolic plateau, and a prolonged ejection period. Prominent S_4 and S_3 are demonstrated, and are associated with the a wave and the rapid filling wave (RFW) respectively, of the ACG. Note the late systolic bulge (LSB) on the ACG. The second heart sound (S_2) is single. A loud grade V systolic ejection murmur (SEM) is present, and is of greatest intensity at the apex. In the right panel, the apical SEM is recorded together with the M-mode echocardiogram. Simultaneous high fidelity left ventricular and central aortic pressures are recorded by catheter-tipped micromanometers. Marked thickening of the interventricular septum and systolic anterior motion (SAM) of the mitral valve are present on the echocardiogram. A large systolic pressure gradient is demonstrated, beginning shortly after the onset of SAM.

Systolic anterior motion (SAM) of the mitral valve is present, and its duration correlates well with the duration and magnitude of the pressure gradient (Pollick *et al.*, 1984).

Frequently on auscultation, the skilled observer has difficulty in deciding whether the systolic murmur is ejection or regurgitant in nature. The explanation for this confusion is readily understood by analysis of the intracardiac sound and pressure recordings shown in Figure 2, as recorded from another patient with HCM. A typical systolic ejection murmur is recorded from the catheter-tipped micromanometer in the left ventricular outflow tract, while a late systolic murmur of mitral regurgitation is recorded from the transeptal left atrial micromanometer. The onset of this latter murmur begins shortly before contact of the anterior mitral leaflet with the septum (SSC) as timed by simultaneous M-mode echocardiography of the mitral valve. Thus, the resulting external murmur recorded by the precordial phonocardiogram is the summation of both murmurs as transmitted to the chest wall.

In patients with dynamic left ventricular outflow tract gradients, the intensity of both the systolic ejection murmur and the mitral regurgitation murmur varies directly with the level of the pressure gradient. Thus, physiological manoeuvres and pharmacological interventions which increase the pressure gradient will

increase the intensity of the precordial murmur and *vice versa*. Decreases in left ventricular preload and afterload, or increases in left ventricular contractility are associated with increases in both the pressure gradient and the intensity of the murmur, while increases in left ventricular preload and afterload or decreases in left ventricular contractility will decrease the pressure gradient and the intensity of the murmur (Glancy *et al.*, 1971; Reddy *et al.*, 1971). For example, both the upright posture and the strain phase of the Valsalva manoeuvre decrease venous return and left ventricular preload, and the murmur increases in intensity. Upon reclining or with prompt squatting, augmented venous return increases left ventricular preload, and the murmur decreases in intensity. Vasodilator drugs, e.g. inhaled amyl nitrate, decrease blood pressure and a marked increase in the intensity of the murmur occurs, whereas vasoconstrictive drugs, such as infused phenylephrine, increase the afterload and the murmur is decreased or abolished. Inotropic drugs such as digitalis and catecholamines increase the intensity of the murmur while beta blockade usually decreases it. The responses to these interventions are relatively specific for HCM, although occasionally patients with a mitral valve prolapse syndrome behave similarly. However, the clinical setting of this entity is easily differentiated and rarely confused with HCM.

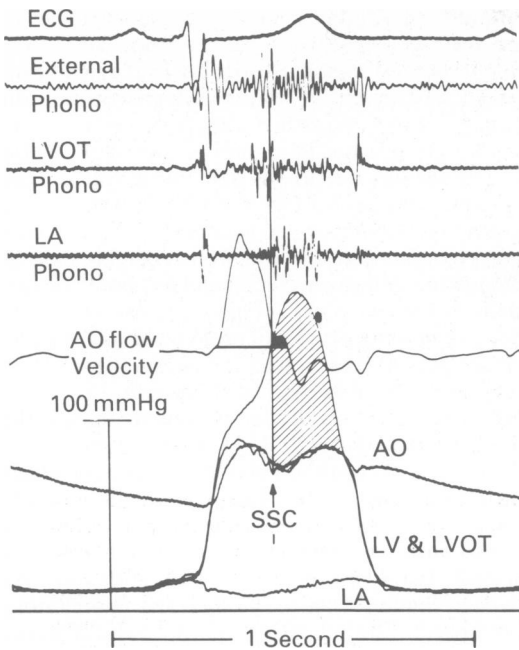


Figure 2 Catheter-tipped micromanometer pressures are recorded from the left atrium (LA), left ventricle (LV), left ventricular outflow tract (LVOT), and aorta (AO), together with external sound and intracardiac sound recorded from micromanometers in the LA and LVOT, in a patient with HCM. Aortic flow velocity is recorded by a flow probe in the central aorta. The vertical line denotes the time of SAM-septal contact (SSC) as determined by simultaneous M-mode echocardiography of the mitral valve. Note that the majority of aortic flow occurs before SSC. A SEM is recorded from the LVOT, and a late systolic mitral regurgitant murmur is recorded from the LA. The external phonocardiogram represents the summation of these two murmurs. (Courtesy of Dr Joseph Murgo, San Antonio, Texas).

In Figure 3, a finding unique to HCM with a resting or provokable gradient is shown. Following the long diastolic pause after a premature ventricular contraction (PVC), there is a significant increase in the intensity of the left ventricular outflow murmur associated with a decreased pulse pressure in the central aortic tracing. The normal response after a long filling cycle would be an increase in the intensity of any systolic ejection murmur associated with an increase in the arterial pulse pressure. The finding of a decreased pulse pressure following a long diastolic pause can be easily appreciated by palpating the carotid pulse at the bedside when extra systoles are present, and is diagnostic of this condition. The increase in the intensity of the externally recorded murmur following the compensatory pause after the

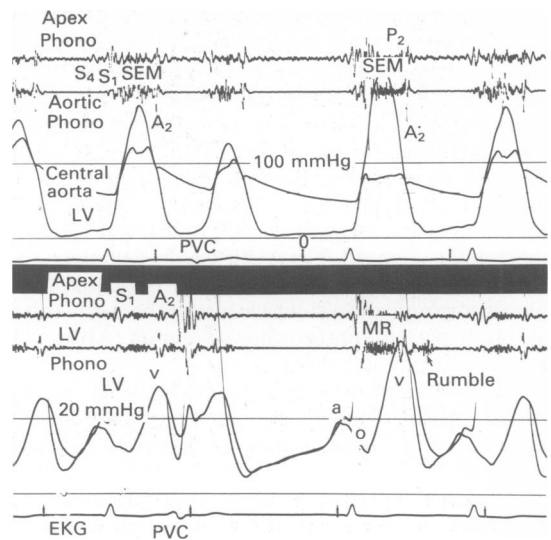


Figure 3 Top panel: Left ventricular and central aortic catheter-tipped micromanometer pressures are recorded, together with the apex phonocardiogram and intracardiac sound from the central aorta in a 71 year old woman with HCM having a dynamic LVOT gradient. Following a long diastolic filling period after a premature ventricular contraction (PVC), there is a marked increase in the LVOT gradient associated with a significant increase in the SEM, as recorded externally and in the central aorta. This is concomitant with a decrease in the pulse pressure recorded in the central aorta. In the lower panel, LV and LA pressures are recorded from equisensitive catheter-tipped micromanometers, together with the apex phonocardiogram and intracardiac sound from the inflow tract of the LV. A mitral regurgitant murmur is recorded from the LV phono, while the left atrial V wave rises to a peak of 20 mmHg. Following the compensatory pause after the PVC, there is a marked increase in the intensity of the mitral regurgitant murmur recorded by the LV micromanometer with a left atrial V wave peaking to 40 mmHg. Note the persistent true early diastolic gradient present in all complexes, and the concomitant early diastolic rumble recorded by the LV inflow phonocardiogram.

premature contraction is due not only to an increase in the left ventricular outflow ejection murmur, but also due to an increase in the degree of mitral regurgitation in the beat following the premature contraction. This is nicely shown in Figure 3 by an increase in the height of the left atrial V wave and by an increase in the intensity of the mitral regurgitant murmur as recorded by the intracardiac micromanometer. Again, the increased intensity of the precordial murmur is a summation of both the systolic ejection murmur and the mitral regurgitant murmur.

In the absence of a left ventricular outflow gradient

at rest or with provocation, the murmur of HCM is much less impressive (Figure 4). Although a short systolic ejection murmur is usually recorded due to the rapid early left ventricular ejection, it is softer and extends through less of systole than when a gradient is present (Murgo *et al.*, 1980). There is also little variation in its intensity with changes in preload, afterload, or contractility. The carotid upstroke is normal in character, and the left ventricular ejection time is not prolonged. The ACG usually demonstrates a prominent a wave associated with an S_4 . SAM of the mitral valve and midsystolic closure of the aortic valve are rarely present on the echocardiogram (Alvares *et al.*, 1984; Doi *et al.*, 1980).

The documentation of reversed splitting of the second heart sound (S_2), having paradoxical movement with respiration, is an important physical finding in HCM. In the 64 patients with idiopathic hypertrophic subaortic stenosis (IHSS) reported by Braunwald and associates (1964), it was present in 22 patients (34%), and was associated with a significantly higher gradient than in those patients with a normal or single S_2 . In a recent study by Alvares and associates (1984), 15 of 84 patients with HCM were studied by

phonoecardiography and were documented to have reversed splitting. Catheterization data was available in 13 of these patients, and in all a significant gradient was present at rest or with provocation. In addition, all 15 patients had systolic anterior motion documented by echocardiography, and 14 of 15 also had midsystolic closure of the aortic valve. The mechanism responsible for the delayed aortic closure sound (A_2) is marked prolongation of the left ventricular ejection time resulting from a combination of prolongation of the contraction and relaxation phases of left ventricular systole (Figure 5) (Alvares *et al.*, 1984). When lesser degrees of prolongation of ejection time are present, the S_2 may be single (Figure 1) or splitting may be physiological (Figure 4).

When normal sinus rhythm is present, an S_4 is the rule in HCM (Figures 1, 4, 6). It is the result of a forceful left atrial contraction into a noncompliant left ventricle (Goodwin, 1982; Reddy *et al.*, 1971). Since massive left ventricular hypertrophy is present in patients with or without left ventricular outflow gradients, the S_4 is common in both situations. As shown in Figures 1, 4, 6, the S_4 is the auscultatory counterpart of the presystolic apical pulsation occur-

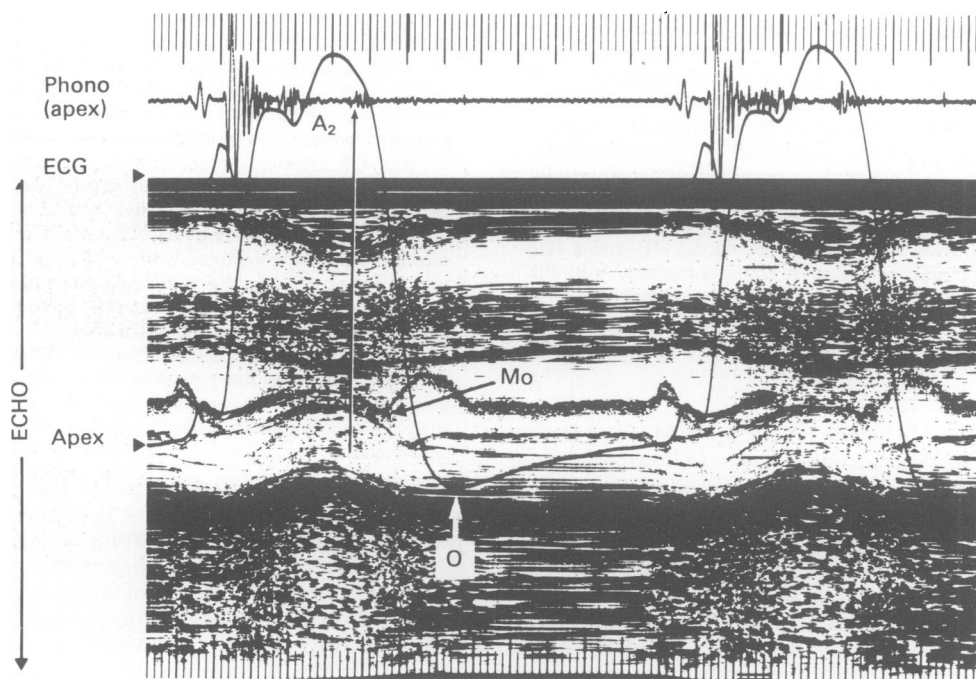


Figure 4 Simultaneous ACG and M-mode echocardiogram are recorded together with the ECG in a 41 year old male with HCM. A short SEM is present. A prominent presystolic pulsation is demonstrated on the ACG, which is coincident with the loud S_4 on the apex phonocardiogram. Splitting of S_2 is normal, and marked prolongation of the isovolumic relaxation period (A_2 -MO) to 110 ms is demonstrated. SAM of the mitral valve is absent. Marked thickening of the septum is present, with asymmetric hypertrophy. No LVOT gradient was measured at cardiac catheterization. (From Alvares *et al.*, 1984, reproduced by permission).

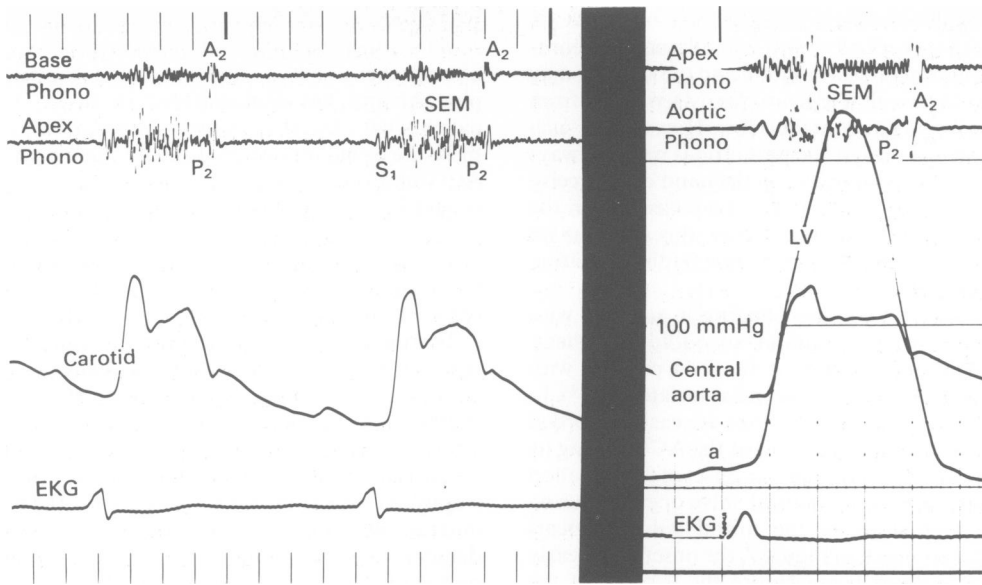


Figure 5 Left panel: Reversed splitting of S_2 is documented by the simultaneous recording of the carotid pulse with the base and apex phonocardiograms in a 71 year old woman with HCM. Right panel: A large resting LVOT gradient was measured with equisensitive catheter-tipped micromanometers in the LV and central aorta. A_2 is markedly delayed, due to both prolonged LV contraction and relaxation (same patient as Figure 3).

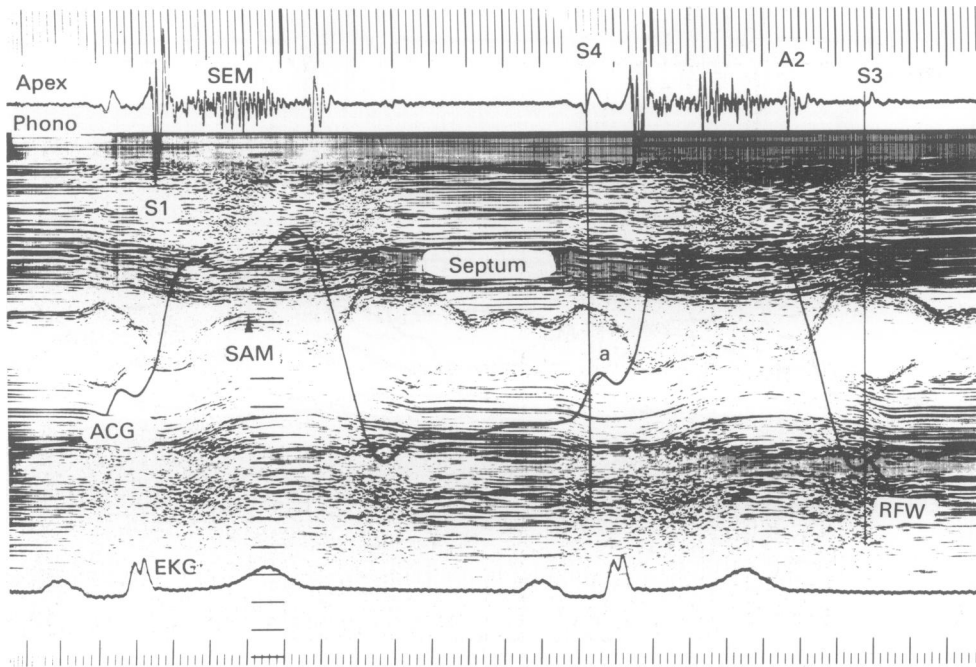


Figure 6 Simultaneous apex phonocardiogram, ACG, M-mode echocardiogram and EKG are recorded in a patient with HCM, having both S_4 and S_3 . The S_4 occurs shortly after the maximal opening of the atrial wave of the mitral valve echocardiogram during the rapid upstroke of the a wave of the ACG. The S_3 occurs 80 ms after the maximal opening of the mitral valve during the RFW of the ACG. Note the late systolic bulge demonstrated on the ACG. A prominent SEM is recorded, and SAM of the mitral valve is observed.

ring during the rapid rise of the a wave of the ACG. Although easy to identify by the simultaneous recording of the apex phonocardiogram and the ACG, it may be difficult to auscult if it is not clearly separated from S_1 , or if its frequency content is low. However, in such cases, the apical presystolic pulsation is nearly always present and easy to palpate, as the hand can perceive low frequency vibrations much more easily than the ear. When atrial fibrillation occurs, this sound is no longer present, and the apical presystolic pulsation likewise disappears.

The S_3 is also a common finding in patients with HCM, either with or without an outflow gradient. Such a sound was present in 38 of 64 patients with IHSS reported by Braunwald and associates (1964). In Figures 1 and 6, typical S_3 gallops are easily recorded and occur well after the 0 point of the ACG during its RFW. As shown in Figure 6, the onset of the S_3 gallop occurs after the maximal mitral valve opening during the early E-F slope of the mitral valve echocardiogram. In contrast, in Figure 7, the onset of the early diastolic vibrations begin before the 0 point of the ACG, and on close inspection, continue for considera-

ble duration, resembling more an early diastolic rumble rather than a discrete low frequency sound. In a comprehensive study utilizing phonoechocardiography, Alvares has demonstrated an S_3 in 19 of 60 patients with HCM in normal sinus rhythm; in 10 of these the S_3 gallop occurred before the 0 point of the ACG and often appeared as a rumble, as shown in Figure 7 (Alvares, 1980). Of further interest, he noted that an S_3 was present in 8 of 9 patients with HCM having atrial fibrillation, and in all it occurred before the 0 point of the ACG, often having vibrations of considerable duration.

The presence of such diastolic rumbles have been reported by other investigators in patients with HCM with clinical findings which mimic mitral stenosis (Shabetai & Davidson, 1972; Smith *et al.*, 1975). However, as pointed out by these reports, careful attention to evidence of marked left ventricular hypertrophy, as well as the typical findings of the echocardiogram allow the clinician to readily make the correct diagnosis of HCM, even when the loud S_1 and diastolic rumble clinically simulate mitral stenosis. It is possible that such diastolic rumbles may be secondary to

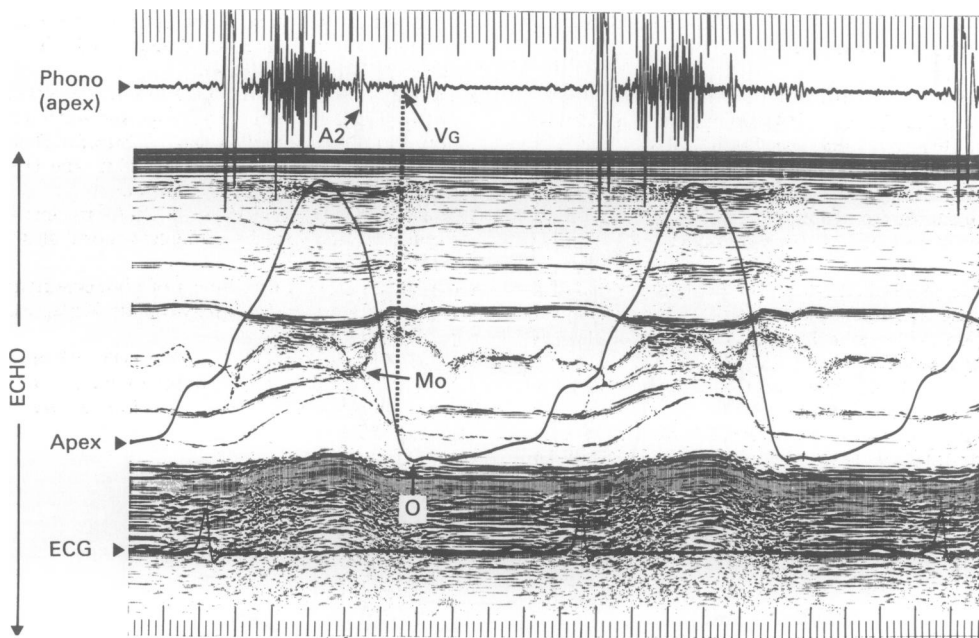


Figure 7 Simultaneous apex phonocardiogram, ACG, and M-mode echocardiogram are recorded together with the ECG in a patient with HCM having prominent early diastolic vibrations occurring well before the 0 point of the ACG. The first heart sound (S_1) is loud, and together with the early diastolic apical rumble, may mimic mitral stenosis on auscultation. However, the prominent presystolic heave and sustained apical systolic pulsation are incompatible with this diagnosis. The echocardiogram unequivocally documents the diagnosis of HCM with massive septal hypertrophy, SAM of the mitral valve, and reduced LV cavity size. A prominent SEM is recorded typical of HCM with a significant LVOT gradient, which was documented at cardiac catheterization. (From Alvares, 1980, reproduced by permission).

increased impedance to left ventricular filling secondary to decreased diastolic compliance of the left ventricle. That such a phenomenon may exist is suggested by the ability to record a diastolic rumble in the inflow tract of the left ventricle during early diastolic filling, when a definite left atrial-left ventricular pressure gradient is demonstrated by equisensitive catheter-tipped micromanometers (Figure 3). In this patient, there was no clinical or echocardiographic evidence of mitral valve stenosis, although the LV cavity was small with marked hypertrophy of the septal and posterior walls and prominent SAM of the mitral valve was evident. As shown in Figures 3 and 5, neither a typical S₃ nor an S₃ with duration was present on the external phonocardiogram. Whether in certain patients the early diastolic gradient and concomitant rumble can be of significant magnitude and intensity to transmit to the chest wall is not known at this time.

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