CASE REPORT

Complete Heart Block in a Case of Idiopathic Hypertrophic Subaortic Stenosis
Noninvasive Correlates with the Timing of Atrial Systole

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SUMMARY A young man with IHSS who developed complete heart block was successfully treated with a permanent pacemaker. Echocardiography and other noninvasive techniques showed marked cycle-to-cycle variation in the evidence of subvalvular obstruction which decreased markedly when atrial systole preceded the ensuing paced complex by an appropriate interval. Because cycle length and therefore afterload were constant, it is concluded that diminished obstruction resulted from augmented ventricular end-diastolic volume produced by atrial contraction. The mitral valve echocardiogram showed unusual movements in diastole dependent upon the timing of atrial systole. Early reopening of the leaflets was a direct result of atrial contraction when the P waves were appropriately timed in presystole, whereas late reopening was passive and a result of ventricular filling in mid-diastole. Variations in intensity of the first heart sound correlated with the position of the mitral valve leaflets at the onset of ventricular systole.

OBSTRUCTION TO THE LEFT VENTRICULAR OUT-FLOW TRACT in idiopathic hypertrophic subaortic stenosis (IHSS) is labile and susceptible to various maneuvers which alter ventricular contractility. Changes in the subaortic gradient have been demonstrated by cardiac catheterization, phonocardiography, and also echocardiography which detects abnormal anterior movement of the mitral valve in systole (SAM). Of the various factors which affect the gradient, alteration in ventricular volume is important and an increase diminishes the gradient, softens the systolic murmur, widens the pulse pressure and diminishes SAM. The effect of atrial contraction in changing end-diastolic volume and the subaortic gradient has occasionally been observed during episodes of A-V dissociation during cardiac catheterization. Similarly, because complete heart block so rarely accompanies IHSS, observations on the effect of atrial systole have only been made by measurements at the time of cardiac catheterization.

In the case of IHSS reported here, permanent pacing was needed to treat complete heart block; variation in cycle length which affects afterload was thus absent. The effect of atrial preloading of the ventricle was studied by echocardiography which also showed unusual movements of the mitral valve in diastole, dependent upon the timing of atrial contraction.

Case Report

The patient, a 20-year-old Asiatic male, complained of severe retrosternal chest pain and exertional dyspnea which had been progressive for the past three years. His father was known to have a heart murmur and his brother had died suddenly in India at the age of 16 years because of a “leaking heart valve.”

On examination the pulse had a rapid upstroke and a readily palpable apical precordial impulse had both forceful presystolic and systolic components. A grade II mid-systolic murmur was heard at the left sternal border and a loud fourth heart sound was present at the apex. The systolic murmur increased in intensity with the Valsalva maneuver and amyl nitrite inhalation but softened with squatting. A chest roentgenogram demonstrated left ventricular enlargement. The echocardiogram showed marked thickening of the interventricular septum but a normal posterior left ventricular wall; at end diastole the ratio of septal to posterior wall thickness was 2:1. The anterior leaflet of the mitral valve (AML) opposed the septum in early diastole, and had a reduced diastolic closure rate. In systole there was pronounced systolic anterior movement (SAM) of both the anterior and posterior leaflets.

He was admitted for cardiac catheterization largely because of unusual electrocardiographic features. The tracings showed sinus rhythm with a P-R interval of 0.20 sec. The mean frontal plane QRS axis was $-90^\circ$ and there was right bundle branch block. In addition there were deep Q waves in standard lead I and lead aVL.

The hemodynamic and cineangiographic findings were typical of IHSS. The cardiac index was 3.25 L/min/m². The pressures (mm Hg) were: right atrium, 10.5; right ventricle, 46/8-16; pulmonary artery, 40/20; wedge mean, 20; left ventricle, 164/16-28; aorta 100/70, with a peak subvalvular gradient of 64 mm Hg. In addition, postectopic pauses produced a marked increase in left ventricular pressures, aggravation of the subaortic gradient and a decrease in aortic pulse pressure with accentuation of the mid-systolic dip. Angiography showed a grossly thickened and trabeculated left ventricular wall, “cavity obliteration” and mild mitral incompetence. The aortogram and coronary arteries were
normal. These findings were considered diagnostic of IHSS.

Following this investigation he was discharged and was symptomatically improved with digoxin and propranolol. Six months later he was readmitted because of dizzy spells and an electrocardiogram showed complete heart block with an idioventricular rate of 32 beats per minute. Digoxin and propranolol were discontinued and seven days later a permanent unipolar ventricular inhibited pacemaker was placed in a right pectoral pouch and the lead passed percutaneously to the right ventricular apex through the internal jugular vein. For the past six months he has remained well and serial electrocardiograms have shown complete dependency on the pacemaker discharging at a rate of 70 beats per minute.

**Echocardiographic Findings**

The studies were performed using a Picker echocardiograph equipped with a 2.25 MHz transducer with an active diameter of one-half inch. The M-mode signals were fed into a 6-channel Cambridge fiberoptic physiological recorder which also recorded on strip chart the simultaneous phonocardiogram (PCG), electrocardiogram (ECG), and carotid pulse tracing (CAP). The PCG was recorded from the apex using medium frequency range.

The examination concentrated on the echocardiographic assessment of left ventricular outflow obstruction manifested by SAM, and abnormal movements of the aortic valve cusps, correlated with simultaneous changes in the systolic murmur and the carotid pulse tracing. Because the patient was entirely pacemaker dependent, cycle length was constant and the effect of atrial contraction could be studied by observing the P wave in relation to the other recorded parameters beat by beat.

**Aortic Valve Echograms**

These were always abnormal showing premature partial closure of the anteriorly visualized cusp and a series of irregular posterior and anterior movements during systole. The timing of the most obvious posterior displacement of this cusp, indicative of premature closure, correlated fairly well with the other parameters of obstruction to left ventricular outflow such as the peak point of intensity of the systolic murmur and the amplitude of the carotid pulse tracing. Because the patient was entirely pacemaker dependent, cycle length was constant and the effect of atrial contraction could be studied by observing the P wave in relation to the other recorded parameters beat by beat.

**Mitral Valve Echograms**

Similar findings were observed from a study of mitral valve movement which also showed previously undescribed unusual and interesting movements of the leaflets in diastole.
SAM, simultaneously softening the systolic murmur and widening the carotid pulse tracing.

In diastole, the pattern of movement of the mitral valve leaflets varied according to the timing of the P wave. When the P wave preceded the QRS complex at an interval of 0.22 sec the usual early atrial reopening movement (A) was observed so that at the onset of systole the leaflets had deflated and were almost apposed, their excursion was therefore minimal and the first heart sound was correspondingly softer (first complex, fig. 3). In those cycles where the P wave was dissociated from the paced beats, the leaflets reopened passively and late, so that at the onset of ventricular systole the leaflets were widely separated, their excursion maximal and the first heart sound was therefore loud (remaining complexes, fig. 3).

Late passive reopening was of greatest amplitude when the P wave occurred just after the mitral valve opened (E point) shortly after S$_2$ (fig. 4). The time interval from such a P wave to the peak of passive reopening (RO) was 0.40 sec compared to the normal P to A time of 0.08–0.12 sec, where A is a consequence of active atrial reopening of the valve.

**Discussion**

With the exception of left anterior hemiblock, conduction defects in IHSS are rare. Excluding those cases of complete heart block induced by operation, we have been able to find four reported instances among series as large as 300 cases (C. M. Oakley, personal communication). Apart from our patient and the case of a 10-year-old boy whose gradient was only manifest after isoproterenol stimulation, the remaining patients were elderly (60, 60 and 71 years, respectively).
Therefore, spontaneous heart block may be either a result of coincidental congenital heart block or senile degeneration of the conduction system rather than a complication of IHSS.

Complete heart block or atroventricular dissociation complicating IHSS provides an opportunity for studying the hemodynamic effects of the loss of the normal relationship between atrial and ventricular mechanical events. In the few studies performed, the data obtained at cardiac catheterization have been somewhat complicated by variation in cycle length (apart from case I of Matlof et al.).8, 10, 11 Long R-R intervals not only increase ventricular volume (preload) but also reduce aortic diastolic pressure (afterload), factors which independently decrease or increase the subaortic gradient, respectively. However, Matlof et al.7 and Johnson and Daily11 have demonstrated in two cases that a properly timed atrial contraction in paced complete heart block decreases the subaortic gradient. This finding is consistent with the concept that increasing ventricular volume by distending the left ventricular outflow tract decreases the subvalvular gradient.1

In our study, cycle length and therefore afterload, was constant and the only variable was the timing of atrial contraction. Appropriately timed atrial systole not only normalized the echocardiographic pattern of aortic valve movement but diminished the degree and duration of SAM and also simultaneously softened the systolic murmur, widened the pulse tracing and diminished its mid-systolic dip. These findings are consistent with a decrease in subaortic gradient due to augmented ventricular end-diastolic volume provided by atrial contraction.

Hemodynamic measurements were not made in our non-invasive study and the echocardiographic assessment of a decrease in subaortic gradient is qualitative rather than quantitative, particularly in relationship to SAM. King et al. have shown that echocardiographic stenosis may occur without hemodynamic obstruction or gradient across the outflow tract; this inconsistency results from uneven septal apposition by the anterior mitral leaflet.19 However, the changes in SAM, taken in conjunction with the aortic leaflet echogram, carotid pulse tracing and phonocardiogram were highly suggestive of changing degrees of outflow tract obstruction dependent upon the timing of the atrial systole.

The variation in intensity of the first heart sound found in our case was clearly related to the diastolic movement of the mitral valve which in turn was dependent on the timing of atrial systole. A properly timed atrial systole produced early active reopening of the mitral valve, allowing time for the leaflets to refloat almost to closed position at the onset of ventricular systole resulting in a normal closure sound. The loud closure sounds were produced by later reopening of the valve so that at the onset of ventricular systole leaflet excision was maximal and the closure sound correspondingly loud.

We believe that this late reopening of the valve (RO) is passive in nature and unrelated to the timing of the P wave. A passive movement of the mitral valve has been shown to occur normally in sinus rhythm by Zaky et al.14 in their figure 3; following initial opening of the mitral valve to the E point the leaflet undergoes a fast closing movement. However, if diastole is long enough the leaflet may reopen passively before being thrown open to a wide-open position by atrial contraction at point A. Edler et al.18 have shown that the interval between the start of the P wave of the ECG and the A wave of the echocardiogram is 0.08 to 0.12 sec. Using this measurement, active and passive reopening of the mitral valve can be clearly identified in complete heart block uncomplicated by IHSS. The passive reopening movement becomes most evident as the P waves occur earlier and earlier in diastole (fig. 5).

In our case, the measured delay from the start of the P wave of the ECG to the A wave of the echocardiogram was 0.12 sec (first complex fig. 3). Allowing for this delay in the next complex where the P wave is closely related to S1, atrial systole occurs just after the mitral valve swings open to the E point during the period of rapid ventricular filling; the peak of the reopening wave, however, occurs 0.44 sec after the start of the P wave making a direct relationship unlikely. This reopening movement was always evident on the echo of the posterior leaflet even when the P waves fell during ventricular systole. Passive reopening of the anterior mitral leaflet was only observed, however, when atrial systole fortuitously occurred at the onset of ventricular diastole as described above. Since the pattern of movement of the posterior mitral leaflet is less sensitive to changes in flow than that of the anterior,18 it is presumed that early augmentation of ventricular filling accelerates early refloating of the
anterior leaflet as in uncomplicated complete heart block (fig. 5) which is then reopened passively and later, independently from dissociated P waves as ventricular filling is completed in mid-diastole.

Only properly timed atrial systoles preceding the paced beats had a significant effect in alleviating obstruction to left ventricular ejection, presumably by producing a significant increase in left ventricular end-diastolic volume and dimension. Although this measurement was not made in our case, Johnson and Daily have demonstrated by echocardiography that appropriately timed P waves in IHSS and complete heart block do, in fact, increase the left ventricular end-diastolic dimension.11 Our patient tolerated ventricular inhibited pacing well, but in elderly patients with long-standing disease, atrial synchronous ventricular pacing may be preferable when the left ventricle is less compliant and highly dependent upon the atrial booster pump.11

References
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Complete heart block in a case of idiopathic hypertrophic subaortic stenosis: noninvasive correlates with the timing of atrial systole.
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_Circulation_. 1977;55:418-422
doi: 10.1161/01.CIR.55.2.418
_Circulation_ is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231
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Print ISSN: 0009-7322. Online ISSN: 1524-4539

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