Echocardiographic Abnormalities in the Mitral Valve Prolapse Syndrome

By Richard L. Popp, M.D., Owen R. Brown, James F. Silverman, M.D., and Donald C. Harrison, M.D.

SUMMARY
Two echocardiographic patterns of the mitral valve have been found to correlate with anatomic and/or angiocardiographic appearance of redundancy of the mitral valve. The echocardiographic abnormality may be found in the presence or absence of auscultatory findings, while the apparent separation of the anterior and posterior mitral leaflets on echocardiogram does not seem to represent anatomic separation leading to mitral regurgitation. Since the echocardiographic abnormality is found in the absence of various parts of the syndrome, it may be used as a tool to diagnose the mitral valve prolapse syndrome, discover its incidence and follow its natural history.

Additional Indexing Words:
Ultrasound
Midsystolic click-late systolic murmur
Mitral regurgitation
Floppy valve syndrome
Angiocardiography
Phonocardiography

Pulsed Reflected Ultrasound, or echocardiography, was first used by Shah and Gramiak1 to study patients with the clinical syndrome of mitral valve prolapse. Echocardiograms from patients with the midsystolic click-late systolic murmur syndrome have also been reported by Kerber, Isaeff and Hancock2 and by Dillon et al.3 In each of these preceding series, patients were known to have specific clinical auscultatory findings, and inferences regarding the mechanism of murmur generation were made from the echograms, invoking apparent separation of the mitral valve leaflets to account for the mitral regurgitation. However, by screening patients prior to cardiac catheterization, a spectrum of clinical features has been found in patients with the echographic findings of mitral valve prolapse. This report describes a group of 20 patients studied clinically, with phonocardiogram and echocardiogram, and having confirmation of mitral valve prolapse by cardiac catheterization, left ventricular angiocardiography and, in six cases, by surgery.

Materials and Methods
The twenty patients selected for analysis were those undergoing diagnostic cardiac catheterization and left ventriculography from September 1971 to May 1972 at Stanford University Medical Center, in whom echocardiograms suggested mitral valve prolapse. All patients admitted for cardiac catheterization at Stanford University Hospital undergo echocardiographic and phonocardiographic studies one to two days prior to catheterization.

Echocardiograms were performed using a Smith-Kline Ekoline 20 Mark II or IIa ultrasonoscope and 2.25 MHz transducer of 0.5 inch diameter, with acoustic lens collimating the sound beam for 5 cm tissue depth. The echocardiograms were recorded on Polaroid photographs of the oscilloscope or strip chart recordings, using an Electronics for Medicine DR8 recorder or Honeywell #1856 visicorder. The patients were studied in the supine position, with the transducer placed on the chest wall at the left sternal border. The interspace chosen would allow recording of the anterior and posterior mitral valve leaflets with the transducer oriented nearly perpendicular to the chest wall. Care was taken to record the anterior and posterior mitral valve leaflets near their free edges. By rocking the transducer at one position on the chest wall, the anterior leaflet was recorded from its annular insertion to its free edge. Recording the posterior leaflet was accomplished by rocking the transducer to encompass the area from the postero-lateral left atrium across the mitral annulus to the posterior left ventricle. In addition, in most cases.

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the transducer was placed in the interspace below that just described, so that the sound beam could be directed through the mitral valve and into the left atrium.

Phonocardiograms were recorded using a Maico contact microphone placed at the cardiac apex and filtered to display frequencies of 120 to 500 Hz and 40 to 200 Hz. Angiocardiograms were performed at approximately 30° right anterior oblique projection, using 0.5-0.7 cc/kg contrast material injected at 10-12 cc/sec. Cineangiocardiograms were filmed at 60 frames/sec.

Results

The results of this study are summarized in Table 1. Thirteen patients showed the echocardiographic abnormality which has been previously reported in the midsystolic click-late systolic murmur syndrome.1-3 That is, during systole the normal pattern of gradual anterior migration of the closed mitral leaflets (fig. 1) is replaced by the pattern of an initial horizontal, slight anterior or posterior motion, followed by an abrupt posterior motion, which results in a configuration that resembles a question mark turned approximately 90° clockwise (fig. 2).2, 3

In 12 of these 13 patients, the association of a mid- or late systolic click or late systolic murmur or combination of midsystolic click and late systolic murmur could be heard and recorded. These findings on the phonocardiogram were variable during a single examination in five of these twelve patients, while the echocardiographic findings were constant. One of the 13 patients showed the echocardiographic pattern just described, but in this patient no phonocardiographic abnormality could be found, despite changes in position, amyl nitrite administration and performance of Valsalva’s maneuver (fig. 3). This patient was being evaluated for chest pain, and the echocardiographic abnormality was an un-

Table 1

<table>
<thead>
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<th>No. pts.</th>
<th>Echocardiogram</th>
<th>Phonocardiogram</th>
<th>Angiocardiogram</th>
</tr>
</thead>
<tbody>
<tr>
<td>12</td>
<td>MSN</td>
<td>MSC, LSM</td>
<td>MV prolapse</td>
</tr>
<tr>
<td></td>
<td></td>
<td>MSC-LSM (5 variable)</td>
<td>(7 mitral regurgitation)</td>
</tr>
<tr>
<td>1</td>
<td>MSN</td>
<td>0</td>
<td>MV prolapse</td>
</tr>
<tr>
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<td>Pan-systolic murmur</td>
<td>MV prolapse and mitral regurgitation</td>
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<tr>
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<td>ESPM</td>
<td>Multiple clicks</td>
<td>MV prolapse</td>
</tr>
<tr>
<td>1</td>
<td>ESPM</td>
<td>0</td>
<td>MV prolapse</td>
</tr>
</tbody>
</table>

Abbreviations: MSN = midsystolic notch; MSC = midsystolic click; LSM = late systolic murmur; MV = mitral valve; ESPM = early systolic posterior motion; 0 = no abnormality.

Figure 1

Mitrval valve echogram from a clinically normal subject. The systolic pattern is of gradual anterior motion toward the transducer (top). Two echoes with the waveform of the anterior leaflet (al) are recorded. One of these echoes (white arrow) is more intense but has less excursion than the second echo. During systole there is apparent "separation" of these echoes. Although the echo showing the posterior leaflet (pl) waveform is near the second anterior leaflet echo during systole, some variable separation is seen (black arrow). The posterior leaflet could be recorded only with the left ventricular posterior wall (LVPW) posterior to it. IVS = interventricular septum; ECG = electrocardiogram.

Figure 2

Simultaneous electrocardiogram (ECG) lead II, apex phonocardiogram (PHONO) and echocardiogram from a patient with a late systolic murmur, T wave abnormalities and angiographic mitral valve prolapse. The anterior mitral valve (MV) leaflet echoes meet the posterior leaflet echoes at the onset of systole (black arrow). These echoes move slightly toward the chest wall (CW) in early systole, but show an abrupt posterior motion toward the left atrium (LA) in late systole, coincident with the recording of the murmur (sm). Note the multiple echoes during systole. IVS = interventricular septum.

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Simultaneous electrocardiogram (ECG) lead II, phonocardiograms (PHONO) with 120-500 Hz band-pass filters from second interspace at left sternal border (PULM) and apex, and echocardiogram. During diastole, echo waveforms of both anterior and posterior mitral leaflets were recorded. Despite apparent separation of the echoes, no systolic murmur was recorded (and none could be heard). The left atrium is recorded posterior to the valve echoes.

The valve. Five of these seven patients had a pansystolic murmur recorded on the phonocardiogram, while one patient showed multiple systolic clicks without murmur (fig. 5). In the remaining patient, with radiographic dilation of the pulmonary artery, the early systolic posterior motion was recorded on the echogram, but no phonocardiographic abnormality could be found with position change, amyl nitrite administration, or Valsalva’s maneuver.

More than two echoes were recorded from the mitral valve leaflets in all patients, and the number of apparent mitral leaflet echoes recorded during systole was greater in these patients than in normals. Both anterior and posterior mitral leaflets were recorded while directing the sound beam into the left atrium in six patients.

All patients had the angiocardiographic picture of mitral valve prolapse (fig. 6). Twelve of the twenty patients had associated mitral regurgitation. There was no correlation between angiocardiographic mitral regurgitation and the apparent separation of the multiple echoes from the mitral valve leaflets after their meeting at end-diastole (the C-point). The diagnosis of “floppy mitral valve” was confirmed at surgery in six patients.

Discussion

In normal patients, the basic systolic mitral waveform is that of gradual anterior migration toward the chest wall (fig. 1).2,3 This seems to be the result of reduction in volume of the right and left ventricles during systole. Consequently, the closed mitral leaflets are closer to the chest wall at end-systole than at end-diastole, although they have not undergone motion independent of the mitral annulus.4 In high-quality echocardiograms, it is possible to see a small initial posterior motion of these leaflets at the onset of systole, which lasts for less than 0.05 seconds (fig. 1). In the patients with mitral valve prolapse, the echogram records posterior motion of the leaflets during systole, which
may be biphasic, forming a midsystolic notch, or monophasic, which is described here as showing posterior motion for a prolonged period (>0.05 sec) but beginning with the onset of systole. Since the usual cyclic changes in ventricular volume still occur during systole, the pattern seen in all of these cases is one of end-systolic motion towards the chest wall before the valves move to their open positions. This suggests that the leaflets reach their most posterior position much later in systole than is normally seen, but after they cease their prolapse toward the left atrium, the ongoing annulus motion toward the chest wall is apparent.

With amyl nitrite administration, the midsystolic notch occurs earlier in the cycle, appearing very similar to the early systolic prolapse which is reported here as the second echographic pattern associated with the angiocardiographic abnormality. Since both patterns have been seen in the same patient, it is felt that the echogram is recording two forms of the same basic abnormality (fig. 7). As there is posterior motion of the leaflets after closure (C point) in some normal patients, it is proposed that the duration of this posterior motion is important for the proper diagnosis of prolapsing leaflets. This suggestion is supported by angiocardiographic observation of a prolonged “rolling” or “inflation” of the redundant leaflets during the early phases of systole. It is this temporal prolongation of posterior motion during systole, as well as the amplitude of the posterior motion toward the atrium, which distinguishes these patients by echocardiography (fig. 6).

This posterior motion during systole leads to an abnormally high opening excursion of the anterior leaflet, which previously has been cited as an echographic criterion for mitral regurgitation. While this finding is associated with mitral valve prolapse, it does not indicate mitral regurgitation in all cases, as shown in this series. The early systolic prolapse pattern is commonly associated with a pansystolic murmur of mitral regurgitation, but it may be present in association with multiple systolic clicks (fig. 5), and in several patients not studied by angiocardiography this waveform has been associated with variable auscultatory findings in this laboratory. Apparent separation of anterior and posterior leaflet echoes may be recorded without auscultatory (fig. 3) or angiocardiographic findings of mitral regurgitation.

Since the mitral valve leaflets are not perfectly smooth and the sound beam is relatively broad, it is possible to get discrete echoes from multiple areas of the same leaflet. This is seen very commonly in normal patients and can be confirmed on strip chart records showing a low amplitude mitral waveform which originates from the valve near the area of the mitral annulus, as well as a higher amplitude waveform which originates near the free edge of the valve undergoing the greater excursion (fig. 8). When the anterior mitral leaflet is tangential to the relatively broad sound beam, separate parts of the leaflet will appear to be at different depths from the transducer and give the appearance of separate structures. It is suggested that the redundancy and interchordal “hooding” which is commonly seen in surgical and autopsy specimens leads to multiple echoes in such a valve (fig. 9). These multiple curved surfaces will reflect sound back to the transducer from multiple depths re-

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**Figure 6**

Left ventricular angiocardiogram, right anterior oblique projection. Left, early systolic frame, mitral valve is closed. Right, late systolic frame showing more posterior inferior position of mitral valve shadow characteristic of mitral valve prolapse. Note the apparent increase in the extent of prolapse during systole. (This patient had aortic root dilatation as well.)

**Figure 7**

Simultaneous pulmonary area (PA) and apex phonocardiogram (PHONO), lead II electrocardiogram (EKG) angiocardiogram (ACG) and mitral valve (MV) echocardiogram from a patient with spontaneous premature ectopic beats. Note the midsystolic notch and mid-late systolic murmur in the sinus complexes. The early beat (third beat) shows early systolic posterior motion and a full length systolic murmur. (The apexcardiogram is technically poor.)
Echographic strip chart recording of the left ventricular outflow tract obtained by slowly rocking the transducer from one point on the chest wall. The beam is swept (left to right) from the body of the left ventricle (LV), below the mitral valve leaflets, denoted by recording echoes from chordae tendineae (c), to the area of the aorta (AO), with its intraluminal cusps and left atrium (LA) posterior to it. Note the relatively intense anterior mitral echo (white arrow) which is first recorded near the aortic root. A similar waveform is seen in the less intense but higher amplitude anterior mitral echoes which are first recorded further from the aortic root. The C-point is denoted by the black arrow. This patient had coronary artery disease but did not have valve prolapse on the angiogram. P = pericardium, ECG = electrocardiogram.

gardless of their orientation to the sound beam. Therefore, multiple parallel echoes may be seen on the echocardiogram, but this does not necessarily represent separation of the leaflets. In addition, since chordae tendineae from each papillary muscle insert into both the anterior and posterior leaflets, it would be very difficult for one leaflet to separate from the other unless the chordae tendineae have become detached or extremely elongated, so that one leaflet may "overshoot" the other. It may be these abnormal surfaces (hooding) that lead to abnormal coaptation of the anterior and posterior leaflets, resulting in mitral regurgitation. Elongated chordae tendineae may permit the posterior leaflet to move toward the atrium and decrease the surface area in contact with the anterior leaflet, but actual separation of the anterior and posterior leaflets during systole, in a way which would allow recognition with the ultrasound technique, seems distinctly unlikely. Since the anterior leaflet abuts the posterior leaflet during systole, it is expected that anterior or posterior leaflet prolapse will give an abnormal echocardiographic waveform from both leaflets. It is extremely uncommon to find only the anterior leaflet or only the posterior leaflet involved in pathologic specimens, as the abnormal process does not stop at the commissures of the valve.

The echographic pattern of a midsystolic notch is striking and quite easy to recognize. The only point of confusion here could be with the echographic pattern of idiopathic hypertrophic subaortic stenosis (IHSS). In a few cases the initial systolic motion of the mitral valve in the prolapse syndrome may be slightly anterior, and in combination with the midsystolic notch it simulates the midsystolic anterior motion associated with IHSS. However, the late systolic position of the mitral valve in the prolapse syndrome is posterior to the C point, while in IHSS the end-systolic position is anterior to the C point. It has been shown that anyl nitrite administration will move the position of the midsystolic notch earlier in systole, and administration of this substance can resolve any question regarding the presence of mitral valve prolapse versus IHSS. The abnormal "U-shaped" early systolic pattern may be simulated by the mitral valve echogram of patients with very low output states, such as that seen in cardiomyopathies. In some of these patients, there is so little change in volume during systole that the anterior migration of the mitral valve may be nearly imperceptible. An additional factor in these dilated hearts is that decreasing left ventricular volume during systolic ejection allows the stretched mitral valve leaflets to move toward the left atrium and simulate the prolapse pattern. However, it is a posterior motion of the leaflets after the C point which is the hallmark of this abnormal pattern associated with prolapse.

So far, we have not seen the echographic pattern of prolapse without angiographic or anatomic findings of this condition when such correlations have been available. Because of technical factors, it may be possible to miss the diagnosis of prolapse echocardiographically when it is present anatomically. However, the recognition of the early systolic prolapse reported here should decrease such false-neg-
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ative results. It is interesting to note that echoes from both anterior and posterior mitral valve leaflets could be recorded clearly in six patients with the sound beam directed up through the valve area and into the left atrium (fig. 2). This suggests that such valves do seat more toward the left atrium than is the normal case, since simultaneous records of the posterior leaflet and left atrium are quite unusual in other groups of patients (fig. 1). However, due to the normal anterior systolic motion of the left ventricular posterior wall, it may be difficult to recognize the posterior systolic motion of the mitral valve on the echogram if the sound beam records these two structures simultaneously. For this reason it is suggested that the transducer be placed lower than the usual position on the chest wall, so that the sound beam will pass up through the mitral valve structures and into the atrium.

This study demonstrates that the echographic abnormality correlates with the angiocardiographic and anatomic abnormalities of the mitral valve prolapse syndrome. This correlation is found in the absolute or temporary absence of phonocardiographic abnormalities. Because of the variable clinical presentation of patients with this syndrome, the constancy of the echographic abnormality may be an important diagnostic feature. The syndrome includes electrocardiographic abnormalities, atrial and ventricular arrhythmias, chest pain which may be quite severe in nature, as well as variable auscultatory features. While it cannot be stated from this study that the mitral valve abnormality is the primary underlying factor in all of these cases, we have seen echographic prolapse in patients presenting with only arrhythmia or only chest pain (with normal coronary arteriograms), as well as the suggestive auscultatory features. Because of the recent evidence that propranolol may be the treatment of choice for the arrhythmias associated with this condition, it is important to recognize the association of the prolapse syndrome with the patient's arrhythmia. The echographic pattern of prolapse is frequently found in our laboratory, and since there is a high prevalence of atherosclerosis in our population, chest pain in association with this echocardiographic abnormality certainly cannot rule out coronary artery disease.

With this noninvasive tool, it may be possible to diagnose mitral valve prolapse before it becomes clinically apparent in patients with minimal auscultatory or clinical findings, so that recommendations regarding antibiotic prophylaxis and possible antiarrhythmic prophylaxis may be made. In addition, now it should be possible to recognize the true incidence and follow the natural history of this condition in order to learn more about the various aspects of the whole syndrome.

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