The Variable Spectrum of Echocardiographic Manifestations of the Mitral Valve Prolapse Syndrome

By Anthony N. DeMaria, M.D., James F. King, M.D., Hugo G. Bogren, M.D., James E. Lies, M.D., and Dean T. Mason, M.D.

SUMMARY

The variety of echographic features associated with the mitral valve prolapse syndrome (MVPS) is not yet completely understood. Therefore, ultrasound recordings were obtained in 33 patients in whom mitral prolapse had been documented by biplane left ventricular cineangiography. Echographic abnormalities of the mitral leaflets during systole were recorded in 26/27 MVPS patients and 6/6 with ruptured chordae tendineae. In MVPS, the midsystolic mitral buckling, emphasized in early echocardiographic studies, was observed in only 12 patients. In our study, the most common aberrancy was abnormal pansystolic mitral motion in 14 patients, which in 12 was similar to the pansystolic bowing observed in all six patients with torn chordae. An additional echographic abnormality in MVPS was localized mitral collapse throughout systole in 10/14 patients with pansystolic prolapse; this finding was the most striking defect noted in five, in two of whom it was the only disturbance. Phonocardiography in MVPS showed typical midsystolic click and/or late systolic murmur in only 15/26 patients of whom ten had midsystolic mitral buckling. A variety of systolic clicks and/or murmurs occurred in the 14 patients with generalized bowing and/or localized collapse throughout systole on echocardiography. Thus, the mitral echographic spectrum of MVPS is comprised of three different abnormal patterns of systolic prolapse: buckling in midsystole, pansystolic bowing, and pansystolic collapse. These echocardiographic disorders commonly occur in the absence of classical auscultatory findings in MVPS and the most frequent abnormality on ultrasound is pansystolic bowing of both mitral leaflets.

Additional Indexing Words:

Idiopathic dilatation of pulmonary artery  Phonomography  Marfan’s syndrome
Ruptured chordae tendineae  Mitral myxomatous degeneration  Straight back syndrome
Mitral regurgitation

Although the Syndrome of mitral valve prolapse (MVPS) with regurgitation has been recognized for only a few years, there has been considerable interest in the prevalence and important clinical aspects of this disorder. While the combination of midsystolic click with late systolic murmur serves as the auscultatory hallmark of MVPS, this syndrome may be accompanied by the less indicative finding of isolated early or midsystolic click, a non-specific pansystolic murmur, or even the absence of abnormalities on physical examination. Since special therapeutic, prognostic, and genetic factors must be considered in the mitral prolapse syndrome, precise identification of this condition is essential.

Echocardiography has recently provided a non-invasive means for the objective detection of mitral valve prolapse. Thus abrupt midsystolic backward displacement of the posterior or both mitral leaflets has been described as the characteristic feature of MVPS by ultrasound. The typical description of mitral regurgitation due to ruptured chordae tendineae, in contrast, has been acknowledged to be pansystolic prolapse with diastolic flail of the mitral valve, in contrast, has been acknowledged to be pansystolic prolapse with diastolic flail of the mitral valve. However, little attention has been directed to the occurrence of pansystolic posterior movement of the mitral valve in the mitral valve prolapse syndrome. The purpose of this report is to delineate the variable spectrum of echographic features in MVPS in patients with angiographic documentation of this condition.

From the Section of Cardiovascular Medicine, Department of Medicine, Physiology and Radiology, University of California, School of Medicine, Davis and Sacramento, California.

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Address for reprints: Anthony N. DeMaria, M.D., Section of Cardiovascular Medicine, University of California, School of Medicine, Davis, California 95616.

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and to emphasize the high frequency with which mitral prolapse may present as pansystolic bowing on the echogram.

**Materials and Methods**

Thirty-three patients undergoing right and left heart catheterization with biplane left ventricular cineangiography form the basis of this study. The group was comprised of 22 females and ten males with an age range of 16 to 56 years and mean age of 38 years. The reason for evaluation was chest pain in 18 patients, effort dyspnea in eight, murmur in three, arrhythmias in two, and prominent pulmonary artery in one.

Biplane left ventricular cineangiography was performed in the 30° right anterior (RAO) and 60° left anterior oblique projections on 35 mm film taken at 64 frames per second using a Philips image-intensifier system with 9 inch magnification. The ventricle was opacified with 0.75 to 1.00 cc/kg body weight of Hypaque M, 75% (Winthrop) containing 25% sodium diatrizoate and 50% meglumine diatrizoate injected at 300 psi. Mitral valve prolapse was documented in all MVPs patients in the RAO view by backward displacement of the mitral valve into the left atrium in ventricular systole.1, 2, 7, 20 Figure 1 illustrates a representative cineangiogram obtained in the right anterior oblique projection in one MVPs patient. Scalloped protrusion of the mitral valve is observed to the mitral annulus.

Echocardiography was performed in the supine position with a commercially available echograph utilizing a 0.5 inch diameter 2.25 megahertz transducer focused at 10 cm with a repetition rate of 1000 per second.1 The ultrasound tracing was displayed and recorded on a multichannel oscilloscope recorder† and none of the echograms illustrated has been retouched. In each case care was taken to obtain recordings in the echo sector traversing the mitral valve and left atrium.21

Phonocardiography was performed utilizing a Sanborn microphone from which the signal was directed into the phono channel of the multichannel oscilloscope recorder†. Recordings were performed with the microphone positioned at the cardiac apex, in the fourth intercostal space along the left sternal border, and in the second intercostal space along both right and left sternal borders at frequencies ranging from 50 to 500 Hz.

**Results**

Normal Echogram

Appreciation of the spectrum of echocardiographic manifestations of mitral valve prolapse is facilitated by clear understanding of the echo pattern observed in normal individuals. Figure 2 demonstrates a representative normal echogram obtained in a subject in whom the absence of heart disease was confirmed by cardiac catheterization and angiography performed for the evaluation of chest pain. Illustrated are

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†Model DR8, Electronics For Medicine, White Plains, New York.
superiorly to the left atrium (right side of figure), the posterior leaflet is no longer visible. At the sector of the left atrium, the anterior mitral echo is recorded considerably forward of the atrial posterior wall echo throughout ventricular systole.

**Mitral Valve Prolapse**

Echographic abnormalities were detected in 32 of the 33 patients with angiographically documented mitral prolapse. The patients were grouped according to the presence of midsystolic or pansystolic prolapse on echogram.

**A. Midsystolic Prolapse in MVPS**

Twelve patients with the MVPS revealed mid to late posterior break of one or both mitral leaflets on echogram. This diagnostic pattern of MVPS has previously been emphasized to be the typical abnormality in this condition. Figure 3 demonstrates a representative echogram obtained from one of our 12 patients with this abnormality. At the onset of systole, the anterior and posterior mitral leaflets joined and pursued a horizontal course to mid systole at which time both leaflets buckled posteriorly. Subsequently, during isovolumic ventricular relaxation, the leaflets moved together anteriorly. They then separated in a normal manner during ventricular filling. However, there was increased forward excursion of the anterior leaflet so that it closely approximated the interventricular septum during early diastole.

In six of these 12 patients, the mitral echo during the first portion of systole prior to the abrupt posterior buckle, exhibited a straight or gradual downward course (fig. 4), in contrast to the upward ascent observed normally. In regard to the sudden mid-systolic mitral break, this motion involved both leaflets in ten patients and only the posterior leaflet in two patients.

Phonocardiograms revealed a midsystolic click and/or late systolic murmur in ten of the 12 patients with echographic midsystolic prolapse. Of the remaining cases, one patient had an early systolic click with midsystolic murmur (fig. 3), while the other had only a pansystolic murmur.

**B. Pansystolic Prolapse**

1. **Ruptured Mitral Chordae Tendineae.** Six of the 33 patients exhibited the characteristic clinical and catheterization features of torn mitral chordae tendineae. Surgery confirmed this diagnosis in four cases. All six patients were found to have pansystolic prolapse on echogram accompanied by diastolic flail motion of the anterior or posterior leaflet. In three patients, the systolic prolapse involved both leaflets. Figure 5 demonstrates the echographic configuration in one of these patients. Throughout systole multiple echoes were recorded, all of which had a posterior hammock-like contour with superior concavity, rather than the gradual forward movement seen in normal echograms (fig. 2). In addition, the completely flailed aspect of a portion of the posterior leaflet was evident in diastole from the anterior movement from its abnormal position in the left atrium. In the three patients in whom pansystolic prolapse involved the anterior leaflet predominantly, this structure exhibited markedly exaggerated excursion and erratic motion in diastole as has been described previously.

Phonocardiography revealed high intensity holosystolic murmurs without clicks in all of the patients in this group.

2. **Mitral Valve Prolapse Syndrome.** In 14 patients with the mitral prolapse syndrome, pansystolic mitral prolapse within the left atrium was recorded on echogram. The pansystolic prolapse involved both leaflets in 13 of these patients. The typical echographic finding was mitral bowing throughout systole recorded from a wide area of the left atrium as shown in figure 6 from a patient in whom surgery confirmed mitral prolapse secondary to mucinous degeneration. In addition, multiple pansystolic echoes emanated from the mitral valves and there was markedly accentuated excursion of the anterior leaflet in early diastole. The pattern of pansystolic bowing is further exemplified in figures 7 and 8A from two young adult females with the common clinical and classic catheterization manifestations of the mitral prolapse syndrome. These echograms demonstrate multiple layers of pansystolic echoes with bowing acquisition.

![Figure 3](image-url)
Figure 4

Echogram in MVPS patient R.S. showing posterior movement of the mitral leaflets from the onset of ventricular systole which is accentuated by mid-systolic buckling (arrows). Note also the multiple layers of mitral echoes during systole. RV = right ventricle. See figure 2 for other abbreviations.

posteriorly and increased mitral valve excursion during ventricular filling.

Figure 9 from a patient with the Marfan habitus demonstrates certain additional features of the variable echographic manifestation of pansystolic mitral prolapse in MVPS. Figure 9A shows the sweep of the echo scan across the left ventricle and left atrium. In the inferior portion of the atrium near the left ventricle, there was typical posterior bowing of both mitral leaflets immediately following the onset of systole which persisted throughout ventricular contraction. However, a different abnormal configuration was obtained from a relatively limited area of the mid-left atrium as the echo scan was directed more superiorly. Thus, in this specific area, the two mitral leaflets coapted posteriorly adjacent to the left atrial wall throughout systole; in addition there was generalized increased motion of the left atrium during systole and diastole. Figure 9B emphasizes the echographic details of this midatrial sector showing localized mitral collapse. Note that the echoes from the anterior and posterior mitral leaflets indicate prolapse deep into the atrium as they coapt at the onset of systole. Regional mitral collapse in the

Figure 5

Representative echocardiogram obtained from patient L.C. with torn chordae tendineae demonstrating pansystolic prolapse of the mitral leaflets (left two-thirds of figure). The solid arrow on the left indicates a portion of the prolapsed posterior leaflet in systole which then moves anteriorly (flail) during diastole. As the echo beam is directed in the left atrium toward the aorta (right one-third of figure), echoes from the mitral leaflets continue to be recorded (broken arrow).
presence of generalized pansystolic prolapse is further exemplified by the patient shown in figure 8. In addition to generalized pansystolic bowing (fig. 8A), the anterior leaflet was noted to plunge posteriorly to the left atrial wall in a limited area of the echographic scan (fig. 8B). This localized pattern of complete backward descent of mitral leaflet coaptation was observed in ten MVPS patients, and was the major abnormality in five patients. In two of these patients, localized predominant collapse was the solitary disorder in systole (fig. 10) which was accompanied by generalized increased left atrial wall movement and accentuated mitral diastolic motion.

Phonocardiograms in pansystolic MVPS provided a variety of findings. Holosystolic murmurs were noted in only two patients. Midsystolic click and/or late systolic murmurs were recorded in five patients. Figure 6 illustrates one such patient in whom a predominantly late systolic murmur was recorded in the presence of pansystolic prolapse on echogram. Two patients with echographic pansystolic prolapse were found to have multiple early and midsystolic clicks with a late systolic murmur. An early systolic click was the solitary finding in four patients in this group (fig. 9A) and one patient was actually free of auscultatory abnormalities.

Discussion

There has been an increased appreciation in recent years of the frequent occurrence and clinical importance of the mitral valve prolapse syndrome. It is now clear that this entity is neither rare nor completely benign and may often result in a variety of symptoms. Thus occurrence of severe mitral regurgitation, idiopathic angina-like chest pain, idiosyncratic diastylemy, tachyarrhythmias, and sudden death has been well documented in MVPS. Therefore, the availability of an accurate and practical method for the detection of mitral valve prolapse is of considerable importance.

Previously the diagnosis of mitral prolapse has been based upon the auscultatory findings of mid-systolic click with late systolic murmur. However, it has recently been shown that aneurysmal protrusion of the mitral valve may be accompanied by a number of auscultatory features and may even occur in the absence of physical findings. Thus, it would appear that only a limited portion of patients with MVPS may be identified by physical examination. In addition, this condition has occasionally displayed a familial occurrence and silent or atypical prolapse may be particularly common among relatives of patients who demonstrate the classical features of the syndrome. Further, the importance of objective detection of occult MVPS by a noninvasive technique is emphasized by the increased incidence of sudden death among these patients.

Echocardiography has provided a precise atraumatic means for the identification of the mitral prolapse syndrome. Previous reports have described a midsystolic mitral leaflet buckling on echogram in many patients with midsystolic click and/or late systolic murmur. The present study extends this observation by delineation of the wide variety of mitral echographic abnormalities in angiographically-proved MVPS (fig. 1) in patients with atypical or absent auscultatory findings, as well as in individuals with typical phonocardiographic features. Thus our findings enhance the utility of echocardiography in
the objective detection of MVPS regardless of concomitant clinical manifestations. Further, this report emphasizes that a variable spectrum of mitral echographic abnormalities occurs among MVPS patients with typical auscultatory characteristics, as well as in those with atypical findings.

The echocardiographic abnormality that was initially reported in mitral prolapse was an abrupt, sharp, mid-systolic posterior buckling of one or both mitral leaflets\(^\text{15, 16}\) (figs. 3 and 4). This abnormality has received major emphasis and has formed the diagnostic cornerstone of this syndrome by ul-

Mitral echograms in MVPS patient M.P. Panel A shows pansystolic bowing of layered echoes recorded from the low left atrium. In panel B, the transducer is directed superiorly and rightward in the left atrium at which location the anterior leaflet is observed to plunge posteriorly at the onset of systole to approximate the left atrial wall. See figure 2 for abbreviations.

Mitral echograms in patient R.F. with Marfan’s habitus and early systolic click. Panel A illustrates an echographic scan from the left ventricle directed superiorly to the left atrium. Pansystolic mitral prolapse is demonstrated at the level of the atrioventricular junction (arrows). In the left atrial sector, the anterior mitral leaflet collapses posteriorly against the atrial wall. LVW = left ventricular wall; AO = aorta. Panel B emphasizes the details of the localized pansystolic collapse of the anterior leaflet in the left atrium.

Ultrasound recording in MVPS patient M.B. in whom localized predominant leaflet collapse was the solitary echographic abnormality of the mitral valve. The arrow indicates associated local mitral prolapse and/or layered echoes from the collapsed mitral leaflets.
transound. Pansystolic posterior hammock-like bowing of the mitral leaflets was observed subsequently on echogram in patients with torn mitral chordae tendineae37-39 (fig. 5). However, pansystolic backward bowing of the mitral leaflets without ruptured chordae has received little attention. In the present study, pansystolic posterior hammock-like mitral movement on echogram was documented to occur with mitral prolapse in the absence of clinical and catheterization evidence of ruptured chordae (fig. 6-8), and more importantly, actually represented the most common echographic manifestation of the mitral valve prolapse syndrome, being present in 14 of 26 (54%) such patients. The increased prevalence of pansystolic bowing in our MVPS patients as compared to other reports may be explained, in part, by the fact that previous studies reported patients in whom the diagnosis of mitral prolapse was based solely upon the presence of typical auscultatory features. However, even some of our patients with classical features of midsystolic click and/or late systolic murmur manifested pansystolic prolapse on echogram (fig. 6). Thus, when examining an echogram for evidence of mitral prolapse, one may expect to observe pansystolic bowing as often as midsystolic break. It should be noted, however, that since our findings were obtained from patients with signs and/or symptoms of MVPS, they may not fully represent asymptomatic patients.

Another important finding which emerged from this study was that rapid pansystolic posterior coaptation of the mitral leaflets adjacent to the left atrial wall was a common echographic manifestation of mitral prolapse (figs. 8-10). When scanning superiorly in the sectors traversing the mitral leaflets and left atrium, this movement was found in a limited area of the left atrium and was observed as a precipitous plunging or backward collapse of the anterior leaflet to meet the prolapsed posterior leaflet against the left atrial wall which exhibited increased excursion. This pattern of both leaflets against the posterior left atrial wall is best seen in the scan in figure 9A. The juxtaposition of the prolapsed posterior mitral valve against the left atrium rendered recognition of this leaflet difficult when the echogram was obtained from a narrow sector (figs. 9B and 10), thereby necessitating a vertical scan for its identification (fig. 9A). This localized abnormality was usually accompanied by generalized pansystolic bowing (fig. 8A and 9A), but in five patients in whom it was the predominant echocardiographic disorder (fig. 10), the mitral valve systolic motion was either flat or minimally bowed in other sectors of the mitral leaflets and left atrium.

The patients in whom this posterior position of mitral leaflet coaptation was the predominant abnormality formed a particularly interesting subgroup. All five patients manifested mild posterior mitral prolapse on angiogram without mitral regurgitation. In addition, all were noted to have musculoskeletal abnormalities suggestive of either the straight back syndrome31 or Marfan’s syndrome.32 Further, two were found to have idiopathic dilatation of the pulmonary artery,33 on standard roentgenogram and by catheterization with angiography. On cardiac auscultation, four patients exhibited solitary early systolic clicks while one had no abnormal physical findings. The possibility that this patient subset represents a normal variant in whom the mitral leaflets dome into the left atrium in systole cannot be completely excluded, but is considered unlikely. Rather the increased mitral excursion and prominent atrial movement on echogram, the presence of chest pain, and the musculoskeletal abnormalities which have been reported in increased frequency in the mitral prolapse syndrome,34 all suggest that these patients manifest a type of localized mitral prolapse, perhaps in some individuals a forerunner to more generalized mid- or pansystolic prolapse. In addition, the complex of cardiovascular and musculoskeletal abnormalities in these patients suggest that auscultatory abnormalities attributed to the straight back syndrome or idiopathic dilatation of the pulmonary artery frequently may actually represent mitral valve prolapse.

Although this study was not primarily designed to evaluate the acoustic events accompanying mitral prolapse in MVPS, several important observations were evident from the phonocardiograms performed. The initial association of aneurysmal protrusion of the mitral valve with the presence of midsystolic click and late systolic murmur established reference to this condition as the “midsystolic click-late systolic murmur syndrome.” Recent reports, however, have indicated that a solitary early systolic click may be the only abnormal auscultatory finding.16,35 Our data confirm this finding (fig. 9A) and, in addition, provide evidence for its mechanism of production since the observation that mitral prolapse may be initiated with the onset of systole would explain the occurrence of the resulting sounds at this time. Thus, early systolic nonejection clicks appear to occur occasionally in MVPS and usually indicate pansystolic prolapse.

Previous reports have noted the occurrence of the systolic click in MVPS to be simultaneous with the maximal prolapse of the mitral leaflets observed angiographically.2 It was of interest, however, that an early systolic click was recorded in one of our patients in whom the echocardiogram clearly revealed midsystolic posterior motion (fig. 3). It is possible that the click was produced by a portion of mitral leaflet which was prolapsing at the onset of systole not detected by
echo, although diligent search for such leaflet tissue was unrevealing. Also, the timing of prolapse on angiography in this patient revealed mid-systolic movement. Whereas an early click with late echo prolapse was observed in only one patient, the reverse of this situation, mid-click and/or late murmur with pansystolic echo prolapse was considerably more common and was noted in five patients (fig. 6). Angiography in these five patients revealed pansystolic posterior motion into the left atrium similar to that recorded on echogram and also confirmed that mitral regurgitation was responsible for the murmur occurring in mid to late systole. It was also of interest that neither the presence nor the degree of mitral regurgitation correlated with the ultrasound pattern of mitral prolapse. Thus, auscultatory events may not coincide temporally with mitral prolapse as recorded on echogram and angiography. These observations suggest that the chordae tendineae may be responsible for genesis of the systolic click in some MVPS patients and are consistent with the concept of “chordal snap.” Since the auscultatory features of MVPS may fluctuate, in the interpretation of these findings it should be recognized that the echo-angiographic correlations were not simultaneous.

Recently attention has been directed toward the frequency with which each mitral leaflet is involved in mitral prolapse. Original reports of this syndrome stressed prolapse of the posterior leaflet, but frequent involvement of the anterior leaflet has subsequently been noted. This question has been complicated by lack of agreement concerning the exact identification of prolapsing leaflets on angiogram, particularly recognition of anterior leaflet prolapse. Echocardiography provides certain advantages in the study of the anterior leaflet which may be difficult or impossible to identify during right anterior oblique left ventriculography. Conversely, the multiple echoes which are often reflected from the valve, particularly in mitral disorders, complicate precise differentiation of the two leaflets. Despite this limitation, both anterior and posterior leaflets or all echoes recorded in the absence of two distinct leaflets manifested abnormal posterior motion in 23 of our 27 MVPS patients (figs. 3-10). Thus it would appear that, from a functional standpoint, both mitral leaflets participate in the posterior prolapse in nearly all patients. The explanation for this phenomenon is probably related to the dependence of each leaflet segment upon the segment with which it coapts for proper function, and perhaps to chordae involvement with the disease process, since the frequency of both leaflets prolapsing on echogram is substantially greater than the occurrence of morphological abnormalities of both mitral leaflets.

Finally, concerning the genesis of the multiple echoes recorded from the prolapsed mitral leaflets in systole (figs. 3-8), it is pointed out that when a broad interface such as the mitral valve is aligned obliquely to the main axis of the ultrasonic beam, several successive echoes may be produced which are recorded on echogram immediately behind each other rather than the actual side-by-side orientation. Although systolic mitral layering may occasionally be observed normally, this phenomenon occurs more commonly in the various patterns of leaflet distortion in mitral prolapse. It is of interest that multiple systolic echoes are also frequently obtained for the same reason from the abnormally forward-moving anterior mitral leaflet in idiopathic hypertrophic sub-aortic stenosis in which the disturbance of mitral valve function during contraction is the antithesis of mitral prolapse.

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