Surgical Repair of Long-Standing Mitral Valve Prolapse: A Case Report and Brief Review of the Literature

Paul S. Pagel, MD, PhD; Nadia Haider, MD; Elizabeth G. Butler, MD; Alfred C. Nicolosi, MD

ABSTRACT
A 57-year-old patient presented with a long history of minimally symptomatic mitral valve prolapse (MVP). The patient eventually developed severe mitral regurgitation (MR) and clinical evidence of congestive heart failure over 10 years duration before admission. He described a 34-year history of MVP. A transesophageal echocardiography examination demonstrated left atrial enlargement and severe prolapse of the posterior mitral leaflet associated with severe MR to the dome of the left atrium. Left ventricular size and function were normal. A quadrangular resection of prolapse segment and placement of a posterior annuloplasty ring were used to successfully repair the valve using a minimally invasive approach. The current case emphasizes that patients with MVP complicated by MR may remain clinically asymptomatic for prolonged periods, only to subsequently develop left atrial enlargement, severe MR, and congestive heart failure late in the natural history of the disease.

INTRODUCTION
The natural history of mitral valve prolapse (MVP) is remarkably heterogeneous.1-2 Many patients with this common valvular disorder have a benign clinical course and normal life expectancy,3 but an unfortunate minority with mitral regurgitation (MR) or related complications often suffer substantial morbidity or succumb to its adverse consequences. Temporal progression of the MR severity resulting from MVP produces left atrial (LA) enlargement with or without atrial fibrillation, left ventricular (LV) dilatation, and impaired LV systolic dysfunction, which may culminate in irreversible congestive heart failure and death. Spontaneous rupture of chordae tendinae or infective endocarditis frequently exacerbates MR and contributes to the development of heart failure or the requirement for urgent surgical treatment.4 In addition, an increased incidence of stroke, ophthalmologic ischemia, and sudden cardiac death have been reported in patients with MVP.5-6 Several clinical investigations examined the natural history of the mitral valve prolapse and stratified patients who appear to be at highest risk of developing such complications.1,5-7-9 The results of these studies provided important prognostic information about risk factors associated with MVP disease progression and highlighted the need to intervene to mitigate or prevent them.

CASE
A 57-year-old, 95 kg, 183 cm man presented for evaluation of progressive dyspnea on exertion and easy fatigability that had gradually developed over 10 years before admission. He described a 34-year history of MVP. MVP is defined using echocardiography by >2 mm leaflet displacement above the annular plane accompanied by leaflet thickening of at least 5 mm.10 The original diagnosis was made in 1974 based on cardiac auscultation findings and M-mode echocardiogram (ECHO), and was later confirmed with 2-dimensional ECHO after this technology became widely available several years later.11 The patient’s primary care physician followed and medically managed his disease over the intervening years, but this past medical record was unavailable. The patient currently described shortness of breath while slowly walking up a slight incline, severe dyspnea after climbing 1 flight of stairs, 2-pillow orthopnea, and ankle swelling. He denied paroxysmal nocturnal dyspnea. The patient also reported frequent episodes of palpitations associated with dizziness that occurred several times per day and were relieved by
After induction of general endotracheal anesthesia, an intraoperative TEE examination confirmed the preoperative findings of posterior mitral leaflet prolapse, P2 flail, and severe mitral regurgitation. Anticoagulation was achieved with intravenous heparin, and the femoral artery and vein were cannulated for cardiopulmonary bypass. Antifibrinolytic therapy consisted of epsilon-aminocaproic acid (intravenous loading dose of 10 g followed by 1 g/h). The heart was approached through a 7 cm right anterior thoracotomy after the right lung was deflated and the pericardium was incised. Myocardial protection was provided using systemic hypothermia (28°C) and intermittent antegrade and retrograde cold blood cardioplegia delivered via cannulas placed in the aortic root and coronary sinus, respectively. Direct inspection of the posterior mitral leaflet during cardiopulmonary bypass confirmed the preoperative diagnoses of MVP and P2 flail resulting from 2 ruptured chordae.

Figure 1. Midesophageal 4-chamber view demonstrating prolapse and flail of the posterior mitral leaflet middle scallop (P2).

Figure 2. Midesophageal long axis view demonstrating prolapse and flail of the posterior mitral leaflet middle scallop (P2).
tendinæ. A quadrangular resection of the flail segment was performed, the posterior annulus was reapproximated with sutures, and a 30 mm Cosgrove-Edwards® posterior annuloplasty ring was placed. After rewarming, the patient separated from cardiopulmonary bypass without inotropic support. A repeat TEE examination demonstrated complete competence of the repaired mitral valve without residual regurgitation. The patient was transported in stable condition to the intensive care unit, where he was extubated later that day. The remainder of his hospital course was unremarkable, and he was discharged on the third postoperative day.

**DISCUSSION**

The prevalence of MVP defined using standard ECHO parameters has been estimated between 0.6% and 2.4% in adults. In contrast to earlier reports suggesting that MVP may be more common in women, no differences in sex distribution were reported in a recent large-scale, community-based ECHO study. Patients with MVP had a higher incidence of MR than those without the disease, but the MR observed in these patients was classified as trace or mild in the vast majority of cases. In fact, severe MR was a relatively rare finding (6.5%) in patients with MVP in the Framingham Heart Study. The frequency with which other adverse consequences of MVP occurred was also low. These findings supported previous suggestions that the diagnosis of MVP was certainly not an ominous one, but the presence of a late systolic murmur suggesting MR was a clear indication for vigilant follow-up and appropriately timed medical or surgical intervention (eg, vasodilator therapy, valve repair, or replacement).

A prospective examination of 316 middle-aged patients with MVP followed for 102 months indicated that 11 patients required mitral valve surgery, 7 developed cerebral ischemia, 6 died of cardiac-related causes, and 2 developed endocarditis. The risk of morbidity or mortality resulting from MVP identified in this investigation was relatively low (approximately 1% per year) compared with previous reports (between 1.9% and 3.7% per year). Male gender, age >45 years, and the presence of a holosystolic murmur and LA enlargement (>4 cm) suggesting severe MR were identified as particularly strong predictors of subsequent complications, including endocarditis, cerebral embolism, and sudden death. All of the aforementioned risk factors were present in the current patient. A more recent comprehensive study of 833 asymptomatic patients with MVP differentiated 10-year mortality, cardiovascular morbidity, and events related to the disease on the basis of primary (moderate to severe MR, LV ejection fraction <50%) or secondary risk factors (mild MR, LA size ≥40 mm, age ≥50 years, flail leaflet, atrial fibrillation). Patients with a primary risk factor had substantially greater 10-year mortality (45±9% versus 5±2%), more frequent cardiovascular complications (18.5 versus 0.5% per year), and a higher incidence of MVP-related events (15% versus 0.2% per year) compared with those with no or a single secondary risk factor. Patients with multiple secondary risk factors also demonstrated substantially increased cardiovascular morbidity. The current patient fulfilled the criteria for markedly elevated risk of cardiovascular morbidity and mortality. The findings of these studies confirmed previous observations suggesting that the risk of complications of disease is higher in men and older MVP patients. A strong correlation between age and severity of MR was also reported, suggesting that temporal disease progression occurs in patients with MVP complicated by MR. Interestingly, moderate to severe MR, atrial fibrillation, chordal rupture, and symptoms of congestive heart failure were more common in patients with posterior compared with anterior leaflet prolapse. Indeed, the current patient with many of these complications also had posterior leaflet prolapse. Patients with the classic form of MVP (leaflet thickening and redundancy) were shown to be a higher risk of infectious and hemodynamic complications of the disease (including severe MR) compared with those without these features. TEE and direct inspection of the mitral valve in our patient also demonstrated a redundant, thickened leaflet structure typical of the classic form of the disease.

Asymptomatic or minimally symptomatic patients with MVP, severe MR, and normal right and left ventric-
ular function were shown to progress to surgical indications for valve repair or replacement at an annual rate of 10.3%. Mitral valve repair compared with replacement enhanced long-term survival (>5 years) in patients with severe MR resulting from MVP. Reoperation was more frequently required for anterior compared with posterior mitral leaflet prolapse. Nevertheless, the overall long-term durability of valve repair suggested that early surgical intervention was appropriate in the setting of MVP-induced severe MR. These surgical results were very similar to those reported for mitral valve repair in patients with other nonrheumatic causes of MR. In contrast to medical management alone, surgical intervention was also shown to substantially reduce the morbidity and mortality associated with MR resulting from a flail leaflet. Similar to the clinical course observed in our patient, who reported progressive exertional dyspnea over a 10-year duration, surgery was required within 10 years of diagnosis of flail leaflet-induced MR in the vast majority of patients examined in this study. These findings contrast with earlier reports indicating that chordal rupture may not be associated with clinical symptoms or hemodynamic decompensation. Current recommendations indicate that asymptomatic MVP patients without MR may be followed clinically at 3- to 5-year intervals without serial echocardiography. In contrast, patients with MVP and symptomatic moderate to severe MR require vigilant follow-up at yearly intervals with ECHO quantification of LA size and valvular dysfunction.

**Funding/Support:** Department funding from the Veterans Affairs Medical Center was used to support the authors as they conducted research and wrote this manuscript.

**Financial Disclosures:** None declared.

**REFERENCES**


The mission of the *Wisconsin Medical Journal* is to provide a vehicle for professional communication and continuing education of Wisconsin physicians.

The *Wisconsin Medical Journal* (ISSN 1098-1861) is the official publication of the Wisconsin Medical Society and is devoted to the interests of the medical profession and health care in Wisconsin. The managing editor is responsible for overseeing the production, business operation and contents of *Wisconsin Medical Journal*. The editorial board, chaired by the medical editor, solicits and peer reviews all scientific articles; it does not screen public health, socioeconomic or organizational articles. Although letters to the editor are reviewed by the medical editor, all signed expressions of opinion belong to the author(s) for which neither the *Wisconsin Medical Journal* nor the Society take responsibility. The *Wisconsin Medical Journal* is indexed in Index Medicus, Hospital Literature Index and Cambridge Scientific Abstracts.

For reprints of this article, contact the *Wisconsin Medical Journal* at 866.442.3800 or e-mail wmj@wismed.org.

© 2009 Wisconsin Medical Society