Communications

Idiopathic Scoliosis and Mitral Valve Prolapse

Michael K. Smith, MD, Rae-Ellen W. Kavey, MD, and John P. Lubicky, MD
Syracuse, New York

Mitral valve prolapse (MVP) is the most common congenital cardiac anomaly, occurring in 6 to 10 percent of the young female population.[1-3] The auscultatory findings of a midsystolic nonejection click and an apical late systolic murmur are due to redundant mitral leaflets and elongated chordae tendineae.[4-7] The pathologic findings of myxomatous degeneration of the mitral leaflets suggest an underlying connective tissue abnormality.[8] MVP has been described with increased frequency in such known connective tissue diseases as Marfan's syndrome,[9] Ehlers-Danlos syndrome,[10] osteogenesis imperfecta,[11] and pseudoxanthoma elasticum.[12]

Both MVP and idiopathic scoliosis occur predominantly in young women. Thoracic skeletal deformities are common in patients with MVP, occurring in 75 percent of one adult series.[13] These facts suggest that MVP should occur frequently in patients with idiopathic scoliosis, important information for such patients, who may require major orthopedic surgery. This study was performed to determine prospectively the incidence of MVP in a population of patients with the diagnosis of idiopathic scoliosis.

Methods

From February 1982 to February 1983, all patients seen in the private practice of one of the authors (JPL) with a diagnosis of idiopathic scoliosis and curvature greater than 20 degrees were considered potential participants in the study. Their scoliosis evaluation consisted of a physical examination and posteroanterior and lateral thoracolumbar spine roentgenograms. The roentgenograms were measured by the Cobb method.[14] Hypokyphosis was defined as a thoracic kyphosis of less than 20 degrees. Letters that briefly described MVP and offered a free evaluation at the pediatric cardiology clinic were sent to 47 of the patients. Thirty-nine patients were reached by the pediatric cardiology clinic, and of these, 33 agreed to an evaluation.

Each patient was examined by a pediatric cardiologist and a pediatric resident. The examination was done in the sitting, standing, recumbent, and left lateral positions. The diagnosis of MVP was made on the basis of the presence of a midsystolic nonejection click with typical postural variation.[15] The finding of a late systolic or holosystolic murmur, if present, was noted, but it was not necessary for the diagnosis. In addition, each patient had a review and examination of cardiovascular systems, a 12-lead electrocardiogram, and an M-mode echocardiogram (Figure 1). Echocardiographic confirmation of the diagnosis was based on the criteria of Meyer.[16] The two examiners were blind to the results of each other's examinations and to the echocardiogram results.

Results

The study group consisted of 31 female and 2 male patients. Mean age was 14 years (range, 9 to 21 years). Fifteen had a family history of scoliosis. Twelve also had hypokyphosis. None of the group had any cardiovascular complaints. No patient was aware of any abnormal cardiac findings. The electrocardiograms were normal in all patients. The auscultatory diagnosis of MVP was made in 26 of the 33 (79 percent). Sixteen had an isolated

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From the Division of Pediatric Cardiology, Department of Pediatrics, and Department of Orthopedic Surgery, State University of New York–Upstate Medical Center, Syracuse, New York. Requests for reprints should be addressed to Dr. Rae-Ellen W. Kavey, SUNY-Upstate Medical Center, 750 E. Adams Street, Syracuse, NY 13210.

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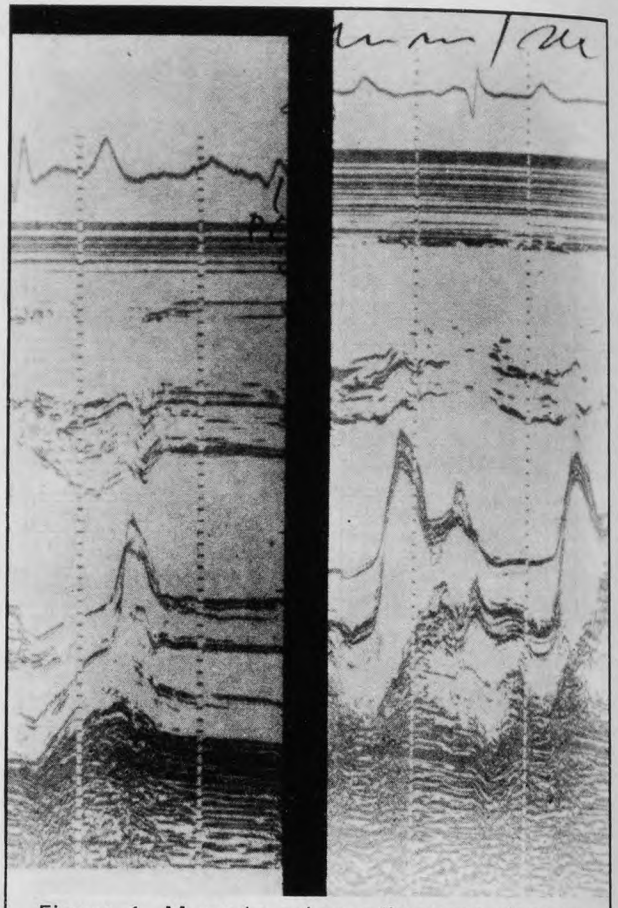

Figure 1. M-mode echocardiograms demonstrating normal mitral valve motion on the left, and mitral valve prolapse on the right. The electrocardiogram at the top of each frame identifies the onset of electrical systole. In the normal mitral valve, seen as an M-shape in diastole, the anterior and posterior leaflets move slowly anteriorly throughout systole (left-hand panel). With mitral valve prolapse, the leaflets remain horizontal rather than moving anteriorly, and there is an abrupt midsystolic posterior dip coincident with the auscultatory click

nonejection click, and 10 had a click with an associated late systolic murmur. The diagnosis of MVP was confirmed by M-mode echocardiography in 24 of these 26 patients (92 percent). All seven patients with normal physical examinations had normal echocardiograms. In four cases the diagnosis of MVP was made by only one examiner; in all four, echocardiography demonstrated typical findings of MVP.

Discussion

Although the association of mitral valve prolapse and thoracic skeletal deformities is well recognized, the incidence of MVP in patients with idiopathic scoliosis remains unclear. The frequency of 79 percent in this series is the highest yet reported. As only 70 percent of the target group was evaluated, perhaps some self-selection did occur despite the denial of symptoms on a cardiovascular review of systems. This frequency far exceeds that in the general population as well as the 29 percent incidence recently reported by Hirschfield et al[17] in another series of patients with idiopathic scoliosis. There were no data on the severity of scoliosis in that study. All patients in this study had at least 20 degrees of curvature. Perhaps the connective tissue abnormality seen in MVP is seen more frequently in adolescents with more severe scoliosis.[8]

MVP is known to occur in a wide variety of connective tissue disorders.[9-12] The results of this study suggest that in some patients a pervasive developmental abnormality of connective tissue may underlie both idiopathic scoliosis and MVP. Further supporting this concept is the simultaneous development of the mitral valve and the thoracic vertebrae, ribs, and sternum early in the seventh week of embryonic life. The common familial patterns in idiopathic scoliosis[18] and mitral valve prolapse[19] suggest that a single collagenous abnormality may influence the development of both the mitral valve and the spine. That the configuration of the thorax does not appear to be a contributing factor was substantiated by the observation that the incidence of MVP in those with lumbar curves only was 80 percent (4/5).

The natural history of MVP is largely benign in children,[15] although ventricular arrhythmias[20] and bacterial endocarditis[15,21] have been reported. Bacterial endocarditis is an important consideration in the context of patients with idiopathic scoliosis, in that they frequently require surgery. If a diagnosis of mitral valve prolapse is made in a patient with idiopathic scoliosis, appropriate antibiotic prophylaxis would be necessary prior to instrumentation.

In no patient in this series was the diagnosis of MVP made prior to the study evaluation. One patient was excluded because of Marfan's syndrome, which was first noted during the cardiac evaluation. The majority of the patients considered family physicians to be their primary care providers, and most had been referred to the orthopedist by these physicians. Study data suggest that careful screening for the presence of mitral valve prolapse is necessary in patients with idiopathic scoliosis.

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