Pulmonary Function Studies in Patients with Prolapse of the Mitral Valve*

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We studied the pulmonary function and chest x-ray films of 20 nonsmokers with idiopathic prolapse of the mitral valve. Forty-five percent of these patients complained of dyspnea. Of the 16 chest films reviewed, 44 percent had radiographic evidence of pectus excavatum and 25 percent thoracic scoliosis. The most common pulmonary function abnormalities were increased residual volume (63 percent) and a reduced steady state diffusing capacity (50 percent of patients). There was no relationship between the presence of dyspnea and either chest wall deformity or abnormality of pulmonary function. These pulmonary function abnormalities may reflect a pulmonary parenchymal disorder possibly secondary to a connective tissue abnormality in the lung.

Some patients with otherwise uncomplicated prolapse of the mitral valve prolapse have unexplained exertional dyspnea as one of their complaints. This symptom, which does not appear to correlate with hemodynamically significant mitral regurgitation, has been reported in 7 to 38 percent of patients. To further elucidate the physiologic mechanisms for this unexplained symptom, we examined chest x-ray films and pulmonary function in 20 nonsmoking patients with prolapse of the mitral valve.

Materials and Methods

Patients

The following criteria were required for patients to be included in this study: (1) mitral valve prolapse, diagnosed by at least one of the following three methods: phonocardiographic confirmation of a midystolic click heard on auscultation; echocardiographic demonstration of midystolic dipping of the mitral valve or pansystolic hamstring of at least 4 mm; angiographic demonstration of pronounced mitral valve prolapse; (2) no known respiratory or other cardiac disorder. Patients with hemodynamically significant mitral regurgitation from their mitral valve prolapse were excluded from the study. (3) no history of cigarette smoking.

Initially, the records of all patients known to us with a diagnosis of mitral valve prolapse were reviewed; of these, 18 had pulmonary function studies performed previously as a part of their diagnostic workup. Of these patients, six met the above criteria and were included in the study. These patients comprise group 1. Two of the 12 excluded patients had coexisting heart disease (atrial septal defect, idiopathic hypertrophic subaortic stenosis), one had bronchial asthma, one pulmonary sarcoidosis and eight were cigarette smokers.

Fourteen patients with newly-diagnosed mitral valve prolapse who met the above criteria were then evaluated with pulmonary function studies. These patients comprise group 2.

The records from patients in group 1 were reviewed for the presence of dyspnea and the patients in group 2 were interviewed at the time of their pulmonary function studies for this complaint. Chest radiographs from both groups were reviewed with particular attention paid to structural thoracic wall deformity. No chest x-ray films were obtained in four patients.

Pulmonary Function Studies

Spirometric flow rates in group 1 patients were determined using a 13.5 liter Collins spirometer while flow rates in group 2 patients were determined with an SRL automated pulmonary function lab. The largest vital capacity measurement was used; predicted normal values for this measurement were from Morris et al. A reduced vital capacity was considered mild if it was from 70-79 percent of predicted, and of moderate severity if from 60-69 percent predicted. FEV₁, was expressed as a percentage of the vital capacity (% FEV₁). The severity of airflow obstruction reflected by a reduced %FEV₁ was estimated according to the criteria of Gaensler and Wright. Predicted normal values for the FEF 25-75 percent were from Morris et al.

Functional residual capacity was determined in Group 1 using the helium dilution method and in group 2 using nitrogen washout techniques. Predicted values for residual volume and total lung capacity were from Goldman and Bellake and values were considered within normal limits if within 20 percent of the predicted normal value. The RV/TLC ratio was considered elevated if greater than 120 percent of predicted value.

Steady state diffusing capacity for carbon monoxide was determined using the method of Filley. In one patient (no
11), arterial Pco₂ was not determined, and alveolar Pco₂ was used to estimate dead space. Healthy volunteers were used as control subjects to estimate normal values for the steady state diffusing capacity, and the diffusing capacities of the patients in the study were expressed as percentages of these values. An impairment in diffusing capacity was considered mild if it was from 60-79 percent of predicted, moderate if from 50-59 percent, and severe if below 50 percent predicted.

Arterial blood gas levels were analyzed using an IL-113 blood gas analyzer. Alveolar-arterial oxygen tension difference P (A-a)O₂ was estimated using the alveolar gas equation and arterial blood gas levels. An upper limit of 18 mm Hg was considered normal for this determination.

Results

Patients

Of the 20 patients evaluated, there were 16 females and four males. Ages ranged from 15 to 59 years (mean 39 years). Thirteen had mid-systolic clicks and seven had silent mitral valve prolapse. Eight patients with mid-systolic clicks also had echocardiographic corroboration of mitral valve prolapse and one patient with a click had positive angiograms for prolapse. Of the seven patients with silent mitral valve prolapse, the diagnosis was made by echocardiography in four and by angiography in three.

Clinical data, chest x-ray film interpretations and pulmonary function results are given in Table 1. Cases 1-6 were from group 1 and 7-20 from group 2.

Dyspnea

Nine of 20 patients (45 percent) had exertional dyspnea as one of their complaints. Four of six from group 1 and five of 14 from group 2 had this complaint.

Chest X-ray Films

Of the 16 chest films reviewed, 10 were abnormal (63 percent). Seven (44 percent) had radiographic evidence of pectus excavatum and four (25 percent) had thoracic scoliosis. One patient (no. 6) had both pectus excavatum and scoliosis and was included in both categories. No straight back deformity was seen. All cases of thoracic skeletal deformity diagnosed radiographically were felt to be of minimal degree. There were no radiographic changes suggestive of chronic obstructive lung disease or diffuse alveolar-interstitial disease.

There was no relationship between the symptom of dyspnea and the presence of thoracic wall deformity; in the seven patients with dyspnea who had chest x-ray films, four films were normal, two showed pectus excavatum and one scoliosis. Seven of 11 patients without dyspnea had either thoracic scoliosis or pectus excavatum.

Pulmonary Function Studies

Lung Volumes: The vital capacity was reduced in four of 20 patients. In one (no 11) it was of moderate severity and in three it was mild. Of the three patients with reduced vital capacities who had chest x-ray examinations, thoracic scoliosis was present in two.

Total lung capacity was determined in 19 cases and was increased in four. An increased residual volume was present in 12 of 19 cases (63 percent), with a range in values for these patients from 123-207 percent of predicted. Of the nine patients with increased residual volumes who had chest x-ray examinations, six had pectus excavatum, while of the six patients with normal residual volumes who had films available, only one had pectus excavatum. This difference was statistically significant (P < .05). The residual volume/total lung capacity ratio was increased in nine of 19 cases (47 percent).

There was no relationship between the symptom of dyspnea and either an increased residual volume or an increased RV/TLC ratio.

Flow Rates: The %FEV₁ was normal in 18 of 20 patients and was mildly reduced in two (68 percent and 69 percent). A reduced FEF 25-75% was present in three of 20 patients (range, 47-64 percent of predicted). Two of three patients with airflow obstruction had dyspnea.

Arterial Blood Gas and A-a O₂ Difference: PaO₂ was mildly reduced in only two of 19 cases. PaCO₂ ranged from 31 to 42 mm Hg. An increased A-a O₂ difference was present in eight; of the eight, it was of mild severity in six (range, 19-25 mm Hg) and of moderate severity in two (39 and 40 mm Hg). Five of eight patients with an increased A-a O₂ difference had dyspnea.

Steady State Diffusing Capacity: The steady state diffusing capacity was reduced in 10 of 20 patients (50 percent). This impairment was considered mild in six, moderate in three and severe in one. There was no statistically significant relationship between a reduced diffusing capacity and the presence of dyspnea, or the presence of pectus excavatum or scoliosis on the chest x-ray film. Likewise, there was no correlation between a reduced diffusing capacity and either an increased residual volume or airflow obstruction.

Discussion

The results of this study lend further credence to the concept that MVP is but one facet of a multisystem disorder involving the thoracic skeleton and pulmonary parenchyma, as well as mitral valve connective tissue. The 45 percent incidence of dyspnea...
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Numbers in parentheses refer to percentages of predicted normal values.
FVC = Forced vital capacity (in liters); %FEV₁ = forced expiratory volume in one second as a percentage of the vital capacity; FEF 25-75% = flow rate (in liters/sec) between 25% and 75% vital capacity; RV = residual volume (in liters); TLC = total lung capacity (in liters); A-aO₂ Diff = alveolar-arterial oxygen tension difference (mm Hg); %Dlco = steady state diffusing capacity for carbon monoxide expressed as percentage of predicted; + (Dyspnea) = dyspnea present; O (dyspnea) = dyspnea absent; Pectus Exc = pectus excavatum; NA = Not available; ND = Not done.

in these patients may be somewhat inflated due to selection bias in group 1, since these are patients with established diagnoses of MVP who had had pulmonary function studies performed as part of a prior diagnostic workup. Nevertheless, 36 percent of those studied prospectively (group 2) had this symptom.

A high frequency of thoracic skeletal abnormalities was demonstrated, with 44 percent of the radiographs showing pectus excavatum and 25 percent thoracic scoliosis. This is in accord with two previous studies showing 61 percent⁴ and 75 percent⁵ incidence of either pectus excavatum, straight back or dorsal scoliosis. There was no relationship between

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the symptom of dyspnea and the presence of thoracic deformity, and except for the statistically significant relationship between an increased residual volume and pectus excavatum, no association was found between thoracic wall deformity and pulmonary function abnormality. It would appear from these data that the chest wall abnormality, although frequent, has little clinical significance other than as a marker for the cardiac lesion.

Pulmonary function studies were abnormal in the majority of patients, with residual volume elevated in 63 percent; RV/TLC ratio high in 47 percent and the diffusing capacity reduced in 50 percent of cases. An increased residual volume may be due to either air trapping in small, collapsible airways at low lung volumes, or to a decrease in the elastic recoil of the lung. Static volume-pressure relationships and more sensitive tests of small airways obstruction would be of further help in elucidating the cause of this increased residual volume.

The steady state diffusing capacity was diminished in 50 percent of the cases. There was no correlation between a reduced diffusing capacity measurement and dyspnea, chest x-ray film abnormality or abnormality in the remainder of the pulmonary functions. In the absence of significant anemia, a reduced diffusing capacity usually implies pulmonary parenchymal disease of the alveolar-interstitium. Pulmonary emphysema is a disease characterized by both increased residual volume and reduced diffusing capacity. Although significant air flow obstruction usually occurs by the time the diffusing capacity is reduced, this is not always the case. Measurements of lung elastic recoil, which would be reduced in generalized emphysema, may be helpful in future evaluation.

Possibly these patients have a generalized disorder of connective tissue, with involvement of the pulmonary parenchyma, as well as the mitral valve. Salazar and Edwards have noted on postmortem studies an association of myxomatous mitral valve lesions with pulmonary emphysema and/or fibrosis in eight of their 37 cases of myxomatous valves. The lung, which is well supplied with connective tissue, would be expected to show profound alterations in mechanics when that connective tissue is abnormal. Interestingly, Marfan’s syndrome, a developmental disorder of connective tissue with similar pathologic findings of the mitral valve, is associated with a variety of pulmonary abnormalities including pulmonary emphysema and air trapping. Connective tissue abnormality in the lung is postulated to underlie these pulmonary findings.

The lack of any significant correlation between dyspnea and either pulmonary function abnormality or chest wall deformity suggests that this symptom may be nonpulmonary in origin. Decreased left ventricular compliance seen in prolapse of the mitral valve is one possible mechanism. Vague “neuropsychiatric” symptoms in these patients have been described previously and testing with a personality inventory test (MMPI) revealed multiple abnormal scores including hysteria, hypochondriasis, and psychasthenia. Whether or not these personality traits are responsible for the dyspnea, of course, remains conjectural.

REFERENCES

5. Goldman HI, Becklake MR: Respiratory function tests—normal values at median altitudes and the prediction of normal results. Am Rev Tuberc 79:457-467, 1959