Mitral Insufficiency in Marfan’s Syndrome*
A Case Report of Surgical Correction

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MARFAN’S SYNDROME, AN HEREDITARY connective tissue disorder, was first described by Marfan in 1896, and is characterized by degeneration of elastic fibers of the musculoskeletal, cardiovascular and ocular systems. To date, over 400 cases have been reported in the world’s literature. Cardiovascular involvement has been recorded in 30 to 60 per cent of these cases, with aneurysmal dilatation of the ascending aorta being the most frequent finding. Mitral valve involvement has been noted in about 35 cases, although premortem documentation of it has been infrequent. Thus, the following case report of a successful surgical correction of mitral insufficiency in a patient with documented Marfan’s syndrome is noteworthy.

CASE REPORT
A 24-year-old mother of three was admitted to the University of Minnesota Hospitals for the

Figure 1a: Preoperative electrocardiogram. Figure 1b: Preoperative vectorcardiogram.
first time on November 11, 1964. A heart murmur had been heard at age four, but the patient remained asymptomatic until four months prior to admission, when easy fatiguability and dyspnea developed. Previous cardiac catheterization had disclosed mitral insufficiency.

She was a tall, slim woman with long slender extremities. Hyperextensibility of all joints and marked kyphoscoliosis with pectus carinatum were noted. On examination of the heart, she had biventricular heaves, atrial fibrillation at 100/minute, and a grade IV/VI regurgitant
apical systolic murmur. Findings compatible with a right femoral arteriovenous fistula were also found at the site of cardiac catheterization one year prior to this admission. Her blood pressure was 90/70 mm Hg.

Hemogram, renal and liver function studies were within normal limits. Her urinary hydroxyproline level was 115 mg/24 hrs. The normal for this laboratory is under 60 mg/24 hrs. The electrocardiogram showed atrial fibrillation, left ventricular hypertrophy, S-T segment depression and T-wave inversion in leads II, III and AVF, (Fig. 1a). The vectorcardiogram showed biventricular hypertrophy, (Fig. 1b). On the chest x-ray and cardiac fluoroscopy, we saw a giant left atrium and extensive calcification of the mitral valve annulus, (Fig. 2a).

On cardiac catheterization at this hospital, there was severe (4+) mitral insufficiency and mitral stenosis as indicated by a 10 mm end-diastolic gradient across the mitral valve (Fig. 3a). Modest dilatation of the ascending aorta and sinus of Valsalva were noted, but there was no angiographic evidence of aortic insufficiency, (Fig. 3b).

The patient underwent open heart surgery on November 24, 1964. At this time, she was found to have a markedly dilated left atrium which partially occluded the superior vena cava. The aortic valve felt normal to palpation. The mitral valve leaflets were redundant and thickened. The mitral annulus was hugely dilated to a diameter of 8 cm, 4 cm greater than our largest Starr-Edwards valve prosthesis, (Fig. 4). Nevertheless, we were able to snug the annulus up to the 4 cm Starr-Edwards valve with the aid of 20 mattress sutures and a Teflon patch to cover a defect between the valve and the mitral leaflet. The leaflets and their papillary muscles were left intact (Fig. 2b). The femoral arteriovenous fistula, which apparently resulted from a femoral puncture done one year previously, was also repaired. This fistula gives added emphasis to the poor quality of elastic tissue in this patient. Microscopic examination of a biopsy of the mitral valve showed thickening and hyaline degeneration with calcification.

Postoperatively, a soft murmur of mitral insufficiency persists. This probably represents leakage about the suture line. However, during the year since surgery, the patient’s exercise tolerance has improved and she can now carry out her household chores with minimal difficulty.

**DISCUSSION**

The patient’s body habitus, musculoskeletal and cardiovascular findings are consistent with a diagnosis of Marfan’s syndrome with mitral insufficiency. Additionally, she had levels of urinary hydroxyproline which were twice that found in our controls. Prochop has reported a similar finding in his cases of Marfan’s syn-

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**FIGURE 4**: Operative photograph looking from left atrium into left ventricle through a dilated mitral valve orifice. Note thickened nodular mitral valve leaflets and chordae tendineae.
drome. Of interest were the electrocardiographic findings of S-T segment depression and T-wave inversion in leads II, III, and AVF. Bowers' had made this observation in patients whose mitral insufficiency was related to Marfan's syndrome in contradistinction to other etiologies. The present case also had redundant thickened nodular valve leaflets and thickened chordae tendineae as reported in previous necropsy cases.19

Clinical documentation of mitral insufficiency on the basis of Marfan's syndrome is rare; the majority of reported cases were found at the necropsy table. To our knowledge, successful surgical correction of this lesion has not been reported previously. In this case, the unusually large mitral valve orifice led to technical problems in replacement with the presently available prosthesis. Because of the dilated orifice, it was necessary to use an unusually large number of sutures and a Teflon patch to obtain proper seating of the valve. In spite of this precaution, the patient has minimal residual mitral reflux, although she has had marked clinical improvement.

Complete reference list will appear in the reprints.

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MECHANICAL VENTRICLE

The authors feel that their data strengthen the belief in the efficacy of mechanical assist devices which can clearly aid in supporting the circulation of patients in acute or chronic left ventricular failure. Further clinical experience, according to the authors, will give increasing indication of the value of such mechanical assistance, particularly in the end stages of coronary artery and hypertensive cardiovascular disease.


TUBERCULOSIS THERAPY WITH ETHAMBUtOl

Of 46 patients under monotherapy with ethambutol, 45 reported clinical improvement, one had no change at all; 35 had radiologic improvement, 11 had no x-ray change. Of 33 with positive culture before treatment, 23 were converted after 90 days of treatment. Such favorable results were still more evident at the end of the second period of 90 days, during which ethambutol was combined with other antituberculosis preparations. A substantial number of cases in this group reported permanent closure of tuberculous cavities.

In the group of 30 patients who received ethambutol with other antituberculosis preparations, 25 reported clinical improvement and five no change at all, 23 had radiologic improvement, and seven no x-ray change at all. Fifteen of the initial 21 positive patients showed conversion of sputum. In this group also there was a satisfactory number of cavity closures. Mycobacterial resistance to ethambutol was observed only to a moderate degree in four patients in the first group only, at the end of the third month of monotherapy. At the end of the following three months of combined treatment, three of these four patients still showed good sensitivity to the chemotherapeutic agent. In the second group, only one case showed a decreased sensitivity to ethambutol upon completion of 180 days of treatment.