CLINICAL PROBLEMS IN CARDIOPULMONARY DISEASE

Management of "Asymptomatic" Mitral Stenosis

Clinical Evaluation by Arthur Selzer, M.D.

Case Summary
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A 19-year-old white woman was well until the fifth month of her first pregnancy. At that time, she noted the onset of nocturnal dyspnea, exertional fatigue, and a chronic cough with hemoptysis. The patient had no previous history of rheumatic fever or known cardiac murmur.

Pertinent physical findings were confined to the cardiovascular system. The patient's blood pressure was 110/56 mm Hg, her pulse was 110 beats per minute and regular, and her respiratory rate was 22/min. Cardiac examination revealed an apical impulse of normal character and location and a prominent parasternal lift. A loud first heart sound was followed by a soft holosystolic apical murmur radiating to the axilla. Also a separate holosystolic murmur was best heard along the left sternal border and increased with inspiration. The second heart sound was narrowly split, with accentuation of the pulmonic second sound, and was followed closely by an opening snap approximately 0.06 second after the aortic second sound (A2). A loud, long diastolic rumbling murmur with presystolic accentuation was heard at the apex. The liver was not pulsatile, but prominent jugular v waves were noted.

The patient was treated with furosemide (40 mg daily), restricted activity, and continued hospitalization for three months. At the end of the period, spontaneous, uncomplicated delivery of a boy weighing 3,827 gm (8 lb 7 oz) occurred.

Four months later, the patient returned for cardiac catheterization. At this time, she was receiving no medication and denied any cardiac symptoms, although she had a minimally active life-style. Physical examination at this time failed to demonstrate evidence of tricuspid or mitral insufficiency. The murmur of mitral stenosis persisted, and the interval between A2 and the opening snap was 0.07 second (Fig 1). An electrocardiogram demonstrated sinus rhythm, with borderline right-axis deviation and a low-voltage S wave in lead V1. A chest x-ray film was compatible with mitral valvular disease and demonstrated clearing of the congestive failure seen during the patient's pregnancy. An echocardiogram was compatible with moderate mitral stenosis. The valve was pliable, and the E-F slope measured 16 mm/sec (Fig 2). The left atrial dimension was 4.1 cm.

Right and left cardiac catheterization disclosed a mean diastolic gradient of 18 mm Hg across the mitral valve at rest; the gradient rose to 29 mm Hg with exercise. Pulmonary arterial pressure was 40/22 mm Hg (mean, 29 mm Hg) at rest and was 48/30 mm Hg (mean, 40 mm Hg) with exercise. Corresponding mean left atrial pressures were 22 mm Hg at rest and 33 mm Hg with exercise. Cardiac output at rest was 3.3 L/min, with a cardiac index of 2.0 L/min sq m; and with exercise, these values rose to 4.3 L/min and 2.6 L/min sq m, respectively. Pulmonary vascular resistance was normal. Biplane left ventricular cineangiographic studies demonstrated normal left ventricular contractility and no evidence of mitral insufficiency. Supravalvular aortic injection disclosed a competent valve.

In summary, this asymptomatic 19-year-old woman had noncalcific pure mitral stenosis with a calculated mitral valvular area of .9 sq cm.

Questions
1. How would you manage this patient?
2. If surgery is recommended, what criteria are you applying, and what operation through which approach would be recommended?
3. If surgery is recommended, what chance is there that surgery would be required in the future?
4. If medical therapy is recommended at present (since the patient is asymptomatic), what later indication would be required to select surgery?
5. Would you advise sterilization?

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The occurrence of severe mitral stenosis in patients under 20 years of age is distinctly rare here, although it is very common in most countries of the "third world." From the epidemiologic standpoint, it would be of interest to know whether this patient was born and brought up in this country.

The problem is that of a patient with all clinical and laboratory findings characteristic of pure mitral stenosis, who was asymptomatic until the middle of her pregnancy and who then developed life-threatening symptoms. Managed medically, she completed gestation, delivered a baby, and then reverted again into an asymptomatic state.

Management of this patient has the following objectives: (1) preservation of her functional capabilities; (2) protection from progression of valvular disease; (3) prevention of cardiac failure in future pregnancies; and (4) prevention of complications. Thus, her management becomes largely prophylactic, rather than remedial. Management thus includes three options, (1) medical treatment alone, (2) medical treatment in association with sterilization, and (3) surgical treatment.

Medical treatment at the age of 19 years and in the presence of an accelerated form of mitral stenosis should emphasize antistreptococcal prophylaxis in the form of intramuscular administration of benzathine penicillin or oral therapy with penicillin. The use of diuretic drugs on a regular basis or intermittently is probably the most effective measure protecting the patient from the effects of retention of fluids. This should be associated with mild to moderate restriction of sodium in the diet, and both interventions should be monitored by the patient by recording daily weights. Therapy with digitalis is not indicated, nor would I give serious consideration to the use of anticoagulant drugs.

The patient's problem arose during pregnancy; the probability is high that it would recur in the course of additional pregnancies. Nevertheless, sterilization would not appear to be a reasonable solution unless it coincides with the patient's personal wishes for family planning. Mitral stenosis is generally considered a disease which, with poor management, increases the risk of pregnancy only slightly. There is a reasonable probability that effective prophylaxis could be designed that would minimize the chances of cardiac failure during pregnancy. This would include considerable restriction of activities and the use of a strict, low-sodium diet reinforced when necessary with the use of diuretic drugs. Should the patient develop cardiac failure in spite of these precautions, mitral valvotomy could be performed during pregnancy.

Surgical treatment (at present an option) is likely to become a necessity sometime in the future, for it

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would be improbable for a patient with severe mitral stenosis at the age of 19 years to remain in stable condition without progression of mitral valvular obstruction. Indications for surgery involve two decisions, the timing of surgery and the type of operation. Mitral valvular surgery, not being of the "curative" type, is seldom performed in asymptomatic patients. Nevertheless, circumstances here (the young age of the patient and the high probability of further child-bearing and the problems arising in connection with it) would make valvotomy a reasonable option. Closed mitral valvotomy is perhaps the ideal type of surgical therapy for this patient; her youth is likely to place her in the most favorable category from the standpoint of mitral valvular anatomy. Unfortunately, closed valvotomy has fallen into disrepute and has been completely abandoned in some surgical centers. In experienced hands, closed valvotomy is associated with a very low surgical risk. While it is true that many patients (probably the majority) will eventually need repeat surgery upon the mitral valve, favorable results of the operation may last as long as 20 years. Open mitral valvotomy has the disadvantage of a slightly higher risk related to the cardiopulmonary bypass; it is uncertain whether or not it is a more effective operation.

In general, the risk-benefit ratio of the more conservative surgical approach to mitral valvular disease is favorable in symptomatic patients. In this case the circumstances listed previously would also place the ratio in the favorable category, despite the fact that the patient is asymptomatic most of the time. On the other hand, the option of replacing the mitral valve with a prosthetic or biologic artificial valve should be strongly opposed. The half-life of prosthetic valves is not definitely known, but it is probably within the range of five to eight years. Replacing the mitral valve in a patient who is 19 years old could only be justified if no other method of treatment existed and if the patient had a disability of class 4.

Subsequent Course

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Mitral commissurotomy was recommended to this patient, but she refused surgery. At a follow-up examination six months after catheterization, she remained asymptomatic and exhibited no change in her clinical findings.

References
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