
Mitral Stenosis with Severe Pulmonary Hypertension

**Sami S. Kabbani, M.D., Tali Bashour, M.D.,
Robert Dunlap, M.D., and Elias S. Hanna, M.D.**

Consecutive catheterizations in 512 patients with mitral valve disease revealed 85 cases of mitral stenosis with systolic pulmonary artery pressures equal to or in excess of 100 mm Hg. Forty-four patients (51.8%) were in New York Heart Association functional Class IV; 30 (35.3%) were in Class III; and 11 (12.9%) were in Class II. Symptoms related to pulmonary capillary hypertension were prominent in 48 patients (56.5%) while the remaining patients showed signs of low cardiac output syndrome. Forty patients underwent surgery: mitral commissurotomy was performed in 26 patients; mitral valve replacement in 10 patients, and double valve replacement in four patients. Nitroprusside was infused into the pulmonary artery in six patients with severe perioperative low cardiac output; four of these patients survived. The overall operative mortality was 15% (six patients). Good long-term results were achieved in 30 cases.

MITRAL VALVE disease is the most common indicator for cardiac surgery in the Middle East. In most of these patients, rheumatic involvement of the mitral valve starts early in life and progresses rapidly, frequently causing the development of severe "reactive" pulmonary hypertension.

Patients with mitral stenosis and extreme degrees of pulmonary hypertension pose special problems of management for both cardiologist and cardiac surgeon. Following is an evaluation of our experience with this syndrome, and some guidelines with regard to management before, during and after surgery.

Patients and Methods

At the Damascus University Medical School, 512 patients with mitral disease were catheterized during a 5½-year period; 85 were found to have mitral stenosis

with severe pulmonary hypertension (Table I). There were 28 men and 57 women in this category, a ratio of 0.49 compared to the 0.65 male/female ratio in the group with general mitral disease. The mean age was 27.1 years in the first group and 25.2 in the latter.

TABLE I. Catheterization Studies: January 1976-July 1981

	Patients with Mitral Valve Disease	Patients with Mitral Stenosis and Severe Pulmonary Hypertension
Catheterized	512	85
Operated upon	319	40

Table II reveals the New York Heart Association classification of the functional status of these patients prior to catheterization. Most patients were in Classes III and

From the Cardiovascular Surgical Center, Damascus Medical School, Damascus, Syria.

Address for Reprints: Sami S. Kabbani, M.D., Western Heart Institute at St. Mary's Hospital, 450 Stanyan Street, San Francisco, California 94117.

TABLE II. Functional Classification of Patients with Mitral Stenosis and Severe Pulmonary Hypertension

Class II	11 (12.9%)
Class III	30 (35.3%)
Class IV	44 (51.8%)
Total	85 Patients

IV; however, a significant proportion were in Class II, underscoring the fact that pathologically critical mitral stenosis *can* be associated with apparently adequate cardiac function.

The presenting signs and symptoms were divided between those related to pulmonary capillary hypertension and those indicative of low cardiac output (Table III). With the exclusion of effort dyspnea that can be attributed to both pathophysiologic mechanisms, manifestations of pulmonary capillary hypertension were predominant in about 55% of patients, and symptoms related to low cardiac output were in the remaining patients. A significant number of patients complained of hemoptysis, chest pain and syncope. Peripheral cyanosis, pul-

TABLE III. Clinical Presentations Related to Pulmonary Capillary Hypertension and Low Cardiac Output

Symptoms Related to Pulmonary Capillary Hypertension	
Effort dyspnea	85
Orthopnea	55
Cough	50
Hemoptysis	15
PA lift	36
RV lift	32
Accentuated P ₂	59
Tricuspid regurgitation murmur	55
Graham-Steel murmur	5
Symptoms Related to Low Cardiac Output	
Effort dyspnea	85
Extreme fatigue	19
Chest pain	10
Syncope	3
Peripheral cyanosis	31

monary artery and right ventricular lift, and tricuspid insufficiency murmurs were also noted in a relatively high percentage of patients.

Besides the pronounced left atrial enlargement that was characteristic of almost all cases of significant mitral stenosis, radiological features (Table IV) included severely dilated pulmonary artery segments in most patients. In about one-third of the patients, there was unusual shifting of pulmonary venous pattern to the upper lung fields, indicating pulmonary hypertension of the highest degree.

TABLE IV. Radiological Findings (Chest Roentgenogram)

Left atrial enlargement	78
Dilated pulmonary artery segment	60
Right atrial and/or ventricular enlargement	60
Reversed pulmonary venous pattern	26

Electrocardiograms revealing severe right axis deviation and R/S ratio larger than 1 in V₁, were quite common in this group of patients. Atrial fibrillation was surprisingly absent in 60% of patients (Table V).

TABLE V. Electrocardiographic Findings

Atrial fibrillation	34
Severe right axis deviation	49
R > S in V ₁	40
Clockwise loop	42
Evidence of biventricular hypertrophy	19*

* All subjects had associated mitral and/or aortic regurgitation.

Cardiac catheterization was followed by low cardiac output in 16 patients (18.8%), three of whom consequently died. Complete mitral stenosis was found in 48 patients (56.5%); combined stenosis and regurgitation in 13 (15.3%); and mitral stenosis plus aortic disease in 24 (28.2%). Tricuspid regurgitation (as evidenced by prominent V wave in the right atrial tracing) was found in 55 patients (64.7%) (Table VI).

TABLE VI. Cardiac Catheterization Findings

Complete mitral stenosis	48 (56.5%)
Mitral stenosis and mitral regurgitation	13 (15.3%)
Mitral valve disease and aortic valve disease	24 (28.2%)
Tricuspid regurgitation	55 (64.7%)

Pulmonary artery systolic pressure ranged from 100 to 160 mm Hg (mean 120 mm Hg) and exceeded the systemic pressure in 40 patients (47.1%). Pulmonary artery diastolic pressure ranged from 24 to 80 mm Hg (mean 52 mm Hg) and exceeded mean wedge pressure in 29 patients (34.1%).

Left ventricular function (estimated by the ejection fraction and left ventricular end-diastolic pressure) was normal in 38 patients (44.7%); moderately depressed in 39 (45.9%) and severely depressed in 8 (9.4%). The abnormal left ventricular function was probably due to both rheumatic myocardial involvement and mixed mitral and multivalvular disease.

Forty of the 85 patients underwent surgical correction. Twenty-six had open commissurotomy; ten had mitral valve replacement; and four had double valve replacements. One underwent tricuspid commissurotomy, and four tricuspid annuloplasty procedures were carried out in combination with mitral commissurotomy (Table VII).

TABLE VII. Operative Procedures

Mitral commissurotomy	26
Mitral valve replacement	10
Mitral and aortic valve replacement	4
Tricuspid commissurotomy	1
Tricuspid annuloplasty	4

The vasodilator drug, nitroprusside, was infused directly into the pulmonary artery in six patients who manifested signs of low cardiac output in the perioperative period. Infusion was administered at a rate appropriate to each case. Four of these patients survived.

As mentioned previously, in 16 patients there were clinical signs of low cardiac output which occurred during or immediately after catheterization. Three of those who developed this complication consequently died in spite of intensive efforts at resuscitation.

Low cardiac output also occurred after angiography in many patients with extreme pulmonary hypertension. Because of this, other clinical and noninvasive methods are now being used to assess left ventricular performance. When using angiography, we suggest application of the least amount of dye necessary to visualize the left ventricle. We also use steroids empirically prior to dye injection to prevent allergic reactions.

Other factors predisposing to poor tolerance of cardiac catheterization were inadequate sedation prior to the procedure, and insufficient supervision of patients afterward. Therefore, after catheterization all patients are now admitted to the intensive care unit for a 24-hour observation period.

Operative results were good or satisfactory in 30 patients and poor in four patients. There was a hospital mortality of 15% (six patients) (Table VIII). No late mortality has been recorded.

TABLE VIII. Operative Results

<u>Good</u>	
Improvement by 2 functional classes or more	26 (65%)
<u>Fair</u>	
Improvement by 1 functional class	4 (10%)
<u>Poor</u>	
No improvement or deterioration	4 (10%)
<u>Mortality</u>	6 (15%)

Discussion

Severe pulmonary hypertension is a serious prognostic finding in cases involving mitral valve stenosis.^{1,2} Great risks are involved in invasive diagnostic interventions and surgical management in this group of patients. Regression of pulmonary hypertension is possible after successful surgical

correction^{3,4}; however, reports of experience with severe pulmonary hypertensive patients are still limited.^{2,5} Most surgeons tend to perform open commissurotomy whenever possible.^{6,7}

A Swan-Ganz catheter should always be introduced into the pulmonary artery prior to surgery so that pulmonary artery pressures can be continuously monitored. If a critical complication occurs during or immediately following the surgical procedure, the Swan-Ganz catheter will register pulmonary artery pressures equal to or above the catheterization levels, and signs of acute right heart failure and low cardiac output resistant to conventional modes of therapy will develop. If the pericardium is open, the pulmonary artery will feel extremely hard, and should ventricular fibrillation or cardiac standstill ensue, defibrillation and cardiac massage may not be effective.

No controlled study to prove the efficacy of sodium nitroprusside has yet been conducted in our center. However, we believe that with its use, we were able to prevent serious after effects in at least two, and probably four of six patients with severely depressed cardiac output. At this institution, sodium nitroprusside is now prepared before each surgical procedure involving mitral stenosis and severe pulmonary hypertension. It is then ready for use whenever signs of low cardiac output develop, and is usually combined with common inotropic agents. The time required to wean a patient completely off nitroprusside has varied from 12 hours to three days in our six cases.

Conclusions

On the basis of our experience with this difficult clinical situation, we recommend that:

1. Patients be in the best possible condition from the medical and emotional stand-

point prior to both catheterization and operation.

2. Angiography be avoided in severely ill patients whenever possible, and that patients be observed in the ICU after catheterization (operation to follow during the same hospitalization period without delay).

3. Continuous monitoring of the PA pressure be observed immediately before, during, and after surgery, and that the shortest possible cardiopulmonary bypass time be used.

4. An infusion of sodium nitroprusside be prepared and kept on a standby basis to help patients who may develop acute right heart failure and low cardiac output from increased pulmonary resistance.

The previous guidelines will help keep the management of operative patients with mitral stenosis and severe pulmonary hypertension at an acceptable risk.

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