Rheumatic Mitral Insufficiency

By Howard B. Burchell, M.D., and Jesse E. Edwards, M.D.

The case on which the present clinicopathologic conference is based involves a 33 year old woman with rheumatic heart disease. The salient points in the history are as follows: At the age of 12 the patient was ill for a year with what was diagnosed rheumatic fever, and at the age of 15 she was ill for three months with this same condition. At the age of 18 she underwent a pregnancy which was unassociated with any recognized disability. At the age of 22 (1941) five days before the delivery of her second child, the patient had pain in the thorax and required treatment with an oxygen tent. She gave birth to a living child but was advised not to become pregnant again.

Symptoms of cardiac disability began at the age of 26 (1945) at the time of an upper respiratory infection which was associated with much dyspnea, cough and the spitting up of small amounts of blood. At this time digitalis therapy was begun. A year later the patient was told that her heart action was irregular, and this irregularity had persisted to the present time (1952). In 1948 she had an induced abortion with ligation of the fallopian tubes. Since that time she had remained ill with epigastric distress which at first was noted with exercise and relieved with rest. In the last two years it had been present more or less constantly. This distress had been temporarily relieved by the injection of mercurial diuretics. During the past year there had been some orthopnea and two episodes of severe nocturnal dyspnea. In the same period she had had slight edema of the ankles and had been hospitalized frequently.

Of possible interest is the fact that in February 1951, following dental extraction, there was rather marked weakness and recurrent sweats. No blood cultures were taken, but the doctor told her that the suspected infection of the heart valves, and she was given penicillin every two hours for five days, followed, evidently, by no persistence of fever.

When she was admitted to the hospital on June 2, 1952, under the care of the Mayo Clinic, her weight was 103 pounds (46.7 Kg.) and her blood pressure 120/70. She was in moderate distress because of dyspnea. The liver was readily palpable and tender and it extended to the umbilicus. Clinically the heart was greatly enlarged, and at the apex a systolic murmur of grade 2 intensity and a middiastolic rumble were noted. The thoracic roentgenogram showed marked enlargement of the heart which seemed to include all the chambers, and there was good evidence of left atrial enlargement (fig. 1a). The problem arose whether the patient had mitral stenosis or mitral insufficiency as the predominant lesion. After careful consideration it was believed justifiable to recommend surgical exploration with the thought that the basic problem might be one of mitral stenosis. Though considered an unfavorable situation, exploration seemed indicated. At operation a high degree of mitral insufficiency was found to be present. The patient’s postoperative course was “stormy” and she died seven days after operation on June 30, 1952.

Comment

The exposure by the surgeon of the physician’s incompetence in diagnosing mitral incompetence (insufficiency) has, in this era of mitral surgery, been more acutely embarrassing to, and less easily forgotten by, the physician than the occasional errors of diagnosis previously pointed out by pathologists. In the scramble to re-entrench themselves the internists have critically examined the traditional and the newer laboratory signs which have been the factors on which a presumptive diagnosis of stenosis or insufficiency or both has been based, and have found them less satisfactory than they had been wont to believe. Some of the problems that are undergoing critical analysis in probably every medical center concern re-evaluation of the significance of the apical systolic murmur, enlargement of the heart generally and of the left atrium and of the left ventricle in particular, studies of the pulsation of the left atrium, the presence of valvular calcification, the electrocardiographic picture, and measurements of left atrial pressures taken either directly, or
indirectly as by the wedging of the cardiac catheter into a pulmonary artery.

It may be pointed out that the person with a small heart and a very typical loud diastolic rumble at the apex has mitral stenosis, and with this diagnostic sign there has been no conflict.

In accord with the plan of this pathologic conference, the various problems which this patient brought up may be listed in terms of questions intended primarily to highlight the features in the clinical differential diagnosis between mitral stenosis and mitral insufficiency.

How may the apical systolic murmur be evaluated as an indication of the presence of mitral insufficiency?

In general, the louder the apical systolic murmur that is heard the more likely it is that the patient has mitral insufficiency. This generalization has greater validity if the systolic murmur is well heard in the axilla and there is no diastolic murmur. However, there are exceptions, and we have seen patients who have had tight mitral stenosis without insufficiency as determined by the surgeon and who had had only a loud (grade 2 to 3) apical systolic murmur. A rare patient with mitral stenosis may have evanescent murmurs and even mimic, at first examination, an idiopathic pulmonary hypertension. It may be pointed out that it has been our teaching for some years that an apical systolic murmur of moderate intensity consequent to, and related to, mitral rheumatic valvulitis is not necessarily indicative of mitral insufficiency. The organic background of such a murmur has been attributed to a distortion of the outflow tract of the left ventricle caused by scarring and distortion of the anterior leaflet.

What is the significance of the size of the heart and of the left atrium as evidence of mitral stenosis or insufficiency?

When the right ventricle is greatly enlarged it may form the apex of the heart, and it is most difficult if not impossible for the roentgenoscopist to recognize with certainty an associated left ventricular enlargement. While characteristically patients with mitral stenosis have smaller hearts than those with mitral insufficiency, there are exceptions.

Enlargement of the left atrium has been properly taught to indicate mitral stenosis, but the enlargement may be just as great in patients with mitral insufficiency. A possible exception to this may be the few tremendous aneurysmal dilatations of the left atrium that have been recorded with mitral stenosis. Also it may be noted that some patients whose mitral stenosis is demonstrable pathologically may have only slight enlargement of the left atrium. It may be emphasized, however, that the left atrium at postmortem examination may not appear large, but if the heart is left intact and the atrium distended with pressure equal to that which may be present during life there is a much better appreciation of its large size.

Is the size of the left ventricle as judged roentgenologically of help in distinguishing mitral stenosis from mitral insufficiency?

The size of the left ventricle is an important feature in considering the differential diagnosis of these two conditions. If the left ventricle shows signs of enlargement and there is not concomitant involvement of the aortic valve or acute rheumatic myocarditis, the diagnosis of mitral insufficiency is favored. In the case being presented the radiologic and electrocardiographic evidence supported the presence of both right and left ventricular enlargement (fig. 1b).

What is the significance of calcifications of the valve?

This patient had an easily recognized area of calcification which was associated with the mitral valve disease (fig. 1a). Clinically we have associated calcification with a stenotic orifice, both in disease of the mitral valve and in disease of the aortic valve, but again the relationship is not one of reliable diagnostically import. A calcified lesion usually indicates that the orifice of the valve is narrower than normal, but the stenosis may not be of functional significance. Rather, an associated
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Mitral insufficiency may be the paramount lesion from a functional point of view.

Of what significance is the increased angle between the main bronchi, viewed roentgenologically, in the diagnosis of left atrial enlargement?

This is a well-recognized roentgenologic sign associated with enlargement of the left atrium, but it is of minor importance. It be angulated and to which its intrinsic shape may be deformed may not be generally appreciated but can be demonstrated very graphically in tomograms.

What is the possibility of a previous subacute bacterial endocarditis causing valvular destruction and mitral insufficiency?

It is generally known that in bacterial endocarditis minimal lesions of the mitral valve,

![Figure](image)

**Fig. 1a.** Roentgenogram of the thorax taken in the right anterior oblique position showing the marked enlargement of the heart, particularly the left atrium and right ventricular outflow region, and calcification of the mitral valve (point of arrow).

**b.** Electrocardiogram showing auricular fibrillation and right axis deviation. The high notched R wave in V\(_2\) and the R deflection with the slight delay in the intrinsicoid deflection in V\(_2\) were interpreted as indicating hypertrophy of both right and left ventricles.

Incidentally the elevation of the left bronchus is believed to play a role, along with the pulmonary artery, in the production of paralysis of the left recurrent nerve which is experienced by some patients with disease of the mitral valve. This patient did not, however, have evidence of such paralysis. The major extents to which the left main bronchus may rather than tight stenosis, are encountered. A corollary of this is that patients with rheumatic mitral disease and auricular fibrillation rarely have subacute bacterial endocarditis. In the case under discussion clinical evaluation had to take note of the previous episode of a febrile illness for which penicillin was administered, since the occurrence of mitral insuf-
ficiency as a manifestation of healed bacterial endocarditis had to be borne in mind.

How reliable is the electrocardiogram in differentiating mitral stenosis and mitral insufficiency?

It is apparent that in the usual case the electrocardiogram should be of great value, and this has been supported in general by our experience with cases of mitral disease in which surgical treatment has been given. With few exceptions, in the cases in which operation for mitral stenosis has confirmed the condition, the experience of our group has been that right ventricular hypertrophy was indicated in the electrocardiogram by right axis deviation and a high R wave in lead V1 or aV_R. However, in cases in which there is mitral insufficiency, the pattern of left ventricular hypertrophy may be expected, but we have observed exceptions in which the evidence of marked right ventricular hypertrophy dominated the electrocardiographic picture. In the case discussed here the electrocardiogram could be best interpreted as indicating both right and left ventricular hypertrophy (fig. 1b).

Of what value is cardiac catheterization in the differential diagnosis?

(The catheterization findings in this case are given in table 1.)

Correlative studies of cardiac catheterization data seem to indicate that pressures in the pulmonary artery are apt to be somewhat lower and the cardiac output less fixed in cases of mitral insufficiency than in cases of mitral stenosis in which there is the same degree of cardiac failure. Such trends, if they exist, cannot be relied upon in evaluating an individual case. The wedge pressure contours for the individual patient, however, may give some clue as to the presence of mitral insufficiency as opposed to mitral stenosis, but again these are not wholly dependable. Connolly and co-workers have found that wedge pressures at the time of operation are practically equal in magnitude and contour to the simultaneously recorded left atrial pressure. In addition even a direct left atrial pressure record may not be diagnostic of an existing mitral insufficiency which the surgeon is able to demonstrate. These observations have caused us to doubt that esophageal pressure records will differentiate with uniform success mitral insufficiency from mitral stenosis.

The data obtained on cardiac catheterization in the patient under discussion are compatible with either mitral stenosis or insufficiency but cannot be considered to favor one condition over the other.

<table>
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<th>Site</th>
<th>Pressure, mm. Hg</th>
<th>Oxygen saturation, %</th>
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<tr>
<td></td>
<td>Maximum</td>
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<tr>
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<td>8</td>
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<tr>
<td>Right ventricle</td>
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<tr>
<td>Pulmonary artery</td>
<td>95</td>
<td>53</td>
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<tr>
<td>Pulmonary artery (onset of exercise)</td>
<td>160</td>
<td>65</td>
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<tr>
<td>Right ventricle (4 minutes after exercise)</td>
<td>115</td>
<td>15</td>
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<tr>
<td>Radial artery</td>
<td>129</td>
<td>84</td>
</tr>
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Cardiac index: rest, 1.34 liters per minute per square meter; exercise, 1.73 liters per minute per square meter.

Wedge pressure not obtained.

* Courtesy of Dr. E. H. Wood.

Of what value are special procedures designed to register the amplitude of the left atrial pulsations?

The problem here is mainly whether the moderately incompetent orifice of the valve might cause the marked systolic pulsation of the left atrium that may sometimes be seen on roentgenoscopy in the presence of very gross incompetency; from evidence gained at the time of operation it would seem doubtful that such pulsations would be discernable with moderate incompetency. With the special procedures of electrokymography and roentgenkymography we have had no experience.

If pulmonary biopsy were feasible, would it be possible from a study of the pulmonary arteries and arterioles to differentiate mitral stenosis and insufficiency?

Becker and we have already published evidence that mitral insufficiency may be
associated with severe obstructive lesions of the small arteries and arterioles identical with those seen in mitral stenosis; hence, even this hypothetic test would be unreliable. It may also be mentioned that the degree of severity of vascular lesions seen in biopsy sections of the lung has not correlated well with the hemodynamic data obtained by cardiac catheterization or with the clinical improvement following surgical treatment.

What did the surgeon find on exploration of the heart?

The surgeon (Dr. John W. Kirklin) noted at thoracotomy that the heart was immense, that it was pushed forward by a very large left atrium, that it probably was the largest left atrium he had seen, and that an expansile systolic pulsation of slight degree could be recognized in it. No thrill was palpable. The pulmonary artery was greatly enlarged and tense. Even before the heart was entered it was thought that the condition was mitral insufficiency; when the valve was explored digitally there was a very forceful turbulent flow coming back through the valve during ventricular systole. Regurgitation seemed to be coming mainly from the site of the medial commissure, and here the posterior leaflet seemed to be greatly deformed and heavily scarred. There was a question of slight stenosis of the mitral valve, since the lateral commissure was elongated by fusion of the leaflets, but further opening of the valve was thought not to be indicated.

What was found at necropsy?

The heart was enlarged, weighing 525 Gm. Most striking was the dilatation of the left atrium which contained about 400 cc. of blood. The left ventricle was moderately dilated and hypertrophied (fig. 2a). The right ventricle was moderately hypertrophied. The condition of the mitral valve was classified pathologically as a combination of rheumatic stenosis and insufficiency, although the degree of stenosis was relatively small, the mitral orifice measuring about 2.5 cm. in diameter. The existing narrowing of the orifice was brought about almost exclusively by changes at the antero-
**Fig. 2a.** Left side of the heart. The left atrium is greatly dilated. The left ventricle is moderately dilated and hypertrophied. There is fusion of the anterior (A.M.) and posterior (P.M.) mitral leaflets at the anterolateral commissure (circle) with calcification. The posteromedial commissure (triangle) is essentially normal. The posterior leaflet is shortened and retracted. The endocardial surface of the lower portion of the left atrium above the posterior leaflet of the mitral valve is at right angles to the endocardial surface of the left ventricle. In this region the epicardial surface of the left atrium bulges beyond that of the left ventricle. The area shown in the rectangle is illustrated as a photomicrograph in e. (Similar abbreviations are used in the other parts of the figure.)

*Fig. 2b.* The mitral valve from the left atrial aspect. The anterolateral commissure (A.L.C.) is deeper than normal because of fusion of the anterior and posterior mitral leaflets. Calcification is present in this region. The posteromedial commissure (P.M.C.) is essentially normal. The mitral orifice is wide.

*Fig. 2c.* The mitral valve from the left ventricular aspect. The mitral orifice is gaping, and the posterior leaflet is short. The chordal changes are relatively mild.

*Fig. 2d.* A longitudinal section through the posterior leaflet of the mitral valve and adjacent left atrium and ventricle of a normal heart. The endocardial surface of the left atrium in this region is in essentially the same plane as the endocardial surface of the left ventricle. The mitral ring lies over the endocardial portion of the left ventricle.

*Fig. 2e.* Section similar to that illustrated in d but from the case presented. The section is taken from the area outlined by the rectangle in a. The left atrial endocardial surface is at right angles to the left ventricular endocardial surface, and the epicardial portion of the left atrium is displaced outward. The mitral ring, which is calcified, is dislocated posteriorly, lying above the epicardial half of the ventricular myocardium and its overlying epicardium. The posterior mitral leaflet (M.P.) is thickened and shortened.
in apposition. Supportive evidence was had from focal thickening in the posterior wall of the left atrium which appeared to represent reactions to the trauma of regurgitant blood.

There were interesting alterations in the tracheobronchial tree where it was associated with the underlying dilated left atrium. One of these was an increase in the angle between the right and left bronchi as they arose from the trachea (fig. 3a to c). It appeared that the
The greatest factor leading to this increase in angle was displacement upward of the left main bronchus by the enlarged left atrium.

In addition to the deviation of the left main bronchus from its normal direction, there was further evidence of compression of this structure: the opposing faces of the left bronchus were virtually in apposition along one aspect of the structure. The posterior aspect of this bronchus had a sharp edge instead of being rounded (fig. 3a), and there was a change in cross sectional contour (fig. 4).

Elsewhere in the heart, there was slight thickening and shortening of the leaflets of the aortic valve. The pulmonary valve was normal. The leaflets of the tricuspid valve were slightly thickened but were not fused or shortened. The foramen ovale was closed anatomically. The venous connections to the heart were normal. The ductus arteriosus was closed. The coronary arteries were essentially normal.

Grossly, the lungs were uniform and slightly reduced in crepitation. The cut surface presented a uniform moist appearance and exuded moderate amounts of blood-stained frothy fluid.

Fig. 4a. Cross section of a normal left main bronchus (hematoxylin and eosin; X6).
b. Cross section of the left main bronchus from case being presented. At the right side of the illustration, which corresponds to the posterior aspect of the bronchus, the superior and inferior walls have been pressed into close apposition, a consequence of the existing dilatation of the left atrium during life. The cross sectional contour is quite different from that of the normal left main bronchus illustrated in a (hematoxylin and eosin; X6).

Microscopic examination of the lungs revealed features similar to those seen in cases in which the lesion is that of mitral stenosis. The lungs were characterized in part by increase in thickness of the alveolar walls by the presence of relatively thick bundles of collagen. In places there were lining cells having cuboidal characteristics. The alveolar spaces, in foci, contained moderate to considerable numbers of macrophages laden with hemosiderin. In the pulmonary vascular system there was
moderate medial hypertrophy of small and medium-sized muscular arteries. The arterioles showed varying, at times severe, degrees of intimal fibrous thickening. Often this fibrous tissue was laid down in a concentric manner. The venules showed changes similar to those in the arterioles. The walls of the small veins were thickened by muscular hypertrophy and increase in density of connective tissue elements of the media. In addition there were varying degrees of intimal fibrous thickening in this class of vessel.

From a pathologic point of view the mitral valvular lesion in this case is to be classified as that of stenosis and insufficiency. The degree of stenosis, however, was relatively minor in that the orifice measured about 2.5 cm. in diameter and probably, for this reason, constituted no significant barrier to ventricular filling. It is to be emphasized that this size of mitral orifice is far greater than that obtained after adequate mitral valvulotomy in cases of predominant mitral stenosis. The mitral insufficiency appears to have been the cause of the cardiac disability.

It is frequently taught that when mitral stenosis is present there is always some degree of mitral insufficiency. While such a statement may be true if the slightest amount of regurgitation is to be considered "insufficiency," the surgeon has now established that mitral stenosis is often a pure lesion unassociated with recognizable insufficiency. Further support of the concept that mitral stenosis frequently exists as a pure lesion is the correlation that is found between the calculated areas of the mitral valve, according to the method of Gorlin and Gorlin, and the surgeon's appraisal of the orifice of the valve.

What is the pathogenesis of chronic rheumatic mitral insufficiency?

Of particular interest is the deformity of the posterior wall of the left atrium observed in this case as in other cases of pronounced dilatation of the left atrium. There is intimate connection between the endocardium of the left atrium and the posterior leaflet of the mitral valve. It appears reasonable, therefore, that dilatation of the left atrium would cause traction on the posterior leaflet of the mitral valve and cause it to be pulled away from the mitral orifice. That this had happened in the case presented is suspected by the fact that the mitral ring was farther posterior than it is normally. To what degree a dilated left atrium might contribute to the initial stages of mitral insufficiency is difficult to determine. It is possible that a chain of events operated as follows: Primarily, shrinkage of the posterior leaflet as a result of rheumatic endocarditis could have initiated a degree of mitral insufficiency. Resulting dilatation of the left atrium would in turn have been followed by retraction posteriorly of the entire leaflet, except where it was fused to the anterior leaflet at the anterolateral commissure, and accentuation of the mitral insufficiency. In this way it is possible that a vicious circle was set up in which mitral insufficiency caused left atrial dilatation and left atrial dilatation further increased the degree of mitral insufficiency.

At this point it might be asked whether, if this reasoning is correct, the dilated left atrium causes mitral insufficiency in cases of predominant mitral stenosis. To this question a negative answer is applicable. With fusion of the two leaflets at both commissures it would appear that the effect of the dilated left atrium upon the posterior leaflet would be to distort the entire mitral valve and to cause the position, but not the size, of the orifice to be changed. It will be recalled that in the case presented there was considerable fusion at the anterolateral commissure. There was little, if any, appreciable fusion between the leaflets at the posteromedial commissure. This would allow the posterior leaflet to be displaced posteriorly by an enlarged left atrium.

Are the bronchial deformities described in this case unique?

The deformity of the tracheal bifurcation, particularly with reference to the position of the left main bronchus and the change in cross sectional contour of the left bronchus, is a feature which has been observed by us in other cases of left atrial dilatation as part of a study in progress. The change in contour of the cross
section of the left main bronchus in this circumstance would seem to cause a degree of stenosis of that structure. The narrowing of the left bronchus in the case presented could conceivably have been overcome by removal of the underlying cause, namely, dilatation of the left atrium. It is also possible, however, that were bronchial ulceration for any reason to have occurred, the healing process could have caused fusion of opposing walls of the bronchus with resulting organic and permanent bronchostenosis.

*Given a heart with stenotic changes in the mitral valve, can the pathologist say with certainty that insufficiency was or was not evident to the surgeon who had had an examining finger in the left atrium?*

In the usual instance with significant mitral insufficiency the pathologist may readily demonstrate inadequacy of the valve leaflets to close the orifice. He may also demonstrate regurgitant “jet lesions” in the endocardium of the left atrium. There is an occasional case, however, in which, to the pathologist, the problem seems to be essentially one of mitral stenosis while at operation the surgeon had clearly felt the regurgitant flow through the mitral orifice.

**REFERENCES**


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