HISTORY

45-year-old woman.

CHIEF COMPLAINT: Increasing dyspnea and fatigue of two years duration.

PRESENT ILLNESS: She had rheumatic fever as a child, with a heart murmur noted since then, and was treated with prophylactic antibiotics. Dyspnea on exertion began at age 35. Digitalis, diuretics and salt restriction were begun at that time with improvement in her symptoms. Recently, intermittent palpitations and paroxysmal nocturnal dyspnea have appeared. An afterload reducing agent was added, with only minor improvement in her symptoms.

Question: What diagnosis is suggested by this history?
**Answer:** The history of rheumatic fever, heart murmur and palpitations in a woman suggests chronic rheumatic mitral valve disease. The palpitations may be due to associated atrial arrhythmia.

The presence of a murmur without symptoms over many years suggests mitral regurgitation (MR), as this left ventricular volume load is often well tolerated for decades.

Paroxysmal nocturnal dyspnea indicates some degree of pulmonary venous hypertension, while fatigue may be a symptom of low cardiac output.

**Proceed**
PHYSICAL SIGNS

a. GENERAL APPEARANCE - Slender woman appearing chronically ill.

b. VENOUS PULSE - The CVP is estimated to be 7 cm H_2O.

Questions:

1. How do you interpret the venous pulse?
2. What is the significance of this patient’s CVP in the presence of symptoms of heart failure?
**Answers:**

1. The estimated CVP is at the upper limits of normal, and the venous wave form is normal.

2. The normal venous pressure means that right atrial pressure is within normal limits. Isolated left ventricular failure or failure compensated by medical treatment is often present with normal right atrial pressures.

A normal CVP with suspected mitral valve involvement is suggestive of chronic MR rather than mitral stenosis (MS). With chronic MR, the regurgitant volume delivered into the left atrium is gradually progressive, and the left atrium may adjust to the increased volume by dilation, with a mild increase in mean left atrial pressure. As a result, pulmonary, right ventricular and right atrial hypertension are minimized.

**Proceed**
c. **ARTERIAL PULSE** - (BP = 130/80 mm Hg)

**Question:** How do you interpret the arterial pulse?
**Answer:** The arterial pulse is normal. If this patient has significant rheumatic MR, a hyperdynamic pulse might be expected, due to decreased resistance to left ventricular ejection. However, concomitant obstruction at the mitral and/or aortic valve level, pulmonary hypertension and left ventricular failure may dampen the typical pulse of MR, so that it is normal.

d. PRECordial MOVEMENT

**Question:** How do you interpret the precordial movement?
Answer: The apical impulse is displaced inferolaterally, enlarged and sustained throughout systole. There is a prominent palpable early diastolic filling wave (arrow).

The apical impulse is consistent with a chronically volume loaded left ventricle. The rapid filling wave is related to left ventricular failure and/or excess flow into the left ventricle, as seen with significant MR.

The character of the apical impulse essentially excludes significant MS, where left ventricular filling is diminished, the left ventricular impulse is small, and filling sounds are absent.

Proceed
**Question:** How do you interpret movement at the mid left sternal edge?
**Answer:** The left parasternal impulse shows late systolic expansion characteristic of MR. Left parasternal impulses are usually due to right ventricular hypertrophy and/or dilation and are most prominent in early systole. With significant MR, the posteriorly located left atrium undergoes a gradual pressure increase and expands in mid to late systole, thrusting the heart forward.

If one simultaneously palpates the apical and left parasternal areas, the early apical impulse followed by the late parasternal impulse imparts a rocking motion to the chest.

**Proceed**
e. CARDIAC AUSCULTATION

**Question:** How do you interpret the acoustic events at the apex?
**Answer:** The first heart sound is obscured by the onset of a high frequency holosystolic murmur. A third heart sound (arrow), coincident with early diastolic ventricular filling, introduces a brief early diastolic rumble. Note the temporal relationship between these acoustic events, the carotid pulse and the late systolic impulse at the mid left sternal edge.

The murmur is characteristic of chronic MR, and typically radiates to the axilla.

It begins with mitral closure (M1) and continues up to and through aortic closure (A2), as left ventricular (LV) pressure exceeds left atrial (LA) pressure throughout this period.

**Proceed**
Answer (continued): In this clinical setting, the S3 may reflect a poorly compliant failing left ventricle or increased mitral valve flow.

The early diastolic rumble may reflect mild MS or an increased volume of blood flow resulting from severe MR. The absence of a prominent first heart sound and an opening snap, and the presence of a third heart sound indicate that the early diastolic murmur does not reflect MS.

In patients with severe MR, the left atrium is overfilled during the previous systole, and a larger than normal volume of blood enters the left ventricle in diastole. This results in an audible S3 and flow rumble. In this patient, these diastolic sounds imply that the MR is severe.

All of these findings also radiate posterolateral to the mitral area over the enlarged left ventricle.

Proceed
The intensity of the systolic murmur in MR does not accurately reflect its severity. The diastolic auscultatory findings, on the other hand, may often reflect the severity as follows:

- **Mild**: ISOLATED HOLOSYSTOLIC MURMUR
- **Moderate**: HOLOSYSTOLIC MURMUR
- **Severe**: HOLOSYSTOLIC MURMUR FLOW RUMBLE

Proceed
**Question:** How do you interpret the acoustic events at the upper left sternal edge?
Answer: The second heart sounds are of normal intensity and are normally split during inspiration. The normal intensity of P2 suggests that significant pulmonary hypertension is absent. Splitting of the second sounds in MR may be wide due to rapid left ventricular ejection against a decreased resistance, causing an earlier aortic closure. However, if left ventricular function is reduced, this finding may be absent.

f. PULMONARY AUSCULTATION

Question: How do you interpret the acoustic events in the pulmonary lung fields?

Proceed
Answer: In all lung fields, there are normal vesicular breath sounds.

**ELECTROCARDIOGRAM**

![ECG Diagram]

**Question:** How do you interpret this electrocardiogram?
**Answer:** The rhythm is sinus, and there is evidence of left atrial enlargement with notched and biphasic P waves. The increased QRS voltage with secondary ST-T wave changes is consistent with left ventricular hypertrophy. The findings are compatible with chronic MR.

To evaluate the patient’s complaint of palpitations, continuous ambulatory monitoring was performed. A rhythm strip taken during a period of palpitations is shown below.

**ELECTROCARDIOGRAM (continued)**

![Electrocardiogram](image)

**Question:** How do you interpret this rhythm strip?
**Answer:** There is atrial fibrillation with a rapid ventricular response. Atrial arrhythmias are extremely common in patients with chronic rheumatic mitral valve disease.

Normally, atrial contraction contributes approximately 15-20 percent of ventricular filling, but the relative contribution increases markedly with tachycardia. In patients with heart disease and limited cardiac reserve, atrial fibrillation may significantly impair cardiac performance.

**Proceed**
CHEST X RAYS

Question: How do you interpret these chest X rays?
Answer: On the PA film there is an enlarged cardiac silhouette with a left atrial “double contour” (arrows), and a prominent appendage (broken arrow). On the lateral film, the left atrium is enlarged and displaces the barium-filled esophagus posteriorly (arrow).

Question: Based on the history, physical examination, ECG and chest X rays, what is your clinical diagnostic impression?
**Answer:** The history, physical examination, ECG and chest X rays are all typical of chronic rheumatic MR, with a large left atrium that tends to absorb left ventricular pressure during systole, minimizing pulmonary hypertension. The patient’s symptoms, cardiomegaly, S3 and diastolic flow rumble, suggest that the MR is severe. The normal carotid pulse is best explained by a decrease in left ventricular function.

**Question:** What is the value of echocardiography in this patient with severe MR?
**Answer:** Two-dimensional echocardiography can show mitral valve anatomy and the status of the other valves, as well as determine left ventricular anatomy and systolic function.

Doppler ultrasound can accurately diagnose mitral regurgitation and estimate its severity. The patient's Doppler study follows.

**Proceed**
Question: How do you interpret this study?

LV = Left Ventricle
RV = Right Ventricle
LA = Left Atrium
Ao = Aorta
**Answer:** This Doppler color flow map shows a mitral regurgitant jet, evidenced by the mosaic (cyan-blue-yellow) coloration (arrow) in the dilated left atrium. The mosaic pattern, signifying a high velocity jet, fills most of the left atrium. This is diagnostic of moderately severe mitral regurgitation.

**Proceed**
**Answer:** The complete two-dimensional echocardiogram showed that the mitral valve was thickened, and that the posterior leaflet moved abnormally, in the same direction as the anterior leaflet. These changes are typical of rheumatic mitral valve disease.

The left ventricular cavity was dilated and the wall motion of both the interventricular septum and posterior wall was hyperdynamic, characteristic of compensated left ventricular volume overload. A markedly enlarged left atrium with systolic expansion of the posterior left atrial wall was also present. These findings are typical of mitral regurgitation. No abnormality of the tricuspid or aortic valve or aortic root was noted.

**Question:** Is cardiac catheterization indicated?
**Answer:** Cardiac catheterization and angiography, while not essential for the diagnosis, were performed to: (a) quantitate the severity of mitral regurgitation, (b) determine the pulmonary artery pressure and vascular resistance, and (c) determine the status of the coronary arteries.

**Proceed**
Simultaneous left ventricular (LV) - pulmonary capillary wedge (PCW) tracing (the PCW is obtained by wedging the catheter in the distal PA, and reflects left atrial pressure).

**Question:** How do you interpret the catheterization data?

**ADDITIONAL DATA:**

- Pulmonary Artery (PA): 45/18 mm Hg (Mean = 32)
- Cardiac Index: 1.9 L/Min/M²
- Ejection Fraction: 50%
**Answer:** There is a large V wave (arrow) in the pulmonary capillary wedge tracing reflecting significant MR. A gradient (hatched area) across the mitral valve is present only in early and mid diastole. This does not imply concomitant MS, since this type of gradient is not unusual in patients with severe isolated MR due to increased flow. Note the correlation of the phonocardiographic findings of an S3 (broken arrow), with the drop in gradient as blood rapidly fills the left ventricle. The flow rumble which follows the S3 is due to the mid diastolic pressure gradient and the normal mid diastolic approximation of the mitral leaflets.

There is mild pulmonary hypertension (normal PA pressure = 25, mean = 15 mm Hg) and a decrease in the cardiac index (normal = 2.5 - 4.0 L/Min/M²), with mild elevation of the left ventricular end diastolic pressure (normal = 4 - 12 mm Hg) and reduction in ejection fraction (normal = 55 - 70%).

**Question:** What factors determine the degree of pulmonary hypertension in patients with MR?
**Answer:** In MR, the level of the pulmonary artery pressure depends upon left atrial pressure, the reactivity of the pulmonary vascular bed, and the cardiac output. Left atrial compliance is a major determinant of the left atrial pressure.

A large, compliant left atrium may accommodate very large regurgitant volumes with little or no elevation of pressure and, hence, minimal symptoms of pulmonary congestion or edema. Conversely, a small, less compliant left atrium (as might occur with acute MR) may have its pressure dramatically increased by a small regurgitant volume, resulting in enormous V waves due to ventricularization of the left atrial pressure curve.

The variation in this “reservoir” function is, in part, responsible for some of the dramatic differences in symptoms between patients with chronic and acute MR. In chronic MR, the left atrial cavity is very large, its walls are thin and it is thus able to “absorb” the left ventricular pressure without reflecting it back into the pulmonary vessels.

**Proceed**
LABORATORY (continued) - ANGIOGRAM

LEFT VENTRICULAR INJECTION - Coronary angiography was normal.

Questions:
1. How do you interpret the angiographic data?
2. How would you treat this patient?

FRONTAL (Ventricular Systole)

LATERAL (Ventricular Diastole)

LA = Left Atrium
LV = Left Ventricle
Ao = Aorta
Answers:

1. The PA view in systole shows dense opacification of a markedly enlarged left atrium (arrows), consistent with severe chronic mitral regurgitation. The aortic outflow tract reveals no changes to suggest concomitant aortic valve disease. The diastolic lateral view shows the posterior position of the left atrium and some thickening of the mitral valve leaflets (arrows).

2. In view of this patient’s symptomatic deterioration while on medical therapy, surgical correction by either mitral valvuloplasty or replacement with a prosthetic mitral valve is indicated. Although the initial surgical mortality is approximately 5%, long term survival is improved. Evaluation for simultaneous surgical treatment of atrial fibrillation is another consideration.

Due to extensive fibrosis and calcification, valve replacement was performed and this patient improved and returned to normal activities on a medical program. Because of her valve replacement she was given anticoagulants to reduce the incidence of thromboembolic complications. She was also instructed in infective endocarditis prophylaxis.

Proceed for Summary
SUMMARY

The mitral valve is by far the most frequently affected in rheumatic carditis, with female preponderance in most series. MR commonly occurs with acute rheumatic carditis. Occasionally it is severe, but most often the systolic murmur of mild to moderate MR is consistent with a long life.

Mitral stenosis, in contrast, is a more remote result of the rheumatic inflammatory and fibrocalcific process, usually requiring years to develop. Once significant stenosis has occurred, however, symptoms tend to occur earlier and progress more rapidly than in patients with MR. In time, with scarring and contracture, the predominantly regurgitant valve usually demonstrates some degree of stenosis. The incidence of rheumatic MR has decreased as streptococcal infections are treated in childhood.

Proceed
SUMMARY (continued)

Since MR results in systolic unloading of the left ventricle, the lesion is well tolerated. Patients with severe rheumatic MR may be asymptomatic for many years. Even when symptoms do appear, there is generally gradual progression of their disability. An acute exacerbation may occur with the onset of atrial fibrillation, the occurrence of rheumatic or infective endocarditis or ruptured chordae. Persistent atrial fibrillation usually occurs with progressive left atrial enlargement.

Appropriate rheumatic fever and endocarditis prophylaxis is important in patients with rheumatic MR. Medical treatment with salt restriction, digitalis, diuretics and vasodilators is indicated, depending upon the severity of the symptoms and findings. The major clinical problem is to anticipate, and thereby prevent, severe irreversible left ventricular failure by recommending surgery at the appropriate time.
Proceed for Case Review

PATHOLOGY

Specimen from a patient with severe chronic rheumatic MR.

AORTA
NORMAL AORTIC VALVE
DILATED LEFT VENTRICLE

THICKENED ANTERIOR MITRAL VALVE LEAFLET
FUSED, THICKENED CHORDAE TENDINEAE
To Review This Case of Severe Chronic Rheumatic Mitral Regurgitation:

The **HISTORY** is typical, with rheumatic fever associated with a murmur in childhood, initial symptoms of left ventricular failure more than 20 years later and the subsequent development of atrial fibrillation.

**PHYSICAL SIGNS:**

a. The **GENERAL APPEARANCE** shows a chronically ill woman.

b. The **JUGULAR VENOUS PULSE** is normal in mean pressure and wave form, indirectly suggesting the absence of significant pulmonary hypertension.

c. The **CAROTID PULSE** is normal, as the hyperdynamic pulse of MR is somewhat dampened by associated mild left ventricular dysfunction.

Proceed
d. **PRECORDIAL MOVEMENT** at the apex reveals an enlarged and inferolaterally displaced systolic impulse and early diastolic filling wave from the volume overload of the left ventricle. The mid to late systolic impulse at the left sternal edge is due to the left atrium thrusting the heart forward as it expands from MR, imparting a “rocking” motion to the precordium.

e. **CARDIAC AUSCULTATION** at the apex reveals the high frequency holosystolic murmur of MR, an S3 due to enhanced diastolic flow and/or associated left ventricular failure, and a diastolic rumble secondary to increased flow across the mitral valve. The absence of an S4 reflects the inability of the dilated left atrium to generate a powerful atrial contraction, in contrast to the common occurrence of an S4 in acute MR with a small left atrium. These findings also radiate posterolateral to the mitral area over the enlarged left ventricle. At the upper left sternal edge, P2 is not increased in intensity, a finding against significant pulmonary hypertension.

f. **PULMONARY AUSCULTATION** reveals normal vesicular breath sounds in all lung fields.

Proceed
The **ELECTROCARDIOGRAM** shows left atrial and left ventricular hypertrophy, with atrial fibrillation on continuous monitoring, typical of chronic rheumatic MR.

The **CHEST X RAYS** show a markedly enlarged left atrium and a moderately enlarged left ventricle, without significant pulmonary hypertension.

**LABORATORY STUDIES** include echocardiography showing typical findings of rheumatic mitral valve disease with thickened leaflets, reduced valve motion and anterior movement of the posterior leaflet. Volume overload is reflected by hyperdynamic left ventricular wall motion, and MR by the large size of the left atrium with posterior systolic expansion.

Cardiac catheterization shows a prominent regurgitant V wave, reflecting the pressure rise in the left atrium due to MR. A small mitral diastolic gradient is present due to flow alone. Left ventricular function is mildly depressed, and pulmonary pressure is mildly elevated. The angiogram shows severe MR with a large left atrium.

**TREATMENT** consists of prosthetic mitral valve replacement.
To Review a patient with *MILD Mitral Regurgitation*, change to disease #8 on the keypad. You will note the following findings:

a. The **JUGULAR VENOUS PULSE** mean venous pressure is normal at 4 cm H$_2$O (normal = < 7 cm H$_2$O). The wave form is normal with a dominant “a” wave due to atrial contraction.

b. The **CAROTID PULSE** is normal in upstroke, peak, and downstroke.

c. **PRECORDIAL MOVEMENT** reveals a normal brief apical impulse in the fifth intercostal space at the midclavicular line, occurring at the time of the first heart sound.

d. **CARDIAC AUSCULTATION** at the apex reveals the high frequency holosystolic murmur of MR. Acoustic events at the aortic, pulmonary and tricuspid areas are all normal.

e. **PULMONARY AUSCULTATION** reveals normal vesicular breath sounds in all lung fields.