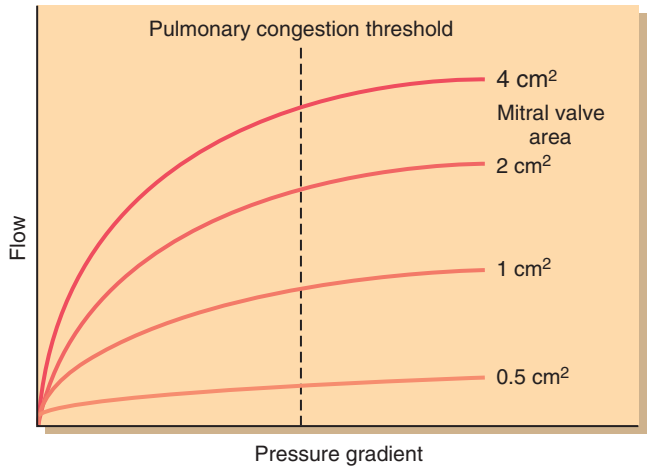

FIGURE 7-5 Graphic illustration of the relationship between the diastolic gradient across the mitral valve and the flow through the mitral valve. As the mitral valve becomes more stenotic, the pressure gradient across the valve must increase to maintain flow into the left ventricle. If the mitral opening is 1 cm^2 or smaller, the flow rate into the left ventricle cannot be significantly increased, despite a significantly elevated pressure gradient across the mitral valve. (Modified from Wallace AG: Pathophysiology of cardiovascular disease. In Smith LH Jr, Thier SO, editors: The international textbook of medicine (vol 1), Philadelphia, 1981, Saunders, p. 1192.)

TABLE 7-4 REVISED JONES CRITERIA FOR DIAGNOSIS OF RHEUMATIC FEVER*

MAJOR CRITERIA	MINOR CRITERIA
Carditis (pleuritic chest pain, friction rub, heart failure)	Fever
Polyarthritis	Arthralgia
Chorea	Previous rheumatic fever or known rheumatic heart disease
Erythema marginatum	
Subcutaneous nodules	

*Rheumatic fever is diagnosed based on the presence of two major criteria or one major and two minor criteria after a recent documented group A streptococcal infection.

leaflet commissural fusion, a funnel-shaped, flow-limiting orifice forms. Although the aortic valve can be affected as well, it is almost never involved without concomitant mitral valve involvement. In less developed countries, the incidence is much higher than in westernized societies, and the time from acute rheumatic fever to symptomatic stenosis is often years instead of decades.

Clinical Presentation

Patients typically remain asymptomatic until the valve area is reduced to approximately 1.5 cm^2 or less. Initial symptoms are usually exertional in nature, with noticeable dyspnea or fatigue that resolves with rest. As the stenosis progresses, symptoms develop earlier with exercise, until ultimately they are markedly limiting. If the cause remains undetected, the markedly elevated left atrial and pulmonary pressures will lead to significant left atrial enlargement. Even in the absence of atrial fibrillation, there is increased risk of thromboembolism. When atrial fibrillation occurs, it carries an approximate 18-fold increased risk of cerebrovascular accident. Secondary changes in the pulmonary vasculature may occur over time, rarely resulting in fixed pulmonary hypertension that is not relieved by valve surgery performed at this late stage. Finally, the right ventricle responds adversely to the chronic severe pulmonary hypertension, and clinical right-sided heart failure ensues.

Diagnosis

Only about 60% of patients with rheumatic heart disease recall having rheumatic fever as a child, and the history cannot be relied on to suspect the diagnosis. Elicitation of the symptoms, followed by confirming physical examination findings, is the cornerstone of diagnosis.

Early in the disease course, there is typically a prominent S_1 sound. As motion of the leaflets becomes increasingly restricted, S_1 may become soft. The opening in diastole begins with a snap and is followed by a low-pitched diastolic rumble, heard best with the bell of a stethoscope at the apex in the left lateral decubitus position. The length of time from S_2 to the mitral opening snap can help in estimating stenosis severity, with shorter intervals indicating proportionally increased severity.

The ECG may show left atrial enlargement and, ultimately, right atrial enlargement; right axis deviation and right bundle branch block are possible in later stages. Left atrial enlargement and dilation of the main pulmonary artery are often identified by chest radiography. In advanced cases, RV enlargement may be evident (see Table 7-2).

Confirmation is typically by echocardiography, and the valve can be evaluated by several methods. The restricted motion of the mitral leaflets in diastole is highly characteristic of rheumatic disease. The valve leaflets appear to dome or form the shape of a hockey stick. The motion of the valve leaflets forms an overall funnel-shaped mitral orifice (Fig. 7-6). Finally, a Doppler examination can be used to estimate the mitral valve area and to measure the mean diastolic gradient across the valve. If doubt remains regarding mitral stenosis severity despite these techniques, invasive pressure measurements taken directly from the left ventricle and atrium will permit calculation of a valve area and mean mitral gradient.

Treatment

In general, symptoms guide the treatment of mitral stenosis. It is believed that asymptomatic cases do not require treatment unless significant pulmonary hypertension has developed (class I indication, level C evidence) or new-onset atrial fibrillation has occurred (class IIb, level C).

In most patients, exertional symptoms drive the need for therapy. As with all other valve lesions, a mechanical (percutaneous or surgical) intervention is required to alter the natural history. In the case of mitral stenosis, an invasive procedure that can be performed in a catheterization laboratory, known as percutaneous balloon mitral valvuloplasty (PBMV), has gained favor over the past 2 decades. A balloon is delivered through the femoral vein across the mitral orifice and inflated (Fig. 7-7); this procedure is repeated with successively larger balloons until the valve area is significantly improved. PBMV has a class I indication for symptomatic patients with moderate or severe mitral stenosis, providing their leaflet morphology is acceptable for this technique.

If the patient is deemed a poor candidate for PBMV, surgical commissurotomy or valve replacement is considered to be a class IIa indication in the symptomatic patient. Because mitral stenosis