Profiles in Dilated (Congestive) and Hypertrophic Cardiomyopathies

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Cardiomyopathies are primary disorders of heart muscle. Although the term cardiomyopathy is sometimes restricted to refer to cardiac muscle disorders of unknown etiology, most cardiologists include disorders of both unknown and known etiology. For example, the cardiac muscle disorder associated with long-standing ingestion of excessive quantities of ethanol is generally termed alcoholic cardiomyopathy, and the disorder resulting from high-dose doxorubicin therapy for malignancy is called doxorubicin or Adriamycin cardiomyopathy.

In general, cardiomyopathies are classified descriptively, as listed in Table 32.1. This chapter discusses only the first two types of cardiomyopathy listed in Table 32.1. Restrictive cardiomyopathy is discussed in Chapter 33, along with constrictive pericarditis, with which it is often confused. Obliterative cardiomyopathy is extremely rare in the United States and is beyond the scope of this book.

DILATED (CONGESTIVE) CARDIOMYOPATHY

The clinical syndrome of dilated cardiomyopathy represents a collection of disorders and is also called congestive cardiomyopathy. The term congestive cardiomyopathy was used previously to refer to this syndrome because the clinical presentation is marked primarily by peripheral and pulmonary edema. Currently, the term dilated cardiomyopathy is preferred because with modern noninvasive techniques (echocardiography, radionuclide ventriculography) it has become possible to diagnose this syndrome before the onset of clinical signs and symptoms of congestion. Also, with effective diuretic and vasodilator management, the congestive component can be eliminated and ventricular filling pressures returned to normal. The ventricular chambers remain dilated, however, with increased end-systolic and end-diastolic volumes and reduced ventricular ejection fraction. Dilated cardiomyopathy is a syndrome that can develop in the setting of a variety of specific cardiac disorders (1), and the hemodynamic profile may vary, depending on the etiology.

CARDIAC CATHETERIZATION PROTOCOL

Study of the patient who is suspected of having dilated cardiomyopathy should include right and left heart catheterization with measurement of pressures, cardiac output, and resistances. As discussed in Chapters 4 and 5, during right heart catheterization the routine measurement of oxygen saturation in blood taken from the superior vena cava and pulmonary artery is essential to detect unsuspected right-to-left shunting. In one elderly patient referred to the author for evaluation of advanced left heart failure, stretching of a patent foramen ovale by high left atrial pressures had led to left-to-right shunting and a $Q_p/Q_s$ of 2.0. Surgical closure of this patent foramen ovale resulted in marked clinical improvement. Angiographic studies will need to be tailored to the individual case, but left ventriculography and coronary angiography are commonly done as a part of the diagnostic study in patients with dilated cardiomyopathy.

Hemodynamic Findings

In the symptomatic patient with dilated cardiomyopathy referred for cardiac catheterization, left and right ventricular filling pressures usually are elevated. As mentioned, however, it is possible that the ventricular filling pressures at rest may be normal; this finding is particularly likely in the asymptomatic patient detected early in the course of his or her disease by noninvasive screening. Also, the patient who has been treated intensively with diuretics (e.g., furosemide and spironolactone) and who is receiving a potent vasodilator (e.g., enalapril) may show little or no
Cardiac output is generally reduced in the patient with dilated cardiomyopathy. In milder cases, the cardiac index may be normal or only slightly reduced and may range from 2.4 to 3.0 L/min/m². In patients with New York Heart Association (NYHA) class III or IV symptoms from dilated cardiomyopathy, it is common to find the cardiac index depressed more severely. Thus a cardiac index of 1.6 to 2.3 L/min/m² can be expected in the usual symptomatic patient presenting with dilated cardiomyopathy, and a cardiac index of 1.5 L/min/m² or less indicates an advanced depression of myocardial function and a poor prognosis.

Elevation in ventricular filling pressures at rest. In general, increases in left and right ventricular filling pressures can be induced easily in such patients by supine bicycle exercise, performed as outlined in Chapter 15. With 6 minutes of supine bicycle exercise, pulmonary capillary wedge pressure commonly rises from 10 to between 25 and 40 mm Hg, and right atrial pressure increases from 6 to between 15 and 20 mm Hg. Supine bicycle exercise places an acute volume and pressure load on the ventricular myocardium and easily brings out underlying loss of contractile reserve.

The *left ventricular pressure waveform* is typically abnormal in patients with dilated cardiomyopathy. Both the rate of rise and the rate of fall of left ventricular pressure are slow, and this is usually visible to the naked eye (Fig. 32.1). Slowing of both the rate of rise and the rate of fall of left ventricular pressure gives a *triangular appearance* to the pressure waveform, with the peak systolic pressure representing the apex of the triangle; end-diastolic and minimal (early) diastolic left ventricular pressures define the triangle's base. This deformity accounts for the brief duration of systolic ejection in dilated cardiomyopathy and contrasts with the trapezoidal, almost square-wave appearance of left ventricular pressure in a normal, vigorous heart. A corollary of the triangular waveform of left ventricular pressure in dilated cardiomyopathy is a normal or low peak systolic pressure. The triangular waveform is associated with reduced values for both the rate of rise (+dP/dt) and the rate of decline (-dP/dt) of left ventricular isovolumic pressure. The marked reductions in +dP/dt seen in dilated cardiomyopathy reflect impaired inotropy and are not increased in response to pacing-induced increases in heart rate, as seen in normal hearts (2), (3). Thus a flat “force/frequency” relation may be demonstrated in the cardiac catheterization laboratory in such patients, paralleling observations in isolated heart muscle (2).

**FIG. 32.1.**

Left ventricular (LV) micromanometer and aortic (Ao) pressure tracings in a 68-year-old woman with advanced dilated cardiomyopathy. Marked slowing of the rates of left ventricular pressure rise and fall give the LV pressure tracing a triangular appearance. Also, the minimal value for left ventricular diastolic pressure is markedly elevated.

A second abnormality of the left ventricular pressure tracing in dilated cardiomyopathy is elevation in the value for left ventricular minimal diastolic pressure (Fig. 32.1). Normally, the left ventricular pressure declines briskly after aortic valve closure, reaching a nadir close to 0 mm Hg shortly after mitral valve opening. This reflects the normal pattern of rapid myocardial relaxation, acting together with *restoring forces* generated by a vigorous systolic contraction, with end-systolic elastic compression and torsion forces being released during early diastolic filling. In the experimental laboratory under conditions of extremely vigorous contraction (e.g., isoproterenol infusion) or hypovolemia (e.g., hemorrhage), the left ventricular diastolic pressure actually may become negative early in diastole, a phenomenon known as *diastolic suction*. In dilated cardiomyopathy, diastolic relaxation is generally slow and incomplete (3), and restoring forces produced by the weakened systolic contraction are minimal. These factors militate against a normal low value for left ventricular minimal diastolic pressure. In addition, end-systolic volume is increased in patients with dilated cardiomyopathy, and this abnormality tends to elevate diastolic volume and pressure above normal.

To appreciate these abnormalities in the left ventricular pressure waveform in dilated cardiomyopathy, one must have pressure tracings of good quality with careful attention to the details discussed in Chapter 7. Micromanometer catheters are not necessary to achieve such high-quality tracings, as can be seen in Fig. 32.2, where fluid-filled and micromanometer tracings are superimposed.

**FIG. 32.2.**
Left ventricular (LV) micromanometer pressure and its first derivative (dP/dt) in a patient with dilated cardiomyopathy before (left) and after (right) intravenous infusion of milrinone. Positive and negative dP/dt have increased without increase in arterial pressure and with a decline in preload, suggesting increased myocardial contractility and relaxation. Left ventricular minimal diastolic pressure is now closer to zero, as is normal. Fluid-filled and micromanometer pressures are displayed simultaneously, indicating the excellent fidelity that can be achieved with fluid-filled systems using the principles described in Chapter 7. (Reproduced with permission from Baim DS et al. Evaluation of a new bipyridine inotropic agent=mmilrinone=min patients with severe congestive heart failure. N Engl J Med 1983;309:748.)

The left ventricular pressure tracing abnormalities just described can be corrected substantially by acute administration of an inotropic drug (4–6). Figures 32.2 and 32.3 illustrate the effects of a phosphodiesterase inhibitor (milrinone) and a beta-adrenergic agonist (prenalterol) on the left ventricular pressure contour in patients with dilated cardiomyopathy. As can be seen, early diastolic relaxation is more rapid and more complete after administration of these drugs, as reflected by the steep decline in left ventricular pressure to a value near zero in early diastole.

**FIG. 32.3.**

Effects of the beta agonist prenalterol on left ventricular and aortic pressure (A) and left ventricular pressure-volume plots (B) in patients with idiopathic dilated cardiomyopathy. The tracings illustrate the restoration of a normal low value for the left ventricular diastolic pressure nadir, as well as a downward shift in the diastolic pressure volume relationship. (Reproduced with permission from Erbel R, et al. Hemodynamic effects of prenalterol in patients with ischemic heart disease and congestive cardiomyopathy. Circulation 1982;66:361.)

Patients with dilated cardiomyopathy often have elevations in pulmonary and systemic vascular resistance. It is common to find pulmonary vascular resistance increased to 150 to 300 dyn-sec-cm\(^{-5}\), and patients with values 400 dyn-sec-cm\(^{-5}\) or more are not rare. These increases in pulmonary vascular resistance result in pulmonary hypertension with mean pulmonary artery pressure commonly of 30 to 50 mm Hg. Systemic vascular resistance is usually 1,500 dyn-sec-cm\(^{-5}\) or more in untreated patients with advanced dilated cardiomyopathy, probably representing a response to combined elevations in serum levels of angiotensin, vasopressin, and norepinephrine. Because cardiac output is reduced, modest increases in systemic vascular resistance do not result in actual elevation of arterial blood pressure but rather tend to preserve arterial pressure at a normal or only slightly reduced level.

Reduction of systemic and pulmonary vascular resistances to normal by administration of vasodilator agents often results in a striking increase in cardiac output and a simultaneous reduction in left and right ventricular filling pressures. As shown in Fig. 32.4, acute administration of sodium nitroprusside (7),(8) or captopril (9),(10) results in an upward and leftward displacement of the left ventricular filling pressure/stroke volume relationship, since heart rate is affected minimally by these agents in the setting of chronic heart failure.

**FIG. 32.4.**

Effects of acute administration of sodium nitroprusside (A) and captopril (B) on left ventricular filling pressure-stroke volume relationships in patients with advanced heart failure. Some of these patients had heart failure on the basis of idiopathic dilated cardiomyopathy, and some had ischemic heart disease. Responses were similar and appeared to be independent of etiology. (See text for discussion.) (Reproduced with permission from (A) Guiha NH, et al. Treatment of refractory heart failure with infusion of nitroprusside. N Engl J Med 1974;291:587; and (B) Davis R, et al. Treatment of chronic congestive heart failure with captopril, an oral inhibitor of angiotensin-converting enzyme. N Engl J Med 1979;301:117.)

During cardiac catheterization in patients with dilated cardiomyopathy, it is often wise to test responsiveness to a vasodilator in the laboratory. This can be done routinely using the following protocol. After measurement of cardiac output and resting hemodynamics and before angiography, if filling pressures are elevated significantly (e.g., pulmonary capillary wedge pressure >= 16 mm Hg) and cardiac output is depressed (e.g., pulmonary artery blood
oxygen saturation \( \leq 65\% \), begin an infusion of sodium nitroprusside as long as arterial systolic pressure is 90 mm Hg or more and has been stable. The starting dose is 15 (g/min through a secure, free-flowing intravenous line, and the infusion rate is increased every 3 to 5 minutes to doses of 25, 50, 75, 100, 150, 200, and 300 (g/min, if needed, until arterial mean pressure has fallen to 20 mm Hg or wedge pressure has fallen by 50% or more or pulmonary artery oxygen saturation has increased to 75% or more. Usually, one of these three endpoints is achieved at a dose of sodium nitroprusside of 200 (g/min or less; however, there are occasional patients in whom 300 (g/min or more is required. If the patient is feeling well during the vasodilator infusion (as is generally the case), continue the infusion during left ventriculography and coronary angiography as a prophylactic measure to protect against pulmonary edema. The dose should be reduced if the arterial systolic pressure is 85 mm Hg or less. A favorable response to sodium nitroprusside in the cardiac catheterization laboratory is not only an aid to the safety of the procedure but also a predictor of a favorable response to an oral vasodilator in the patient's long-term management.

### Angiographic Studies

The hallmark of dilated cardiomyopathy as seen on left ventriculography is left ventricular enlargement (increased end-diastolic and end-systolic volumes) with a reduced ejection fraction. Left ventriculography in patients with dilated cardiomyopathy classically reveals extensive hypokinesis, which, although usually diffuse in nature, is commonly associated with regional wall motion abnormalities that suggest a heterogeneity of the myocardial injury and mimic coronary artery disease. This may represent the consequence of asymmetric injury initially, and in this regard it is of interest that myocarditis may be focal in its inflammatory effects. We have seen several patients in whom biopsy-proven acute myocarditis mimicked regional ischemia and infarction, with left ventriculography showing discrete areas of akinesis or even focal aneurysm formation. These areas of regional dysfunction also could represent the result of coronary emboli from mural thrombus because the occurrence of left ventricular mural thrombus is increased in patients with dilated cardiomyopathy.

Angiographic abnormalities associated with dilated cardiomyopathy include dilatation and loss of the normal eccentric shape of the left ventricle. Normally, the ratio of long axis \( L \) to the minor axis \( M \) is 2:1 for the left ventricular chamber at end-diastole. In dilated cardiomyopathy, \( L/M \) approaches 1:1. This change tends to increase meridional wall stress (see Chapter 16) but has an unpredictable effect on longitudinal wall stress, depending on the extent of associated ventricular hypertrophy. In this regard, left ventricular hypertrophy is common in patients with dilated cardiomyopathy (11). Some authors have reported a substantial beneficial effect of hypertrophy on survival in patients with dilated cardiomyopathy and have suggested that protection against increasing wall stress might have a protective role for these patients (11).

### Endomyocardial Biopsy

Enthusiasm for obtaining endomyocardial biopsy as a part of the diagnostic workup in patients with suspected dilated cardiomyopathy often reflects the experience of a particular laboratory in performing the procedure. Endomyocardial biopsy is done almost routinely in many laboratories as part of the diagnostic study in patients with advanced heart failure. In more than 100 endomyocardial biopsies in nontransplant patients with advanced heart failure at Beth Israel Hospital in Boston, specific heart muscle disorders (inflammatory myocarditis, amyloidosis, hemochromatosis) were found in approximately 15% of cases.

Viral myocarditis is widely regarded as a precursor and etiologic agent for many patients with dilated cardiomyopathy, although evidence confirming this hypothesis definitively remains elusive (12). Endomyocardial biopsy early in the course of viral myocarditis shows a characteristic inflammatory cell infiltrate. (See Chapter 20 for details.) However, in the chronic phase (days 15 to 90 in experimental studies) the cellular infiltrate largely disappears, followed by the appearance of myocardial fibrosis. Special studies can detect persistent viral DNA in this stage. There is also evidence that apoptosis is found in the chronic stage, accounting for the ongoing loss of myocytes in the absence of cell necrosis (12).

The technique of endomyocardial biopsy and additional specific diseases it can detect are described in detail in Chapter 20. In one study of 100 consecutive endomyocardial biopsies carried out to evaluate heart failure of uncertain etiology (13), the pathologic information obtained was judged to be clinically useful in 54 patients and not useful in 46 patients. Specific diagnoses that could be made from histologic examination of the biopsy material...
In summary, a variety of hemodynamic, angiographic, and histologic features can be defined precisely in the course of a single diagnostic cardiac catheterization procedure in patients with suspected dilated cardiomyopathy. Findings from such a diagnostic study yield valuable information about prognosis (11,13–15) and help direct appropriate therapy.

HYPERTROPHIC CARDIOMYOPATHY

Cardiac hypertrophy develops to some extent in a wide variety of cardiac diseases. In hypertrophic cardiomyopathy, however, the development of cardiac hypertrophy proceeds without an obvious inciting stimulus, or develops out of proportion to the magnitude of the stimulus or stimuli that can be identified. Hypertrophic cardiomyopathy was noted early on to be familial in most cases (16) and is transmitted as an autosomal-dominant trait; nevertheless, in clinical practice many cases appear to be sporadic. Studies of molecular genetics have shown that mutations in any of at least seven different genes can cause hypertrophic cardiomyopathy; each of these “culprit” genes encodes for proteins essential for the formation of the normal cardiac sarcomere (17). Expression of some of these genes in transgenic mice has duplicated some of the features of hypertrophic cardiomyopathy. However, we are still at an early point in this research, and it is not yet possible to link a particular genetic mutation with a specific hemodynamic/anatomic phenotype. Most authors distinguish between obstructive and nonobstructive forms of the disorder based on the presence or absence of a resting (unprovoked) systolic pressure gradient within the left ventricle (16), and the presence of a gradient has caused this disorder to be called idiopathic hypertrophic subaortic stenosis (IHSS) or hypertrophic obstructive cardiomyopathy (HOCM). There remains controversy as to whether true “obstruction” occurs in this condition (18) because there is some evidence that most of the left ventricular stroke volume has been ejected before development of a significant gradient. There is general agreement, however, that the pressure gradient, when present, has several adverse consequences, including increased systolic wall stress (in cardiac muscle proximal to the site of septal/mitral leaflet contact) and increased myocardial oxygen consumption.

Hypertrophic cardiomyopathy may be diffuse and symmetric, involving all regions of the left ventricle equally, or it may be asymmetric. Asymmetric hypertrophic cardiomyopathy commonly involves the high interventricular septum, which is disproportionately hypertrophied so that the ratio of thickness of the diastolic septal wall to that of the free (lateral or posterior) left ventricular wall is 1.3 or more. Another form of asymmetric hypertrophic cardiomyopathy, which has been reported from Japan (19), involves massive apical hypertrophy of the left ventricle. A characteristic electrocardiographic feature is the presence of giant negative T waves in the precordial leads. The apical form of hypertrophic cardiomyopathy has now been recognized to occur in Europe and North America (20). (21).

FIG. 32.5.

Left ventricular (LV) catheter pullback to the aorta in a patient with hypertrophic cardiomyopathy. There is a significant systolic gradient within the left ventricular cavity, and the LV outflow tract and aortic pressure waveforms exhibit a spike-and-dome contour. (Reproduced with permission from Braunwald E, et al. Idiopathic hypertrophic subaortic stenosis: a description based on an analysis of 65 patients. Circulation 1964;30[Suppl 4]:3.)

Hemodynamic Findings

As in the patient with suspected dilated cardiomyopathy, cardiac catheterization in the patient being evaluated for hypertrophic cardiomyopathy should include right and left heart study. Right atrial and right ventricular pressures usually are normal in patients with hypertrophic cardiomyopathy. Rarely, involvement of the right ventricle is said to result in a systolic gradient within the right ventricular chamber, although I have never seen such a case personally. If the hypertrophic process involves the right ventricle or if the pulmonary capillary wedge pressure is substantially elevated, right ventricular diastolic pressures may be elevated.

Left ventricular end-diastolic pressure may be normal in patients with hypertrophic cardiomyopathy but is usually
Cardiac output is usually normal or increased in patients with hypertrophic cardiomyopathy, except in the late stages of the disease, when contractility decreases.

The most dramatic hemodynamic features of hypertrophic cardiomyopathy are those related to the systolic intraventricular pressure gradient. As seen in Fig. 32.5, the pressure gradient is present between the body and the outflow tract of the left ventricle. A key feature of this systolic gradient and of most of the associated findings is their variability. Most patients with hypertrophic cardiomyopathy do not have a systolic pressure gradient at rest but may develop one with appropriate provocative maneuvers as listed in Table 32.2.

**FIG. 32.6.**

Left ventricular (LV) and femoral artery (FA) pressure tracings in a woman with hypertrophic cardiomyopathy and asymmetric septal hypertrophy illustrating the increase in gradient and development of a spike-and-dome configuration in the arterial pressure waveform following an extrasystolic beat. Also, arterial pulse pressure clearly narrows in the postextrasystolic beat compared with the control value in the beat before the extrasystole. This narrowing of pulse pressure is known as the Brockenbrough-Braunwald sign.

It should be emphasized that the presence of a systolic gradient at rest or following provocation is a hallmark of only one variety of hypertrophic cardiomyopathy: that form with asymmetric septal hypertrophy. The diffuse hypertrophic variety and the variety associated with massive apical hypertrophy do not exhibit true left ventricular outflow tract gradients at rest or with provocation (19,21). However, catheter entrapment can develop easily in patients with apical as well as symmetric forms of hypertrophic cardiomyopathy, giving the false impression of an outflow gradient (21).

An interesting aspect of the systolic gradient is an associated deformity that develops in the aortic pressure waveform. This deformity consists of an initial rapid rise in aortic pressure to give a spike early in ejection, followed by a dip in pressure and a secondary rounded or dome-shaped tidal wave before the dicrotic notch. This spike-and-dome configuration is seen in the central aortic pressure and is transmitted to the carotid pulse and peripheral arterial tracings. It is most evident following an extrasystolic contraction (Fig. 32.6) but is also seen during Valsalva maneuver (Fig. 32.7) and at other times (Fig. 32.8). The mechanism for this spike-and-dome configuration may be related to blending of an initial hyperdynamic ejection velocity leading to the development of a Venturi effect that sucks the anterior mitral leaflet into the outflow tract, thereby impeding middle and late systolic ejection velocity.

**FIG. 32.7.**

Left ventricular (LV) and femoral artery (FA) pressure tracings in the patient illustrated in Fig. 32.6. Valsalva maneuver produces a marked increase in the gradient, as well as a change in the femoral arterial pressure waveform to a spike-and-dome configuration.

**FIG. 32.8.**

Left ventricular (LV) and left brachial artery (LBA) pressure tracings in a 64-year-old woman with hypertrophic cardiomyopathy. A: The effect of a spontaneous change from nodal rhythm to sinus rhythm. The short arrows show LV end-diastolic pressure. With restoration of sinus rhythm and a presumed decrease in the obstruction, LV stroke volume increases as reflected in the improved LBA pulse pressure. Also, the loss of atrial kick in patients with a stiff ventricle leads to an acute reduction in cardiac output. (continued)
In addition to developing a spike-and-dome pattern, the aortic pulse pressure fails to widen in a postextrasystolic potentiated beat (16). Normally, a potentiated left ventricular contraction has a larger stroke volume than the preceding sinus beats, and this increased stroke volume results in an increased aortic pulse pressure.

Patients with hypertrophic cardiomyopathy, however, develop a spike-and-dome configuration in which pulse pressure is unchanged or actually reduced following an extrasystolic beat (Fig. 32.6). This sign, which was described by Brockenbrough et al. (28) in 1961, is known as the Brockenbrough-Braunwald sign and is believed to reflect worsening of obstruction of the left ventricular outflow tract during the potentiated beat, with diminished stroke volume and aortic pulse pressure.

**FIG. 32.8.**

The impaired left ventricular diastolic relaxation seen in hypertrophic cardiomyopathy (22–26) can be dramatic and can affect the contour of the left ventricular diastolic pressure tracing (Fig. 32.9). The patient illustrated in Fig. 32.9 was a 55-year-old woman with a family history of hypertrophic cardiomyopathy who presented with advanced congestive heart failure manifested by paroxysmal nocturnal dyspnea, marked fatigue, and peripheral edema. An echocardiogram showed asymmetric septal hypertrophy. At cardiac catheterization, there was no outflow tract gradient at rest or with provocation. Right atrial mean pressure was increased (11 mm Hg), reflecting pulmonary hypertension (60/30, 40 mm Hg), which in turn reflected a markedly increased mean pulmonary capillary wedge pressure (32 mm Hg). Arteriovenous oxygen difference was wide (71 mL O2/L), and cardiac index was depressed (2.0 L/min/m²). Left ventricular ejection fraction was reduced at 41%, a finding sometimes seen in late-stage hypertrophic cardiomyopathy. As seen in Fig. 32.9, the left ventricular diastolic pressure did not exhibit its normal rapid decline to a nadir near zero. Instead, early left ventricular diastolic pressure was increased at approximately 35 mm Hg and continued to decline after mitral valve opening until atrial systole produced a diastolic pressure rise coincident with the a wave. The diastolic abnormalities of hypertrophic cardiomyopathy are improved by calcium channel blockade (22,23,29–31), although occasional serious adverse effects have been seen with verapamil (32). Combined treatment with beta blockade and verapamil is currently regarded as the pharmacologic treatment of choice. However, disopyramide has been reported to have beneficial hemodynamic effects in some patients.

**FIG. 32.9.**

Left ventricular (LV) and aortic (Ao) pressure tracings and rate of LV pressure rise (dP/dt) in a 55-year-old woman with hypertrophic cardiomyopathy. There is no resting pressure gradient. LV diastolic pressure waveform is very abnormal, suggesting marked impairment in myocardial relaxation. Fluid-filled and micromanometer LV tracings are both shown.

Diastolic dysfunction is also prominent in hypertensive hypertrophic cardiomyopathy of the elderly, a syndrome described by Topol et al. (33). This condition represents a form of hypertrophic cardiomyopathy seen in elderly patients with mild to moderate hypertension who exhibit severe concentric hypertrophy, a small left ventricular cavity, supernormal systolic function characterized by excessive left ventricular emptying, and marked abnormality of diastolic relaxation. In describing this syndrome, Topol and coworkers (33) observed that several of their patients with this condition improved when treatment with digoxin and diuretics was stopped. In contrast, beta-adrenergic blocking agents often were effective in relieving dyspnea and chest pain. Left ventriculography in such patients shows a severely hypertrophied ventricular chamber, which demonstrates cavity obliteration at end-systole.

Although I will not discuss restrictive cardiomyopathy in this chapter, it is perhaps of value to point out that some unusual forms of infiltrative cardiomyopathy may present with features of both restrictive and hypertrophic cardiomyopathy. Miller et al. (34) reported a patient with eosinophilic heart disease who had a left ventricular subaortic gradient of 90 mm Hg, a spike-and-dome pattern in the central aortic pressure tracing, and a systolic
murmur that increased with either amyl nitrate inhalation or Valsalva maneuver. Left and right ventricular end-diastolic pressures were elevated at 28 and 16 mm Hg, respectively, and left ventricular ejection fraction was markedly increased at 94%. Treatment with prednisone and warfarin resulted in substantial improvement over a 4-month period (34).

FIG. 32.10.

Left ventricular (LV) angiogram in the lateral position in a patient with hypertrophic cardiomyopathy with obstruction. The anterior leaflet of the mitral valve moves toward the interventricular septum in systole (arrow), producing marked narrowing of the LV outflow tract. Mitral regurgitation into the left atrium (LA) is present. (Reproduced with permission from Braunwald E, et al. Idiopathic hypertrophic subaortic stenosis: a description based on an analysis of 65 patients. Circulation 1964;30[Suppl 4]:3.)

Angiographic Findings

The angiographic findings in hypertrophic cardiomyopathy are rather unique and help to explain some (but not all) of the unusual hemodynamic features just described. In hypertrophic cardiomyopathy with asymmetric septal hypertrophy, left ventriculography shows a thickened intraventricular septum bulging into the left ventricular outflow tract in diastole and systole. In addition to this abnormality, patients with hypertrophic cardiomyopathy in whom a systolic gradient is present within the left ventricular chamber generally show systolic anterior movement (SAM) of the mitral valve's anterior leaflet (Fig. 32.10).

In contrast to hypertrophic cardiomyopathy with asymmetric septal hypertrophy, the patient with asymmetric apical hypertrophy does not show systolic anterior motion of the mitral leaflet. In patients with apical hypertrophic cardiomyopathy, the left ventricle shows marked thickening of its anteroapical wall, giving the ventricle a spade-shaped appearance (Fig. 32.11).

FIG. 32.11.

Left ventriculogram at end-diastole (A) and end-systole (B) in a patient with apical hypertrophic cardiomyopathy. There is a spadelike configuration at end-diastole with a marked increase in free wall thickness and an extremely vigorous contraction with almost total cavity obliteration at end-systole. (Reproduced with permission from Yamaguchi H, et al. Hypertrophic non-obstructive cardiomyopathy with giant negative T waves (apical hypertrophy): ventriculographic and echocardiographic features in 30 patients. Am J Cardiol 1979;44:401.)

Patients with asymmetric septal hypertrophy have a distortion of the left ventricle that in the right anterior oblique view often resembles a banana, partly because of the large papillary muscles, which appear as filling defects.

In addition to exhibiting abnormal shapes (spade, banana) and systolic anterior movement of the mitral valve, patients with hypertrophic cardiomyopathy often exhibit mitral regurgitation on left ventriculography. This is usually mild but may progress to become hemodynamically significant. Coronary angiography may show characteristic abnormalities in hypertrophic cardiomyopathy, with marked systolic compression of septal branches of the left anterior descending artery (35). In addition, a “sawfish” systolic narrowing of the left anterior descending artery has been reported by Brugada et al. (36) and is illustrated in Fig. 32.12. The indentations of the left anterior descending artery associated with systolic narrowing of the vessel may represent the effect of contracting hypertrophied and disorganized muscle fiber bundles in the vicinity of the coronary artery.

FIG. 32.12.

Left coronary angiogram in right anterior oblique projection with caudocranial angulation. Diastolic (A) and systolic (B) frames are shown. A “sawfish” appearance of the left anterior descending artery is seen in association with

**Catheter-Based Therapy for Hypertrophic Cardiomyopathy**

Surgical resection of part of the interventricular septum (myotomy/myectomy) was one of the first treatments developed for patients with idiopathic hypertrophic cardiomyopathy, and this operation is widely regarded as a last-resort therapy for patients with refractory symptoms. Based on the concept that the hypertrophied, hypercontractile interventricular septum may be playing an important role in the pathophysiology of this condition, a catheter-based therapy was developed (37) in which ethanol is infused into the septal artery(ies) supplying the hypertrophic myocardium. In one report (38) of 33 symptomatic patients with hypertrophic obstructive cardiomyopathy and a resting gradient of 40 mm Hg, 2 to 5 mL of absolute ethanol infused into the septal artery distal to a local septal artery balloon occlusion induced a focal myocardial infarction with creatine kinase elevations to nearly 2,000 U. Complete heart block developed in 11 patients, who required permanent pacemaker placement. All patients were improved symptomatically, with NYHA class decreasing from 3.0 ± 0.5 to 0.9 ± 0.6. This promising therapy requires further study but may be an effective substitute for surgical myotomy/myectomy in the patient with refractory symptoms.