Cardiomyopathies in the Elderly

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Definition

The term cardiomyopathy refers to a diffuse or generalized myocardial disorder of unknown cause. Patients who meet this criterion have idiopathic or primary involvement of the myocardium. Secondary myocardial diseases resulting from metabolic, nutritional, endocrine or ischemic causes are excluded. However, it is quite possible for valvular, hypertensive or coronary artery disease to coexist with primary cardiomyopathy.

Classification

Using a pathophysiologic classification based on the dominant type of ventricular dysfunction, there are three broad types described, although overlap among the types may be observed.

1) Dilated or congestive cardiomyopathy. Primary characteristics are impaired systolic function with dilation of chambers and increased muscle mass without complete compensatory increase in wall thickness. In some cases dilation may be minimal or absent.

2) Hypertrophic cardiomyopathy. This is characterized by normal or small chamber size, increased wall thickness, hyperdynamic systolic ejection and impaired diastolic filling characteristics.

3) Restrictive or infiltrative cardiomyopathy. The myocardium shows increased stiffness secondary to infiltrative morphologic abnormalities that lead to diastolic dysfunction. The atria are dilated and systolic function is impaired in late stages.

Myocardial Function and Aging

The effects of aging on myocardial function are not well defined in humans. There is some evidence of diastolic dysfunction, although systolic function and cavity dimensions are unaffected (1). Studies in animals support some reductions in diastolic function with aging. The intrinsic heart rate (that is, after removal of autonomic influence on the sinoatrial pacemaker) is reduced. A diminished inotropic response to catecholamines has been reported (2), as has a decrease in ventricular response to hemodynamic stress during beta-adrenergic blockade (3).

The effects of aging on the myocardium appear to include diastolic dysfunction, reduced responsiveness to catecholamines and slowing of the intrinsic heart rate. These, alone, are not sufficient to result in clinical evidence of cardiac dysfunction. However, they may play an additive role in elderly patients with systolic hypertension, mild degenerative valvular disease, anemia or other concomitant disorders.

Dilated Congestive Cardiomyopathy

This condition is thought to be uncommon in the elderly, although approximately 10% of patients in large series are >65 years (4–8). It is possible that the published reports are biased in favor of younger patients. Congestive heart failure is often erroneously attributed to ischemic heart disease or hypertension. Alternatively, the reported age distribution represents a selection process such that few people with dilated cardiomyopathy survive beyond age 65.

Diagnosis. The diagnosis is generally confirmed by echocardiographic evidence of four chamber dilation with depressed ventricular systolic function. The diagnosis of dilated cardiomyopathy further rests on the exclusion of etiologic factors that can result in diffuse myocardial systolic dysfunction. Important among the factors to be excluded are adriamycin toxicity, postirradiation damage and chronic alcoholism. Although alcoholic cardiomyopathy is sometimes considered a secondary myocardial disease characterized by dilated cardiomyopathy, the myocardial depressant effects of alcohol excess may precipitate or exacerbate heart failure in patients with *idiopathic* dilated cardiomyopathy. Dilated cardiomyopathy may also be misdiagnosed as heart failure secondary to coronary artery disease. Angiography should be considered in patients who are thought to have dilated cardiomyopathy who have chest pain. There is a propensity for thrombus formation and subsequent systemic embolic events with dilated cardiomyopathies; this should be considered as a cause of stroke or transient ischemic attacks in the elderly.

Treatment. Treatment is essentially that of heart failure due to systolic dysfunction and anticoagulation. Digitalis is especially useful in the presence of atrial fibrillation or other supraventricular tachyarrhythmias. Caution is advised in prescribing vasodilator drugs, because excessive hypotension may prove hazardous.

Acute myocarditis. Acute myocarditis appears to be rare in the elderly and is difficult to diagnose clinically, although an acute onset with a short history is suggestive. The common etiologic agents are viruses, particularly Coxsackievirus, and rickettsia. Endomyocardial biopsy can demonstrate conspicuous cellular infiltrates with cellular degeneration and necrosis. The role of endomyocardial biopsy both in diagnosis and as a guide to therapy, particularly in the elderly, is not known.

Immunosuppressive therapy has been proposed to treat subacute and chronic myocarditis diagnosed by biopsy. However, the diagnostic criteria are uncertain and therapeutic efficacy remains to be determined. Furthermore, immunosuppressive agents may be poorly tolerated in the elderly (9).

Hypertrophic Cardiomyopathy

Although early reports suggested that this condition was rare in older patients, patients beyond the seventh decade are included in many large series (10–12). Widespread use of echocardiography may uncover more patients with hypertrophic cardiomyopathy with few or no symptoms.

Symptoms. Symptoms in older patients are similar to those in younger subjects; these include dizziness, syncope, dyspnea, chest pain and palpitation. A correct diagnosis is often delayed in the elderly because dizziness and syncope may be attributed to cerebrovascular causes, chest pain to coronary artery disease and dyspnea to associated lung disease. This may explain why the diagnosis of hypertrophic cardiomyopathy was considered in only a third of patients before referral for evaluation (11).

Physical findings. Patients with left ventricular outflow "obstruction" with intraventricular pressure gradients may have characteristic physical findings: bifid carotid pulse, double apical impulse, a fourth heart sound and a systolic murmur that increases with the Valsalva maneuver and decreases with squatting. Aortic regurgitation due to coexisting aortic valve calcification occurs in some elderly patients with hypertrophic "obstructive" cardiomyopathy and should not exclude this diagnosis. Coexisting mitral anulus calcification with mitral regurgitation may mask the diagnosis of hypertrophic cardiomyopathy.

Sudden death. Most natural history studies of hypertrophic cardiomyopathy have emphasized the rarity of sudden death in elderly patients with this condition (13). This may represent survival of a subgroup with a low propensity

to sudden arrhythmic death. Alternatively, less vigorous physical activity in the aged may be protective.

Pathology. The morphologic features of hypertrophic cardiomyopathy in the elderly are similar to those in younger patients (14), but hypertrophy is often more pronounced and may be concentric with severe free wall hypertrophy equal to that of the septum. Fibrous endocardial thickening of the upper interventricular septum and of the anterior mitral leaflet is common. Associated abnormalities in the elderly include mitral anulus calcification and degenerative calcific aortic valve disease. More extensive valvular calcification may result in the coexistence of valvular and myocardial disease. An awareness of such coexistence is necessary for the correct diagnosis and management of hypertrophic cardiomyopathy in the elderly.

Echocardiography. Unsuspected hypertrophic cardiomyopathy may be detected by echocardiography obtained for other indications because the classical physical signs are often absent or masked, or the diagnosis is not considered. The typical combination of echocardiographic findings includes asymmetric left ventricular hypertrophy, hyperdynamic left ventricular function, systolic anterior motion of the anterior mitral valve leaflet and left atrial enlargement. Doppler ultrasound evidence of an impaired diastolic filling rate and an intraventricular pressure gradient increase the diagnostic accuracy. Usually echocardiography is the only diagnostic test necessary.

Treatment. The therapy of the elderly patient with hypertrophic cardiomyopathy is similar to that of the young patient (15,16). Asymptomatic patients without high grade arrhythmias require no treatment. Symptomatic patients are usually managed with beta-receptor blocking agents; calcium channel blockers are also useful, although these drugs have increased adverse effects in the elderly, such as, constipation, ankle edema and hypotension. These adverse effects may be accentuated by surgery or vasodilator or diuretic drug therapy. Surgery, namely myotomy and myectomy, may be indicated in symptomatic patients with a demonstrable outflow gradient in whom medical therapy has failed. The surgical approach often results in gratifying symptomatic improvement with an acceptably low operative mortality rate (17,18). Endocarditis prophylaxis is warranted for appropriate surgical procedures.

Hypertensive hypertrophic cardiomyopathy of the elderly. Topol et al. (19) recently described elderly patients with the syndrome of "hypertensive hypertrophic cardiomyopathy of the elderly." All patients had a history of hypertension; chest pain or dyspnea was the presenting complaint in the majority. Echocardiographically, the syndrome was characterized by severe concentric cardiac hypertrophy, a small left ventricular cavity and supernormal indexes of ventricular systolic function. Systolic anterior motion of the mitral valve and cavity obliteration were noted in many patients. Impaired left ventricular diastolic function as manifested by a prolonged early diastolic filling period and a

reduced rate of diastolic filling appears to be the most likely cause of the presenting symptoms of pulmonary congestion. Congestive heart failure with normal systolic function may similarly be due to hypertrophic cardiomyopathy or early stages of restrictive cardiomyopathy (20,21). This syndrome is not distinct from hypertrophic cardiomyopathy except for concentric hypertrophy and associated hypertension. It may be seen at any age, but appears to be more frequently recognized in the elderly.

Restrictive Cardiomyopathy

Amyloidosis. Primary amyloidosis is a frequent cause of restrictive cardiomyopathy, but is not common in the elderly. The characteristic features of this condition include small ventricles and large atria with elevated filling pressures and thickened walls; a characteristic "sparkling" of the ventricular myocardium on the echocardiogram; the ECG may show low QRS voltage, atrial fibrillation and conduction defects. Diagnostic confirmation can be obtained by biopsy of rectal, gingival or endomyocardial tissue. There is no specific treatment.

Senile cardiac amyloidosis is a separate pathologic entity seen after 70 years of age. Its prevalence increases with age, reaching nearly 80% in those >95 years of age (22). The macroscopic appearance is often normal. Microscopically, the earliest amyloid deposits are seen in atrial capillaries. In advanced cases, small deposits may be present in the valves. Conducting tissue is affected late, if at all. It is unclear if senile amyloidosis has any functional clinical significance, although atrial fibrillation may coexist with larger atrial deposits.

Irradiation. A restrictive type of physiology may occasionally result from irradiation damage. This may be clinically evident months or years after the radiation therapy.

Postoperative pericarditis. An increasing number of patients in this age group are likely to develop postoperative restrictive cardiomyopathy, presumably secondary to post-coronary artery bypass graft or other surgical pericarditis.

Summary

Cardiomyopathies in the elderly have certain characteristic features. The dilated form appears to be less common than in younger patients. Hypertrophic cardiomyopathy is more often associated with severe and concentric hypertrophy (14). The prognosis of hypertrophic cardiomyopathy seems to be better in the elderly, because they appear to have a lower incidence of sudden death. Restrictive cardiomyopathies are not common in the elderly, and senile amyloid heart disease rarely, if ever, results in congestive heart failure. A syndrome of clinical heart failure with reduced diastolic compliance and preserved systolic function is more common in elderly patients.

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