THE CARDIOMYOPATHIES: Part I. INTRODUCTION

By

S. SULTAN AHMED,* M.D., FRCP(C)., FACP., FACC., FCCP

Ever since William Harvey's discovery, the physiologists have viewed the heart as a muscular pump inserted into the circulation to provide continuous flow of blood to the tissues of the body. From this point of view, valves are considered essential to give direction to the stream and the cardiac performance is characterized predominantly in terms of its pumping efficiency. In light of the new dimensions in the past decade or two-namely the mechnical and biochemical properties of contracting and passive heart muscle, the state of left ventricular myocardium is considered critical in maintaining the integrity of heart as a pump. A decrease in its contractile power may eventuate heart failure even when no other parts of cardiovascular system are affected.

The great vulnerability of myocardium, in the basis of its function and structure in relation to injurious agents has received substantial emphasis. A large group of diseases may, in addition to involving various parts of other systems, affect the myocardium with cardiac symptoms dominating the clinical picture. Another group of diseases with no obvious etiologies may primarily involve the myocardium, without evidence of the disease elsewhere in the body. Largely because endocardium and pericardium are not significantly involved in any of these disorders, this latter group has been collectively designated as primary diseases of the myocardium. A number of other terms have been used synonymously with the above. The multiplicity of variation in terminology along with inconsistencies in classification, has contributed to the past and present confusion surrounding the subject of myocardial disease.

To simplify, cardiomyopathy means heart (cardiac) muscle disease (myopathy). In a broad sense the diseases affecting the myocardium can be divided into two major categories: (1) primary myocardial disease; (2) circulatory disease, which have their primary allegiance elsewhere in the cardiovascular system, with secondary effect on the myocardium.

The term "primary myocardial disease," refers to those diseases of diverse etiology which specifically and primarily involve the myocardium rather than other areas of cardiovascular system. If the etiology of the cardionyopathy is unknown, the designation, "idiopathic cardiomyopathy" or "idiopathic primary myocardial disease" is used, if the etiology is known, the cardiomyopathy should be so designated.

Primary myocardial disease is best understood within the framework of some type of orderly grouping. Accordingly, classification has

*Professor of Medicine. Co-Director, Cardiac Catheterization Laboratory. Director, Exercise Testing Laboratory.

Department of Medicine CMDNJ-New Jersey Medical School Newark, N. J., U.S.A.

been approached from two points of view: (1) pathophysiologic, that is, how the myocardium is affected in terms of functional and structural derangement, and (2) etiologic, that is, presumed causes of the myocardial abnormalities.

The pathophysiological varieties of primary cardiomyopathy include the following: hypertrophic, congestive and restrictive. For practical purposes, left ventricular or biventricular involvement is with rare exception, always present. The main feature of hypertrophic cardiomyopathy is the massive ventricular hypertrophy which affects principally the septum but may extend to involve all parts of the left ventricle and sometimes also the right ventricle. This may be genetically determined. Whereas the systolic function of the heart remains good until late stages of the disease, the ventricular filling is significantly compromised. Congestive cardiomyopathy is characterized by clinical expression of failure of the heart as a pump. Unlike the hypertrophic variety, the ventricular hypertrophy is moderate rather than extreme and the left ventricle is markedly dilated. No evidence of genetic basis is noted. Restrictive cardiomyopathy indicates that both ventricular filling and contraction are impaired, similar to constricitive pericarditis. Despite the limitation to distension in diastole, filling pressures are elevated and the rate and degree of fibre shortening are reduced. Whether the diseases affecting this disorder are on the basis of inheritance or merely a reflection of impaired diastolic distension is unknown and functionally irrelevant. This condition is relatively rare but commonly produced by infiltration or fibrosis of the myocardium with or without obliteration of the cavity. An etiologic classification of primary myocardial disease is shown in the table 1.

Table 1: CARDIOMYOPATHIES

I. Primary or Idiopathic (affecting the myocardium primarily).
   1. Hypertrophic with or without obstruction.
   2. Familial.
   3. Endomyocardial fibrosis, fibroelastosis.
   4. Peri and postpartum.

II. Secondary (associated with systemic disorders).

A. Inflammatory (myocarditis) due to:
   1. Infectious agents (viruses, bacteria, fungi, etc).
   2. Toxins (diptheria).
   3. Immune response (collagen vascular diseases).
   4. Granulomatous disease of unknown etiology.
   5. Hypersensitivity reactions (sulfa, other drugs).
   6. Unknown agent-idiopathic.

B. Non-inflamnatory
   1. Metabolic
      (*a*) Endocrine: diabetes, hyper-and hypothyroidism, pheochromocytoma, acromegaly and adrenal cortical insufficiency.
      (*b*) Nutritional: Kwashiorkor, avitaminosis, hypervitaminosis-D, obesity.
   2. Neuromuscular: Friedrich's ataxia, progressive muscular dystrophy, myotonia atrophica, myasthenia gravis.
   3. Infiltrative processes: amyloidosis, glycogen and lipid storage, hemochromatosis, primary and secondary tumors.
   4. Physiochemical agents
      (*a*) Physical: radiation, burns.
      (*b*) Chemical: emetine, metals (mercury, cobalt, antimony), phosphorous, antibiotics, hypokalemia, alcohol, carbon monoxide.
   5. Ischemia.

The clinical diagnosis of primary myocardial disease is best approached by considering the etiology and patholophysiology. The recognition is simplest when comparatively young, normotensive individuals present with cardiomegaly, left ventricular hypertrophy, tachyarrhythmias, conduction disturbances, or heart failure without a prior cardiac history and without clinical evidence of valvular heart disease, congenital heart disease or disease of the coronary arteries, lungs or pericardium. Such individuals may be asymptomatic or may have a gamut of symptoms with the ultimate of unremitting heart failure.

As for the management, ideally early diagnosis with identification of specific etiologic agents, sets the stage for best therapeutic results. Treatment of cardiomyopathy is best considered under three categories: (1) supportive management of cardiovascular symptoms, (2) control of factors aggravating the cardiac insufficiency, and (3) treatment of specific etiologic causes.

The supportive management involves four considerations, namely, control of congestive heart failure, rate, rhythm or conduction disturbances. The use of digitalis, diuretics and sodium restriction are, as a rule, beneficial in treating the congestive heart failure of both the acute and chronic cardiomyopathies. Concern of increased sensitivity to digitalis especially in acute myocarditis, speaks for the use of rapidly acting, rapidly excreting preparations such as digoxin with small loading doses and with comparatively large maintenance doses. Serious tachyarrhythmias and conduction disturbances should not be managed by standard pharmacological regimens but instead should be handled in intensive care units until control has reduced the threat of sudden death.

The management of contributory and aggravating causes in patients with myocardial disease is of considerable importance. A number of factors are relevant in this regard, namely, exercise, obesity, mental distress, heat and humidity, hypoxia, respiratory infections of bacterial etiology, and coexisting systemic and embolic complications deserve close attention. Limitation of physical activity during the acute phase of myocarditis and in the presence of overt congestive heart failure seems to be appropriately based on simple hemodynamic considerations alone. Value of prolonged bedrest as advocated by Burch has considerable merit but remains an unsettled issue. The precise information regarding the cardiocirculatory effects of protracted physical immobilization in normal man in lacking. Judicious use of anticoagulants both for, and in anticipation of, pulmonary embolism as in congestive heart failure and for systemic embolism from mural thrombi especially with cardioversion cannot be emphasized enough. The use of low dose intramuscular heparin (50 mg/12 hours) as a prophylaxis should be considered in cardiomyopathic patients who are confined to bed. The logic of treating bacterial pneumonias and other coexistent systemic diseases is obvious. Specific treatment of cardiomyopathies is dependent upon the early recognition of the etiological cause.

References

1. Wigger C.J. Circulatory dynamics. New York Grune, 1951, p.1-24.

2. Sonnenblick E.H. Implications of muscle mechanics in the heart. Federation Proc. 21:975-990, 1962.

3. Hudson R.E.B. The cardiomyopathies: Order from chaos. Am. J. Cardiol 25-70-77, 1970.

4. Fowler N.O. Differential diagnosis of the cardiomyopathies. Prog Cardiovasc Dis. 14:113-128, 1971.

5. Goodwin J.F. Prospects and predictions for the cardiomyopathies. Circulation 50:210-219 1974.

6. Ahmed S.S., Regan T.J. Cardiomyopathy: The spectrum of etiology and clinical management. New Horizons in Cardiovascular Diseases, Chapt 6, pp. 213-248, 1980.

7. Burch G.E. DePasquale N.P. Recognition and prevention of cardiomyopathy. Circulation 42:A47-A53, 1970.

———:o:———