SESSION I

Chairman: Dr. E. G. J. Olsen

Introduction, problems and aims of the Multicentre Research Project

J. F. Goodwin*
M.D., F.R.C.P., F.A.C.C.

Royal Postgraduate Medical School, London

There is growing awareness of the importance of the cardiomyopathies as a common form of heart disease with global distribution.

By definition the cardiomyopathies are diseases of the myocardium of unknown cause (Goodwin, 1970, 1974; Goodwin and Oakley, 1972) which means that in congestive cardiomyopathy the diagnosis has often to be one largely of exclusion. In other types, particularly the hypertrophic form, a positive diagnosis can be made by clinical and investigative means even though the cause or causes remain unknown.

The cardiomyopathies can be conveniently divided into four types depending upon their structural and functional characteristics. The types are: hypertrophic (obstructive); congestive (dilated); obliterative; restrictive (Goodwin, 1970). The hypertrophic and congestive types are entirely different from each other and the other forms but considered together are the commonest types of cardiomyopathy to be found and rival coronary heart disease in importance as serious cardiovascular problems.

The Workshop today is concerned solely with congestive cardiomyopathy.

Because of the lack of knowledge of causation, the treatment of congestive cardiomyopathy is unsatisfactory and knowledge regarding aetiology is urgently needed. Before this can be obtained, information about incidence, geographical distribution, epidemiology, natural history and associated circumstances or illnesses is needed. To obtain this information it is necessary to arrive at a firm diagnosis and to eliminate confusion between different types of cardiomyopathy.

Congestive cardiomyopathy (COCM) is characterized by dilated ventricles with poor systolic function, and often by atrio-ventricular valvar regurgitation. It is a diffuse disease involving both ventricles. It usually presents as heart failure with signs of low cardiac output, cold blue extremities, raised central venous pressure and gallop rhythm. The ventricular volumes are increased and the chambers contract poorly when studied by echo cardiography and angiography. The coronary arteries are widely patent, large and of smooth bore. Some cases may be detected before the onset of clinical heart failure by the presence of cardiomegaly and gallop rhythm in association with symptoms of dyspnoea and fatigue. Since congestive cardiomyopathy commonly presents as heart failure of unknown cause it must be distinguished from 'rare specific heart muscle disease' (secondary cardiomyopathy) in which the myocardium is damaged and heart failure results from a general system disease or poison that also involves the heart such as sarcoidosis, connective tissue disorders and drugs such as doxorubicin (Goodwin, 1976).

The incidence of congestive cardiomyopathy is unknown because of lack of uniformity of diagnostic criteria. It may be confused with certain forms of coronary heart disease and may require coronary arteriography for elucidation. In most cases, however, a diagnosis of congestive cardiomyopathy can be made on clinical and investigative grounds if care is taken to rule out other cardiac disorders and diseases elsewhere in the body. Since congestive cardiomyopathy has a widespread geographical distribution it merits extensive study.

Several years ago I suggested that a multi-national trial should be undertaken to determine the prevalence of the disease, to refine the diagnostic criteria and to examine the epidemiology and myocardial characteristics. It was proposed that this study should be carried out by the Scientific Council of the International Society and Federation of Cardiology under the guidance of Dr Eckhardt Olsen and Dr...
Celia Oakley. Experts from a wide geographical area have enthusiastically entered the project and you will hear today of the special problems in certain areas of the world, and especially about the incidence, diagnosis, natural history and prognosis. The results of special investigations such as endomyocardial biopsy, immunological and enzyme studies will be presented. By pooling knowledge of all these aspects a pattern of the disease should emerge which may be expected to lead to advances in treatment as a result of improved diagnosis and understanding.

It seems clear that congestive cardiomyopathy is a multifactorial problem. Related factors, which may be causal, are: systemic hypertension, alcohol, virus infection, pregnancy and the puerperium, and immunological disorders. Hereditary factors appear to play little if any part. Cumulative or conditioning factors may operate to produce the disease in an individual who has more than one factor, such as, for example, a heavy alcoholic intake and previous virus infection, or an auto-immune disturbance resulting from a previous virus infection. These are, however, postulates that require substantiation.

While systemic hypertension is present in the minority of patients and may be a cause in some (Brockington and Edington, 1972), most patients are not hypertensive at any time. Alcohol is not usually a factor but can produce congestive cardiomyopathy if taken in excess (Brigden and Robinson, 1964). Congestive cardiomyopathy is well known to occur in the latter half of pregnancy or puerperium in certain patients (peripartum cardiomyopathy) (De Makis and Rahimtoola, 1971). Virus myocarditis has been thought possibly to lead to congestive cardiomyopathy although proof is lacking but fluorescent antibodies have been demonstrated in the myocardium (Kawai, 1971). Professor Waterson will describe the association between high blood titres to Coxsackie B4 infection and congestive cardiomyopathy. Immunological abnormalities have been described in congestive cardiomyopathy (Das et al., 1972) but it is not known whether these are due to previous infection.

The striking normality of the coronary arteries which may appear to be more healthy than would be expected for the normal range in an equivalent age and sex group, suggests that there may be some clue here to aetiology, but if so it has not yet emerged.

The concerted efforts of experts in many fields in many countries with pooling of data should yield important results. This Workshop today will clarify the data obtained so far and illuminate the way to further research.

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References
Congestive cardiomyopathy. Introduction, problems and aims of the Multicentre Research Project.

J. F. Goodwin

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