# Doppler assessment of the left ventricular diastolic function in cases of dilated cardiomyopathy

submitted for partial fulfillment
of the Master Degree of Cardiology



By

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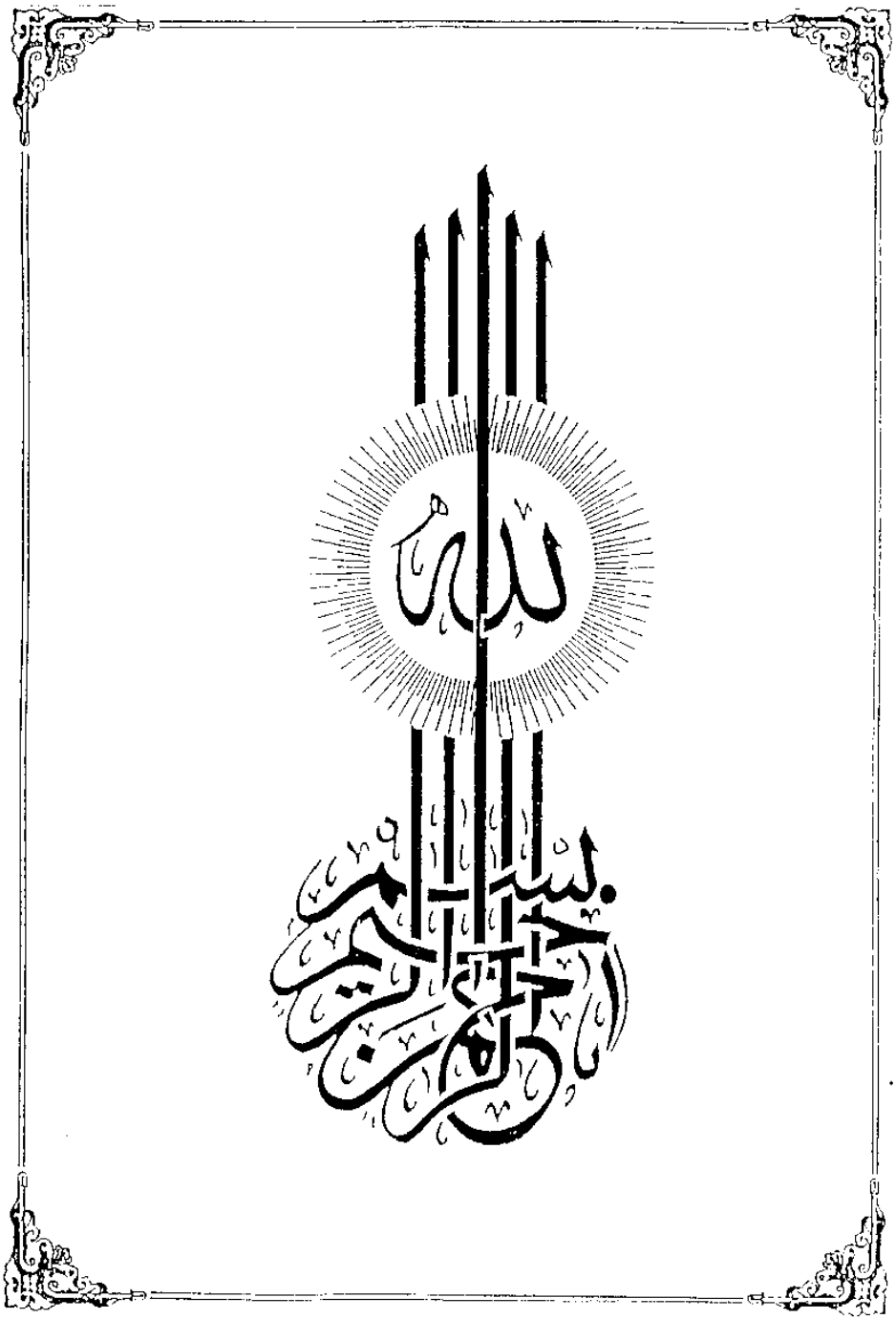

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# INTRODUCTION AND AIM OF WORK

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Congestive heart failure is a clinical syndrome associated with a broad spectrum of myocardial dysfunction patterns, with varying components of systolic and diastolic impairment *(Dougherty et al., 1984).*

Diastolic dysfunction of the left ventricle is defined as an impaired capacity to accept blood or fill without a compensatory increase in left atrial pressure. On its mildest form, diastolic dysfunction may appear as a slow or delayed pattern of relaxation and filling, with little or no elevation of left ventricular diastolic pressure (there is generally little or no systolic dysfunction); thus, in patients with left ventricular hypertrophy or coronary artery disease, an alteration in diastolic filling can serve as a sensitive and early indicator of the disease. Diastolic dysfunction may also appear as overt congestive heart failure, even in the presence of normal or near normal systolic function *(Stauffer et al., 1990).*

*Echeverria et al., (1983),* using echocardiographic techniques have noted normal systolic function in 40% of 50 consecutive patients with congestive heart failure. In addition, *Dougherty et al., (1984)* noted a normal or near normal radionuclide ejection fraction (> 7.45%) in 36% of a consecutive series of patients with congestive heart failure.

*Waronowicz et al., (1983)* noted that 10 of 39 patients with acute myocardial infarction and pulmonary edema manifested normal left ventricular ejection fraction before hospital discharge.

*Dougherty et al., (1984)* have suggested that the clinical manifestations of congestive heart failure are more closely associated with diastolic than systolic properties of the heart. Similarly, no correlation between severity of symptoms and ejection fraction was found. Also, ejection fraction is not predictive of the treadmill exercise duration in patients with congestive heart failure.

Unfortunately, it is commonly believed that the clinical examination can reliably separate those individuals with congestive heart failure with primary diastolic dysfunction from those with primary systolic dysfunction. Jugular venous, pressure, basal lung rales, a displaced apical impulses, a $S_3$ gallop, and edema do indeed identify the average patient with classic systolic dysfunction and a big, baggy heart. However, the current concept suggests that the majority of patients with principally diastolic dysfunction will have predominantly left-sided findings with a forceful, minimally displaced apical impulse, a fourth heart sound, and no third heart sound. While this may be true in some patients, clinical distinction between heart failure associated mainly with systolic versus diastolic dysfunction usually cannot be made at bedside *(Kessler, 1988).*

Recent studies have shown the utility of Doppler echocardiography in assessing left ventricular diastolic function. Indices of diastolic function obtained with this technique correlate well and are probably superior to those obtained by contrast or radionuclide angiography *(Vanoverschelde et al., 1990).*

Patients with dilated cardiomyopathy have been shown to have, not only abnormal left ventricular systolic function, but also abnormal

diastolic function. Impaired left ventricular relaxation and abnormal diastolic left ventricular compliance have been reported *(Takenaka et al., 1986).*

A complicating factor is that patients with dilated cardiomyopathy frequently have mitral regurgitation, which affects left ventricular filling by increasing left atrial pressure in early diastole *(Takenaka et al., 1986).*

## Aim of the Study

The aim of this work is to assess the left ventricular diastolic function in cases of idiopathic dilated cardiomyopathy using Doppler echocardiography.

# REVIEW OF LITERATURE

# REVIEW OF LITERATURE IN DILATED CARDIOMYOPATHY

## Historical background

The term cardiomyopathy started to appear in the literature following Brigden paper *(Emmanuel et al., 1983).*

***Brigden*** published his lecture in 1957 on uncommon myocardial disease; the non coronary cardiomyopathy and pointed out the diversity of the disorder and the difficulty of classification and was among the first to use the term cardiomyopathy *(Goodwin, 1982).*

***In 1961 Goodwin and his colleagues*** published their first paper defining cardiomyopathy and attempting a classification. Then they defined cardiomyopathy as acute, subacute or chronic disorder of heart muscle of unknown etiology often with associated involvement of endocardium or sometimes the pericardium, but not of atherosclerotic origin and the term primary myocardial disorder was then used *(Goodwin, 1982).*

Later in conjunction with ***Dr. Celia Oakley (Goodwin, 1982),*** the definition of cardiomyopathy was simplified to **"a disorder of cardiac muscle of unknown cause".**

Classification based upon disorders of structure and function was introduced in 1964, suggesting that cardiomyopathies may present clinically in one of three ways; congestive, constrictive, or obstructive as

separate distinct entities, not merging from one into the other. The group of obliterative cardiomyopathy was introduced indicating the effect of endomyocardial fibrosis in obliterating the cavity of the ventricle. Furthermore, three changes have been made recently in the classification and the term congestive is now felt inappropriate as cases can now be recognized before overt congestive failure has occurred, however, the word is retained for its frequent usage and the possibility of confusion on its omission. The word obstructive has been omitted and is simply known as hypertrophic. The restrictive and obliterative types are combined into one group restrictive/obliterative type when resulting from endomyocardial fibrosis *(Goodwin, 1982).*

## Definition and Classification of Cardiomyopathy

Cardiomyopathies are defined as heart muscle diseases of unknown cause according to the *World Health Organization, International Society and Federation of Cardiology task force WHO/ICSF.* They are classified into three major groups on the basis of their clinical features, hypertrophic, dilated (congestive) and restrictive types *(Kawai et al., 1983).*

The dilated or congestive type is not necessarily associated with congestive heart failure, but rather characterized by ventricular dilatation and poor systolic function with greatly reduced ejection fraction *(Keren, 1985).*

## Etiology of Dilated Cardiomyopathy

The specific etiology of dilated (congestive) cardiomyopathy remains obscure *(Ozick et al., 1984).*

In contrast to the definition of the *WHO/ICSF* task force the term dilated cardiomyopathy was not reserved for the primary type but was used for secondary forms such as alcoholic or postmyocarditis disease *(Maish et al., 1983).*

It is still controversial whether dilated cardiomyopathy is the terminal stage of etiologically different diseases or if a single etiologically, yet unknown, myocardial disease is provoked by different conditioning factors such as viral infections and immunologic effector mechanisms *(Maish et al., 1983).*

There are possible associated or conditioning factors related to cardiomyopathy in adults including high and prolonged alcohol intake, high blood pressure, pregnancy or puerperium and a disorder of cellular immunity resulting from infections *(Goodwin, 1982).*

Cardiomyopathy has long been thought to involve immune dysfunction but the immunopathy is not completely understood *(Kawai et al., 1983).*

Reduced suppressor cell activity has also been reported in congestive cardiomyopathy and myocarditis *(Kawai et al., 1983).*

The percentage of total T-cells, helper/inducer cells and suppressor/cytotoxic cells was studied using monoclonal antibodies in 20 African patients with dilated cardiomyopathy and in 20 aged matched normal control subjects. The percentage of helper/inducer cells was significantly higher in idiopathic dilated cardiomyopathy patients than in normal subjects, suggesting that an excessive immune reaction was

involved in the pathogenesis of this disease in Africans *(Sanderson et al., 1985).*

Genetic factors in dilated cardiomyopathy are little understood and are certainly less important than in hypertrophic type *(Emmanuel et al., 1983).*

Familial incidence in dilated cardiomyopathy has been reported in the literature and there is some evidence that it may be inherited as a recessive character, but nothing of substance in the literature has suggested that it has an autosomal inherited form *(Emmanuel et al., 1983).*

In a case report, monozygotic twins had related cardiomyopathy with evidence of an autoimmune process involving both the heart and thyroid gland *(Ozick et al., 1984).*

One of the most attractive hypotheses concerning the etiology of dilated cardiomyopathy, having many supporters, links the disease to prior viral myocarditis. It is presumed that a viral infection, either overt or clinically silent attacks cardiac antigenic structure stimulating an autoimmune response leading after a latent period to cardiomyopathy *(Kopecky et al., 1987).*

Fortunately, acute viral myocarditis results in complete recovery in the vast majority of patients but in a small percentage, heart failure may occur acutely, persist and be indistinguishable from cardiomyopathy except by endomyocardial biopsy. In other patients, there may be a latent period after recovery during which the autoimmune process develops and results in the established syndrome of cardiomyopathy

years later. Some patients may have a genetically determined deficiency in cellular immunity rendering them liable to viral attacks on the heart *(Goodwin, 1982).*

Myocarditis as a cause of acute congestive cardiomyopathy has been described following a wide variety of rickettsial bacterial, protozoal and metazoal diseases. The most common agents producing myocarditis in North America are viruses. In the Western World, myocarditis is considered a viral disease until proven otherwise *(Kereiakes, 1984).*

Infectious agents may cause myocardial damage in three ways; by invasion into the heart muscle, by production of a myocardial toxin, or via autoimmune reaction *(Kopecky et al., 1987).*

The picorna viruses (echovirus, poliovirus, coxsackie A and B viruses) are the most common causes of viral myocarditis with the coxsackie B group being the most frequent with approximately 5% of all coxsackie viral infections involving the heart *(Kopecky et al., 1987).*

### *Incidence*

In developed countries, the annual incidence of cardiomyopathy ranges from 0.7 to 7.5 cases per 100,000 population, and the prevalence in England has been reported as 8.317 cases per 100,000 population. In less developed countries, especially in the tropics, cardiomyopathies are more prevalent and also constitute a greater if not dominant fraction of heart disease. This greater prevalence is largely in the form of dilated cardiomyopathy and to some extent restrictive cardiomyopathy *(Abelmann et al., 1989).*

The incidence of dilated cardiomyopathy in Western Hemisphere is approximately 7 - 10 per 100,000 persons per year, but there is a lack of epidemiologic data particularly as regards to Africa and Asia and newer methods of diagnosis and earlier recognition of the disease will likely alter the incidence rate *(Kopecky et al., 1987).*

### *Pathology and Histological Findings*

Pathology yields no clues; the heart is dilated and overweight but the thickness of the ventricular wall is not increased, there are no valvular or coronary artery diseases *(Goodwin et al., 1982).*

The heart weights at autopsy are usually 400 gm (normal weight is 300 gm) but may be in excess of 1000 gm. The coronary arteries are usually normal and there is often dilatation and thickness of the mitral and tricuspid valve rings with chronic regurgitation. Thrombi are found in over 50% of non anticoagulated patients and are usually multiple and located in the left ventricular apex. The histological findings are abnormal but non specific and the diagnosis pathologically is one of exclusion when valvular, pericardial, coronary and congenital diseases have been excluded *(Kopecky et al., 1987).*

Indications of myocardial cellular degeneration (fibrosis, lipofusin pigment accumulation) and intracellular vacuolization are seen more commonly in the left ventricle than in the right. The number of interstitial inflammatory cells does not vary between patients with dilated cardiomyopathy and controls *(Kopecky et al., 1987).*

*Amorism and colleagues, (1982)* made pathological studies in seven hearts of patients with idiopathic dilated cardiomyopathy and they