

# **Correlation Between Incidence of Arrhythmias and Left Ventricular Systolic and Diastolic Functions in Idiopathic Dilated Cardiomyopathy**

**Thesis**

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of master degree in Cardiology

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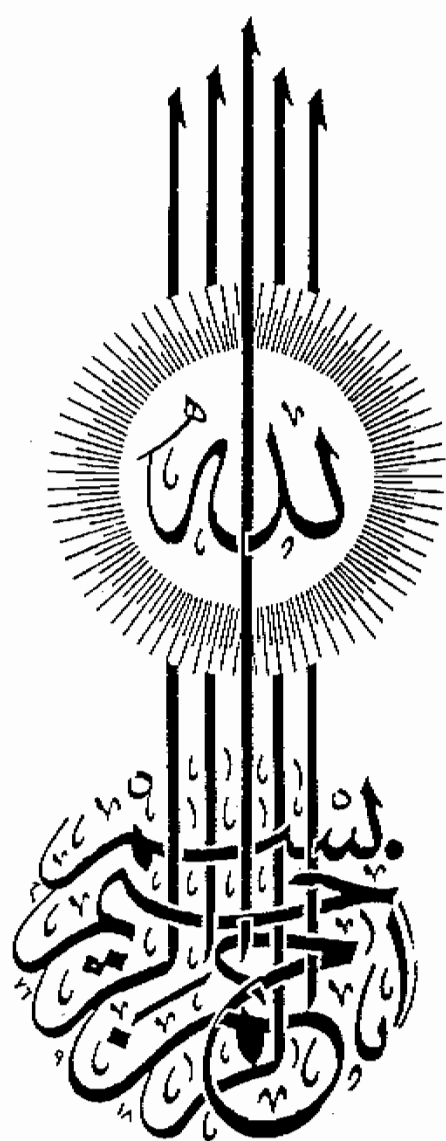
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## TABLE OF CONTENTS

	Page
ACKNOWLEDGMENTS .....	1
INTRODUCTION .....	1
A AIM OF THE WORK .....	3
REVIEW OF LITERATURE .....	4
SUBJECTS AND METHODS .....	51
RESULTS .....	58
DISCUSSION .....	87
SUMMARY AND CONCLUSION .....	96
REFERENCES .....	102

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## INTRODUCTION

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Cardiomyopathy is a disease of heart muscle caused by a spectrum of known (secondary) or unknown (primary or idiopathic) factors (Brandenburg Ro. et al., 1981). A variety of schemes for classifying the cardiomyopathy have been proposed Goodwin's classification into congestive, hypertrophic and restrictive cardiomyopathy has been widely accepted [Goodwin J.F., 1970].

More recently the term dilated has been substituted for congestive as the preferred terminology that is idiopathic dilated cardiomyopathy (I.D.C.) [Fuster V. et al., 1981].

Studies on dilated cardiomyopathy have focused primarily on congestive heart failure relatively little attention have been given to arrhythmias despite considerable evidence that ventricular arrhythmia and sudden death are important problem in this disease (Jhonson et al., 1982).

Long term dynamic electro cardiographic (Holter monitoring) has been useful in detecting the significance of cardiac arrhythmias in different diseases [Anderson K P et al., 1978] [McKenna Wj et al., 1981]. With this diagnostic tool many studies have been focused on the prognostic significance of V. P. C's and V. T. in patients with coronary artery diseases and hypertrophic cardiomyopathy [McKenna Wj et al., 1981] [Maron BJ et al., 1981].

However little is known about the prevalence of ventricular arrhythmias evaluated by long term Holter monitoring and their clinical significance in patients with I.D.C.

In the past evaluation of the myocardium has been limited to examining systolic functions of the heart. Recently however, investigations have demonstrated that abnormalities of diastolic functions of the heart provide an important contribution to the signs and symptoms expressed by patients with heart disease. In addition abnormalities in diastolic functions may precede abnormalities of systolic functions in early stage of disease [Nishimura et al., 1989]. Evaluation of diastolic filling of the heart has been difficult because its complexity and numerous interrelated contributing factors. Previous studies have depended on high fidelity, invasive measurements of instantaneous pressure, volume, mass, and wall stress which could not be done on a routine clinical basis. With the advent of Doppler echocardiography intracardiac blood flow velocities can now be done non invasively [De Maria et al, 1987 and Nishimura et al, 1989].



## AIM OF THE WORK

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## REVIEW OF LITERATURE

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## THE FRONTIERS OF CARDIOMYOPATHY

Three decads ago the frontiers of cardiomyopathy were ill defined and nebulous because no working definition or classification had been suggested and elemination was imprecise [Goodwin J.F., 1982]. In the United States of America, Mattingly, Burch and Proctor Harvey were selecting and studying cases in the 1950's.

In 1957 Brigden published his lecture on "Uncommon myocardial diseases, the non coronary cardiomyopathies" [Goodwin J.F., 1982]. Brigden pointed out the diversity of the disorder and the difficulty of classification and was among the first to use the term "Cardiomyopathy".

In 1961 Goodwin and his colleagues; Gordon, Hollman and Bishop published their first paper defining cardiomyopathy and attempting a classification. They defined cardiomyopathies as acute, subacute, or chronic disorders of heart muscle of unknown aetiology 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 Okley, the definition of

cardiomyopathy was simplified to "a disorder of cardiac muscle of unknown cause" while myocardial disorders that were part of a general systemic disease were termed "rare specific heart muscle diseases". Thus, automatically any condition in which the cause or pathological process could be defined clearly was excluded from the definition of cardiomyopathy. This concept may be rigid for the clinician because the clinical features of a given cardiomyopathy are often identical to those of one of the specific heart muscle diseases, but this concept received approval from the joint task force of the WHO and ISFC.

The term secondary cardiomyopathy may be used to identify those patients with a specific heart muscle disease that clinically closely simulates an idiopathic "primary" cardiomyopathy. In 1964, classification based on disorders of structures and function was introduced suggesting that cardiomyopathies may present clinically in one of three ways: Congestive, Constrictive or obstructive as separate distinct entities. The word obstructive has been omitted and is simply known as hypertrophic. The term congestive cardiomyopathy is derived from the common late clinical manifestation of congestive heart failure. However, dilatation of the cardiac chambers (particularly the left ventricle) is an earlier and predominant feature, and for this reason the term dilated cardiomyopathy is preferred (ISFC/WHO 1978).

## DEFINITION

Dilated cardiomyopathy has been characterized as: A form of cardiomyopathy in which there is impaired function of the heart as a pump with dilatation of the ventricles and reduced systolic function [Goodwin, 1982].

Mildly dilated cardiomyopathy is an unusual subtype of dilated cardiomyopathy in which there is severe L.V. dysfunction but only mild dilatation. A family history of dilated cardiomyopathy is frequent and females are more frequently affected than males, prognosis is poor [Treasure CB. et al 1990].

## INCIDENCE

There is a paucity of epidemiologic data dealing with the incidence and prevalence of dilated cardiomyopathy.

In 1982 in the United States a total of 10,345 deaths, 46,000 hospitalizations, and 410,000 days of hospital care were attributed to cardiomyopathy according to the National Center for Health Care Statistics. Approximately 80% of these patients were diagnosed as "primary" or idiopathic dilated cardiomyopathy [Gillum RF, 1986].

In 1982 the age adjusted prevalence rates for persons aged 35 to 74 years were as follows: white males 11 per 100,000; white females 4.4 per 100,000; black males 27 per 100,000 and black females 11 per 100,000. In this age bracket, the male to female ratio was 2.5 : 1 in whites and 2.4 : 1 in blacks. The black to white ratio was 2.4 : 1 in males and 2.6 : 1 in females. One of the few studies was done by Torp et al at the hospital in Malmo, Sweden, the investigators found a prevalence rate of 7.5 cases per 100,000 inhabitants per year between 1970 and 1979 with a male to female ratio of 3 : 1 and a mean age of 47 years.

In two studies done in Denmark in 1980-1981, 41 cases of dilated cardiomyopathy were identified giving an estimated incidence of at least 0.7 cases per 100,000 residents (Bagger JP 1984). The prevalence of dilated cardiomyopathy in England has been estimated at 8.3 cases per 100,000 population (Williams DG, 1985). A Mayo clinic population-based study in Olmsted County, Minnesota, found an incidence rate of 5.4 per 100,000 population and rate of 32.3 per 100,000 (Codd MB, 1987).

In summary, the incidence of dilated cardiomyopathy in the Western hemisphere is approximately 7-10 per 100,000 persons per year but there are a lack of epidemiologic data particularly in regard to Africa and Asia. Newer methods of diagnosis and earlier recognition of the disease will likely alter the incidence rates.

## **PATHOLOGY**

### **GROSS PATHOLOGY**

The typical features of dilated cardiomyopathy are dilatation of all cardiac chambers, the ventricles are more dilated than the atria [Roberts W.C., 1987]. Although in the majority of patients, enlargement of the left ventricle is greater than that of the right, selective right ventricular dilatation has been noted [Fitchett DH, 1984].

Recently in a small group of patients, clinical and hemodynamic findings that are similar to those seen in dilated cardiomyopathy have been reported, though this group had only minimal cardiac dilatation [Keren A., 1985] i.e. Mildely dilated cardiomyopathy.

Heart weights at autopsy are usually 400-750 gm. (normal weight is 300 gm.) but may be in excess of 1000 gm.

Hypertrophy is present despite normal wall thickness due to the presence of some degree of dilatation. The development of left ventricular hypertrophy appears to have a protective or beneficial role in dilated cardiomyopathy because it may serve to reduce systolic wall stress and protect against further cavity