

بِسْمِ اللَّهِ الرَّحْمَنِ الرَّحِيمِ

وما أوتيتهم من العلم الا قليلاً

سورة الإسراء (آية ٨٥)

RETROSPECTIVE STUDY  
OF PATIENTS WITH CARDIOMYOPATHY  
DIAGNOSED AT THE PAEDIATRIC CARDIOLOGY  
OUT-PATIENT CLINIC OF AIN-SHAMS UNIVERSITY



Thesis

Submitted in partial fulfilment for the  
Master Degree in Paediatrics

By

Mohamed Mamdouh Mostafa  
M.B.B.Ch.

Supervisor

Prof. Dr. ABD EL KHALIK KHATTAB  
- Faculty of Medicine  
Ain Shams University

1986

618.92124  
M.M

24939

35.9

27 up 1986

A.E. 1986

7/11/86



To My Parents

### *ACKNOWLEDGEMENT*

*I would like to express my sincere gratitude to Prof. Dr. Abd El-Khalek Khattab for his kind guidance and supervision.*

*Mamdouh Eldighaidy*

*1986*

## AIM OF WORK

## Aim of Work

The aim of this work is to find out the exact incidence of various types of cardiomyopathy, i.e., congestive, hypertrophic, and restrictive types. Also, we try to find out the value of clinical examination and invasive and non invasive techniques in the diagnosis of cardiomyopathy.

## CONTENTS

	Page
Introduction .....	1
Definition .....	6
Classification .....	7
Review of Literature	
* Dilated Cardiomyopathy .....	10
* Hypertrophic Cardiomyopathy .....	20
* Restrictive Cardiomyopathy .....	38
Material and Methods .....	47
Results .....	48
Discussion .....	57
Summary and Conclusion .....	63
References .....	65
Arabic Summary	



## **INTRODUCTION**

The existence of primary myocardial disease has been recognized in the world for more than 100 years ago (Massumi et al., 1965). In 1891, "Krehl" an outstanding clinician and pathologist and perhaps the father of pathological physiology was one of the first to investigate and write about "idiopathic disease" of heart muscle. His clinical and pathologic descriptions on a series of cases which he observed, fit well into what is described today as "idiopathic cardiomyopathy" or "idiopathic myocardial hypertrophy". He believed that the pathological changes were chronic and non inflammatory which eventually led to myocardial failure.

Shortly after the above reports by "Krehl", observation and reports by "Fiedler" in 1899 and by "Josserand" and "Gallavardin" in 1901, resulted in the introduction of other new terms to describe forms of idiopathic myocardial disease (Mattingly, 1973).

The first published description of idiopathic hypertrophic subaortic stenosis (I.H.S.S.) appears to be that of Schmincke of "Wurzburg" Germany, who in 1907, presented gross pathologic findings in two adult women. Each had diffuse "hyperplasia" of muscle mass constituting the wall of the left ventricular, outflow tract.

Three years later after Schmincke's report, "Bernheim" included several patients with eccentric left ventricular

hypertrophy of unknown etiology, in whom the hypertrophied septum bulged into and tended to obliterate the Rt. ventricular cavity. In retrospect, it is likely, although not certain, that some "Bernheim's" cases were examples of "I.H.S.S." (Braunwald et al., 1964).

Howland reported on young children in whom idiopathic myocardial hypertrophy and failure were only findings at autopsy (Braunwald et al., 1964).

"Brigden" (1957) was the first one who used the term "cardiomyopathy" to describe disorders of the myocardium of unknown cause.

Brock (1957) published a report of a patient believed to have been suffering from aortic valve disease but who at operation was found to have muscular hypertrophy of the interventricular septum.

Tear (1958) described eight patients whose heart at necropsy showed asymmetric hypertrophy of the heart. This condition had already been described in the literature of the 19th century.

In 1960, Goodwin et al., studied patients with idiopathic asymmetrical hypertrophy like that previously reported by Tear and observed similar haemodynamic findings. He concluded that ventricular septal hypertrophy produced a functional obstruction to the outflow of one or both ventricles. They called the condition "obstructive

cardiomyopathy" to differentiate it from congestive and constrictive forms of cardiomyopathy.

In 1962, "Braunwald" et al., introduced the term "idiopathic hypertrophic subaortic stenosis. This term remains the most frequently used term in U.S.A. whereas Goodwin's term of "hypertrophic obstructive cardiomyopath" is more generally used throughout the rest of the world (Mattingly, 1973) .

The ventricular septal thickness to left ventricular posterior wall ratio was first described echocardiographically by Abbasi et al. (1972), and a year later Henry et al. (1973) claimed that a ventricular septal thickness to posterior wall ratio greater than or equal to 1.3 which he termed "asymmetric septal hypertrophy" was a pathognomonic echocardiographic abnormality of hypertrophic cardiomyopathy (Doi et al., 1980).

Alcohol was recognized in the U.S.A. as a potential cardiotoxic agent as early as 1855 (Wood, 1855). Alcoholic cardiomyopathy was described in the United States in 1956 by "Eliaser" and "Griansiracusa".

Post partial cardiomyopathy has been recognized for many years (Hull & Hafkesbring, 1973 ; Turner, 1938 and Walsh et al., 1965).

The first account of Ht disease secondary to coxsackie B. virus infection in adults came from Northern Ireland (Fletcher & Brennan, 1958).

The clinical syndrome of ischemic cardiomyopathy has been recognized in England by Ratteny, Banks and Aram (1969).

#### Incidence of Cardiomyopathy in the World

The precise incidence of cardiomyopathy is not known in the united states, nor in other countries of the world, although heart muscle disease is one of the most common cardiac and clinical problems confronting man today.

By 1958, only 72 patients with idiopathic cardiomyopathy were apparently reported in the world literature (Spodick & Littmann, 1958). The world health organization indicated in 1965 that even population studies would not provide reliable data concerning the true incidence of the various types of cardiomyopathy (Bull, WHO, 1965). This may be due to failure to obtain accurate histories from patients, lack of recognition of myocardial disease by the physician, inadequate diagnostic facilities, insufficient time spent with the patient and other factors resulting in adequate and inaccurate data (Burch & Giles, 1974).

Torp (1978) studied a total population of 250,000 during a period of eight years. This study was based on cases investigated at Malmo general hospital. The city of Malmo is located in the south of Sweden. The diagnosis of congestive cardiomyopathy could be firmly established in 95 cases during

cardiomyopathy could be firmly established in 95 cases during this period. This gives us an idea about the incidence of the disease of  $(3/100,000)/\text{year}$ . If cases disclosed at post-mortem during the same period are included the frequency of the disease will equal  $(5/100,000)/\text{year}$  (Torp, 1978).

## **DEFINITION**