ELECTROCARDIOGRAPHIC STUDY IN CHILDREN WITH VENTRICULAR SEPTAL DEFECT

THESIS

Submitted for Partial Fulfilment for the
MASTER DEGREE OF PEDIATRICS

By

Dr. Younes Mohamed Morsy
M.B., B.Ch.

Faculty of Medicine
Ain Shams University

618.92 12.75 4
Y. M

30159

Under Supervision of

Prof. Dr. Amal Ayoub
Professor of Cardiology
Ain Shams University

Dr. Shereen Abd Al Fatah
Assistant Professor of
Pediatric
Ain Shams University

Dr. Said Khaled
Lecturer of Cardiology
Ain Shams University

1989

Central Library - Ain Shams University

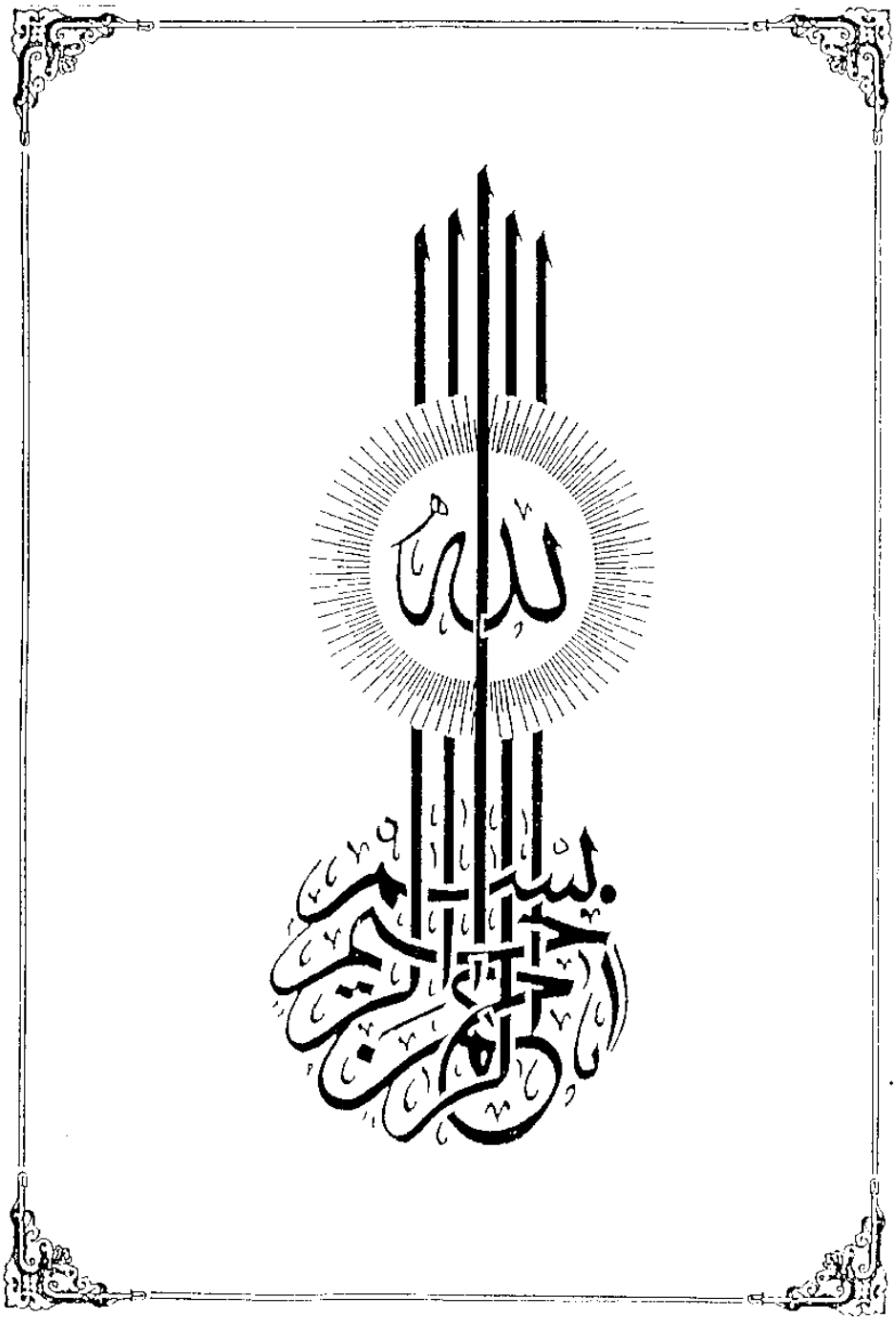
ACKNOWLEDGEMENT

I wood like to express my deepest gratitude to Prof. Dr. Amal Ayoub, Professor of Cardiology, Ain Shams University Hospitals for her kind guidance, encouragement and supervision all through this study.

I would like also to express my thanks and appreciation to Dr. Shereen AbdelAl Fatah , Assistant Professor of of Pediatric and Dr. Said Khaled, Lecturer of Cardiology, Ain Shams University Hospitals for great help in preparing and writing this thesis.

I would also like to thank all the members of the Department of Cardiology and Pediateric .

Central Library - Ain Shams University



Central Library - Ain Shams University

CONTENTS

Central Library - Ain Shams University

HISTORY AND INTRODUCTION

Central Library - Ain Shams University

HISTORY AND INTRODUCTION

Duper in 1891 presented the first case diagnosed during life and confirmed by autopsy. He proposed the name "Maladie de Roger" , (Cooley and Hallman, 1975).

The term "Maladie de Roger " or "Rogers" disease" is generally considered synonymous with asmall V.S.D., (Fowler, 1980).

Isolated V.S.D. is the most common congenital heart defect, occuring at a rate of slightly over 2 per 1,000 live births and constituting over twenty of all forms of congenital heart disease. (Wells and Lindesmith, 1985).

The effect of V.S.D. on the patient depends primarily on the size of the defect, and in large communication on the resistance to blood flow through the lungs. (Graham et al., 1983).

About 25 % children with V.S.D. develop symptoms, usually during infancy. The remainder will be symptom free through childhood. Even among the former, the possibility of a relative decrease in size or of spontanous closure of the defect is appreciable. (Dickinson et al., 1981).

The high incidence of spontaneous closure or reduction in the size of the defect introduces an interesting challenge in managing infants and children with

Central Library - Ain Shams University

ventricular septal defect. The physician caring for these patients often must match the risks of development of pulmonary vascular disease, bacterial endocarditis, and the risks of surgery itself against the chances of spontaneous closure (Rudolph, 1978).

Central Library - Ain Shams University

REVIEW OF LITERATURE

Central Library - Ain Shams University

Prevalence and Aetiology :

*Twenty percent of congenital heart patients have venticular septal defect as a solitary lesion (Graham et al., 1983).

* The incidence of V.S.D. in all live births is approximately 1.5- 2.5 /1000. The lower the prevalence found by groups caring for adult with congenital heart diseases is probably in large part due to spontaneous closure of a significant number of defect (Graham et al. 1983).

* Ventricular septal defect is found slightly more frequently in Females as compared to males with the experience of Hoffman and Rudolph representive of most series (56 % females 44 % males) (Hoffman and Rudolph, 1965).

* Ventricular septal defect is the most common lesion in the majority of the described chromosomal syndromes including the 13- Trisomy, 18- Trisomy and 21-Trisomy groups. In the majority of patients with V.S.D. (more than 95 %), the defect is unassociated with chromosomal abnormality, and the cause is unknown. Amultifactorial aetiology has been assumed in which interaction between hereditary predisposition and environmental influences results in the defect (Nora and Fraser, 1974).

Central Library - Ain Shams University

I- Chromesomaal Causes :

* Down syndrome is the most common chromosomal abnormality.

The incidence of cardiovascular malformation in Down syndrome is about 40 % V.S.D. makes up approximately 28- 33 % of patients with down syndrome who are found to have congenital heart disease (Spicer 1984).

* Trisomy- 13 also shows cardiac malformation in about 80 % of patients mostly in the form of dextrocardia with V.S.D. and pulmonary hyper tension (Tayler, 1968).

II- Single Mutant Gene :

* Mendelian disorders still appear to account for a very small (3 %) but very important subset of congenital heart disease (Nora and Nora, 1978).

III- Primary Environmental Factors :

* Only 2 % of congenital heart disease have an environmental, actiology with very little if any genetic contribution (Nora, 1983).

* However, certain environmental risks have been implicated.

Central Library - Ain Shams University

III- Congenital cardiovascular diseases as :

1. Drugs

Alcohol, Amphetamines, sex hormones and thalidomide.

2. Infections : as Rubella

3. Maternal conditions :

as Diabetes, phenyl ketonuria.

(Nora and Nora, 1978).

IV- Multifactorial Inheretance :

* The most popular concept regarding aetiology is that cardiac defects are due to a combination of genetic and environmental interaction. This multi factorial aetiology requires a genetic predisposition and an environmental Teratogen to which the susceptible fetus is exposed in the vulnerable period of cardiogensis. (Nugent et al., 1986).

* The persumed vulnerable period for teratogenic influence on the inter ventricular septum is between 14-44 days of gestation. (Nora, 1983).

Central Library - Ain Shams University

EMBRYOLOGY

The process of separation of the ventricles is intimately related to that of the aortic and pulmonary orifices at the distal end of the bulbus, and to the division of the Truncus arteriosus into pulmonary and aortic channels. These processes are so closely interdependent that they must be considered before the separation of the two ventricles.

The separation of the two ventricles from each other leaves the right ventricle in communication with the right atrium and with the pulmonary artery, and the left ventricle in communication with the left atrium and the aorta. This involves a series of complex changes in which three distinct factors contribute to the formation of the adult ventricular septum- (a) The ventricular septum, (b) The proximal bulbar septum, and (c) the atrioventricular endocardial cushions. (Warwick and Williams, 1973). At the 5 mm stage of embryonic development during the latter part of the fourth week. The heart has a common ventricular chamber. At this stage it begins to be partitioned by a primitive ventricular septum. Gradually this primitive septum is transformed into the definitive ventricular septum. This occur by the fusion of the primitive septum above with the endocardial cushions and the aorto pulmonary septum (Plauth, 1982). Untill this fusion is complete,an

Central Library - Ain Shams University

opening exists between these two structures, this is the interventricular foramen.

*In the process of obliteration of the interventricular foramen, failure of fusion between the aorticopulmonary septum and the primordial ventricular septum results in Type I and Type II defects.

Imperfect fusion between the endocardial cushions and the primordial ventricular septum produces a Type III defect. Faulty formation of the primordial, ventricular septum results in Type IV defect (Cooley, 1975).

Anatomy and Pathology :

The ventricular septum is considered as possessing muscular and membranous portions, the muscular septum itself is divided into Inlet, Trabecular and outlet (infundibular) components. The inlet muscular septum separates the septal cusps of the mitrals and tricuspid valves. The infundibular muscular septum separates the right and left ventricular outflow tracts. The trabecular septum, which is the largest part, separates the finely trabeculated left ventricular apex from the more coarsely trabeculated right ventricular apical zone. The membranous septum is a small structure that is divided into two parts by the insertion of the septal leaflet of the tricuspid valve.

Central Library - Ain Shams University

This insertion produce an atrio-ventricular and inter-ventricular components . (Soto et al., 1980).

Ventricular septal defect varies in anatomic location and is best considered in relation to morphologic land morks of the normal heart. The most important landmark is the crista supraventricularis, the muscular ridge that separates the main portion or body of the right ventricular cavity from the infundibular or outflow portion. Ventricular septal defects lie either above or below this ridge2 Defects that lie above this ridge are called supracristal and constitute about 8 % of isolated ventricular septal defect. Such defects are situated immediately beneath the pulmonary orific (sub-pulmonic) so that the valve forms part of the superior margin of the interventricular communication. When viewed from the left side, the supracristal defect is found just below the commissure joining the left and right aortic cusps (Perloff, 1978).

Some authors call these supracristal ventricular septal defects infundibular, conal, or sub-pulmonary (Moss, 1983).

Defects that lie below the crista are called infracristal and may be located either in the area of the membranous septum or the muscular septum, or in the site that would be occupied if the defect occurred as part

Central Library - Ain Shams University

of an endocardial cushions malformation (Perloff , 1978).

The most common variety of the ventricular septal defect is infracristal , lying below and posterior to the crista in the region of the membranous septum. These defeets are often partially hidden by the septal leaflet of the ticuspid valve. When viewed from the left side, they are found beneath the aortic valve(subaortic) close to the commissure joining the right and non-coronary cusps . The atrioventricular bundle as it travels from the AV node to the ventricles, passes along the posterior and inferior rim of the defect. Ventricular septal defects in this position are generally reffered to as membranous, although they usually involve an adjacent portion of the muscular septum (Perloff, 1978).

These defects constitute approximately 80 % of defeets seen at surgery or at autopsy (Moss, 1983). With the membranous defects, there can be avariable degree of malalignment between the infundibular septum and the interior ventricular septum such as aortic valve appears to override the defect. In addition when the septal commissure of the tricuspid valve is deficient at its attachement

Central Library - Ain Shams University