HISTOPATHOLOGICAL STUDY OF HEPATOMEGALY IN EGYPTIAN CHILDREN

THESIS

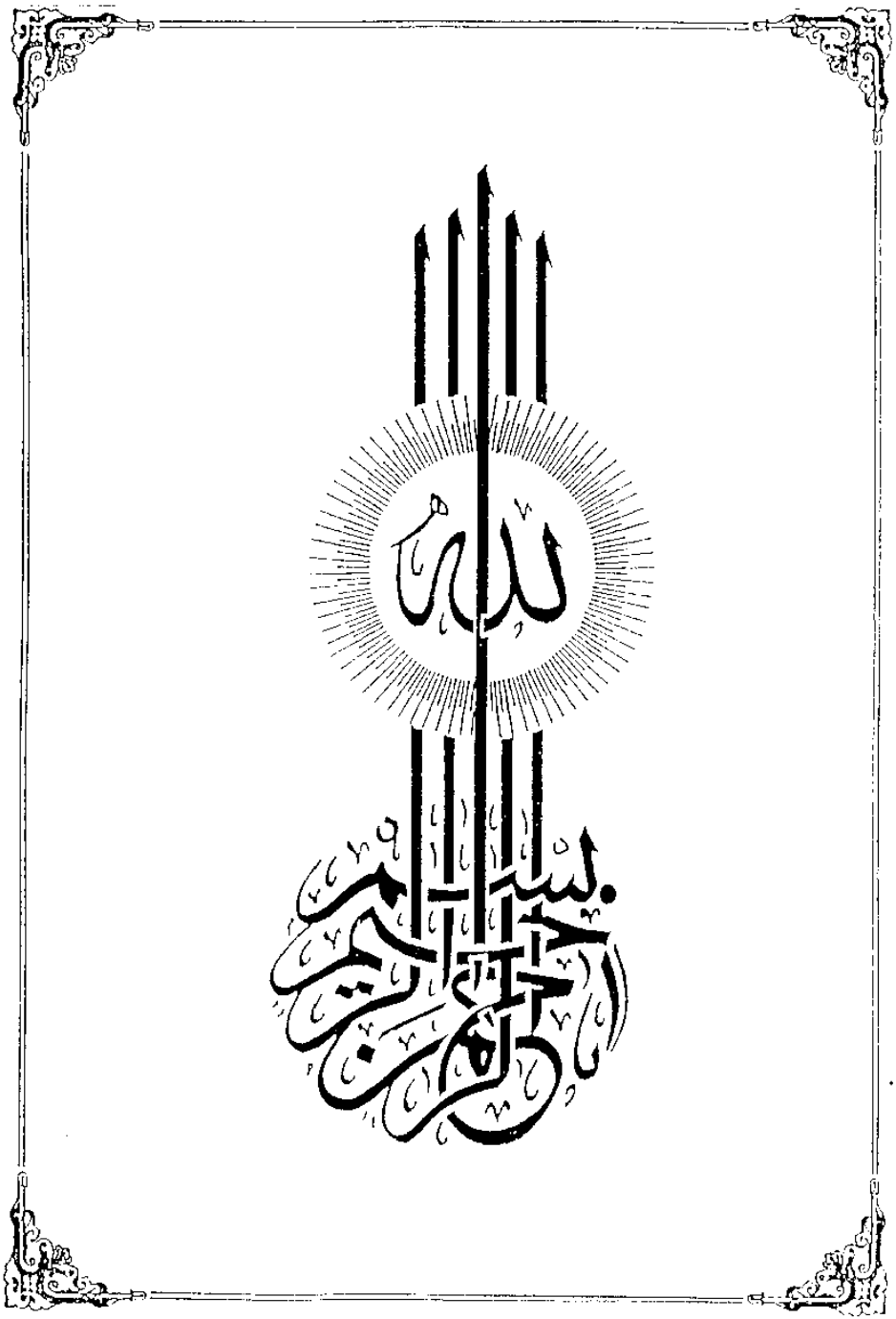
Submitted For Partial Fulfilment
of Master Degree of
(PAEDIATRICS)

By
ASHRAF ABDEL BAKY SALAMA
M. B., B. Ch.

Supervised by
Prof. Dr. SAADIA MOHAMED ABDEL FATTAH
Prof. of Paediatrics, Ain Shams University
Prof. Dr. SAWSAN M. AMIN ELSOKKARY
Prof. of Paediatrics, Ain Shams University
Dr. SHIRIN ABDEL FATTAH
Lecturer of Paediatrics, Ain Shams University

Faculty of Medicine
Ain Shams University

1987



ACKNOWLEDGEMENT

I wish to express my gratitude to Professor Dr. SAADIA ABD EL FATTAH Prof. of Paediatrics, Ain Shams University for giving me the privilege of working under her supervision and her patience and kind guidance throughout this work.

I am also indebted to Prof. Dr. SAWSAN EL SOKKARY Prof. of Paediatrics, Ain Shams University who was kind enough to offer me much of her time, valuable advice and experience.

I am really grateful to Dr. SHIRIN ABD EL FATTAH Lecturer of Paediatrics, Ain Shams University for her helpful efforts and instructions.

My deep thanks to Dr. HODA LOTFY, Lecturer of Paediatrics Ain Shams University for her meticulous revision of this work and her useful suggestions.

CONTENTS

* * *

ABBREVIATIONS

AAT	:	Alpha one antitrypsin
ALP	:	Alkaline phosphatase.
ALT	:	Alanine aminotransferase.
AST	:	Aspartate aminotransferase.
Bd	:	Bile duct.
CAH	:	Chronic active hepatitis.
CLH	:	Chronic lobular hepatitis.
Cmm		Cubic millimeter.
CMV	:	Cytomegalovirus.
CPH	:	Chronic Persistent hepatitis.
CV	:	Central vein.
ELISA	:	Enzyme Linked Immunosorbent Assay.
EPV	:	Epstein Barrvirus
GGT	:	Gamma Glutamyl transferase.
ha	:	hepatic artery
HAV	:	Hepatitis A virus
HBcAg	:	Hepatitis B core antigen
HBsAg	:	Hepatitis B surface antigen
HBV	:	Hepatitis B virus
HLA	:	Human leucocytic antigens
Ig	:	Immunoglobulin
Kg	:	Kilogram
ml	:	millilitre
MPS	:	Mucopolysaccharidoses

MRI	:	Magnetic Resonance Imaging
NANBV	:	Non A-non B viruses
PT	:	Prothrombin time
PV	:	Portal vein
Tc	:	Technitium
VH	:	Viral hepatitis
+ve	:	Positive
-ve	:	Negative
VOD	:	Veno occlusive disease.
CT	:	Computerised tomography.
ICC	:	Indian childhood cirrhosis.

INTRODUCTION AND AIM OF THE WORK

INTRODUCTION AND AIM OF THE WORK

Hepatomegaly is a relatively common finding in the paediatric age group. Diseases which may be associated with hepatomegaly are numerous. Clinically the diagnosis of hepatomegaly is best based on the liver span measurements. Laboratory data and useful imaging techniques are very important in the differential diagnosis of hepatomegaly. Liver biopsy inspite of being invasive technique is a very valuable investigation (Sherlock, 1985).

The aim of this work is to present cases of hepatomegaly due to different causes in the Paediatric Hepatology Clinic of Ain Shams University, in order to compare between the clinical, laboratory and sonographic data of these patients with the results of interpretation of their liver biopsies and to assess the diagnostic value of liver biopsy.

REVIEW OF LITERATURE

ANATOMY OF THE LIVER

Gross Anatomy:

The liver is the largest gland in the body. It accounts for 2% of the body weight in adults. In infants and young children, it is proportionately larger weighing approximately 5% of the body weight during the first year of life and gradually reducing in size over the subsequent years (Nayak and Ramalingaswami, 1979)[A].

The liver is situated in the right upper quadrant of the abdomen. It possesses five surfaces: The superior, anterior, lateral, inferior and posterior surfaces. There are two anatomical lobes, the right lobe being about six times the size of the left. Lesser segments of the right lobe are the quadrate lobe on its inferior surface and the caudate lobe on the posterior surface. The right and left lobes are separated anteriorly by the falciform ligament, inferiorly by the fissure for ligamentum teres and posteriorly by the fissure for ligamentum venosum (Elias, 1963).

Surface Anatomy:

The upper border extends from the fifth rib medially to the right of the mid clavicular line to the sixth rib in the left mid clavicular line. Its lower margin

crosses the epigastrium midway between xiphisternum and the umbilicus (Sherlock, 1985).

The lower border of the liver can be normally palpated 3 cm below the costal margin up to six months of age and less than 2 cm in children between four and ten years (Athreya, 1980).

Blood supply:

The blood supply is by hepatic artery, a branch of coeliac axis, and the portal vein. They enter through porta hepatis empting their blood into the sinusoids.Venous drainage is by hepatic veins into the inferior vena cava. (Elias and Sherric, 1969; Finlayson and Richmond,1984).

Histology of the liver:

A liver lobule is described as a polygonal prismatic formation constituting a mass of liver parenchyma which has in its centre a central vein and which is demarcated by planes connecting the adjacent portal tract. Liver cell plates are radially arranged, they are one cell thick and they are exposed on either side to hepatic sinusoids which converge towards central vein.

Rappaport(1963) on the other hand had put forward the concept of functional unit, "The acinus". This is centered around an axis comprising terminal portal venules, hepatic arterioles, bile ducts, lymph vessels and nerves. The axis extends from a small triangular portal tract and is extending in three dimensions,almost pear shaped.

An acinus has peripherally located central veins and is composed of parts of the adjoining lobules. (Poulsen and Christoffersen, 1979).

Hepatocyte is polygonal in shape. The nucleus is single or in mitosis. It has three surfaces: one facing the sinusoids, second facing the canaliculi and the third facing the neighbouring hepatocytes. The walls of the sinusoid consist of endothelial and phagocytic cells of reticuloendothelial system. The flat cell component is known as Kupffer cell (Sherlock 1985).

MECHANISMS ACCOUNTING FOR HEPATOMEGALY

Hepatomegaly is a physical finding which may suggest either intrinsic liver disease or may represent a component of a more generalised disorder. Hepatomegaly in the paediatric age group is a situation which may require extensive evaluation in order to distinguish benign self limited disease processes from serious life threatening conditions involving the liver (Chandra, 1979).

The pathophysiologic mechanisms involved in the sudden or gradual onset of liver enlargement are varied and complex. These include inflammation, infections, congestion, storage diseases, tumours and malignant infiltrations (Ewerbeck and Remischovsky, 1980).

Inflammation and infections:

An infection should be considered first if hepatomegaly is noted in children. The liver may be enlarged and firm to palpation in almost every viral infection in the infant because of intense response of the reticuloendothelial system to the infectious stimulus. Hepatomegaly may persist for a long time. This should remind the pediatrician to repeat the liver function studies since viral hepatitis as well as its sequelae are often