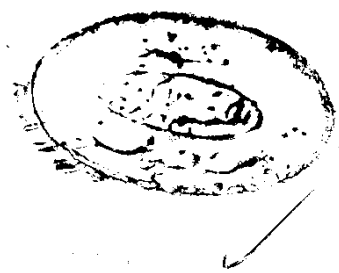
PORTAL HYPERTENSION IN INFANCY AND CHILDHOOD THESIS

Submitted For Partial Fulfilment Of
The Master Degree Of Pediatrics



BY

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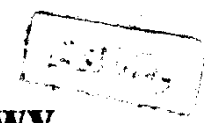

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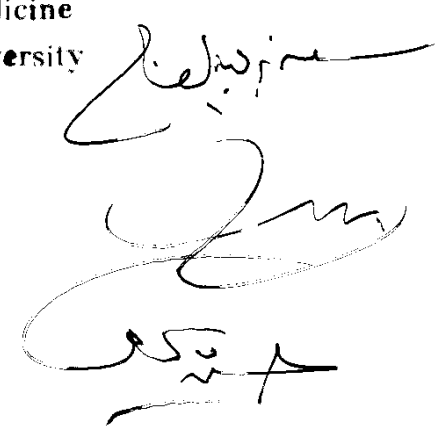

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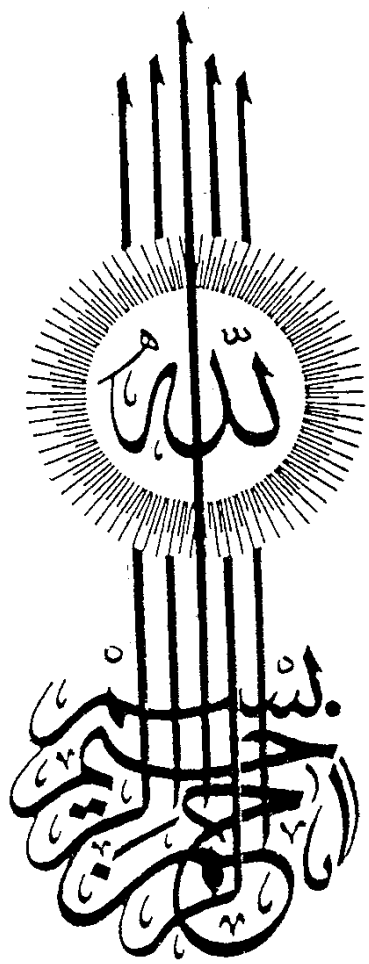

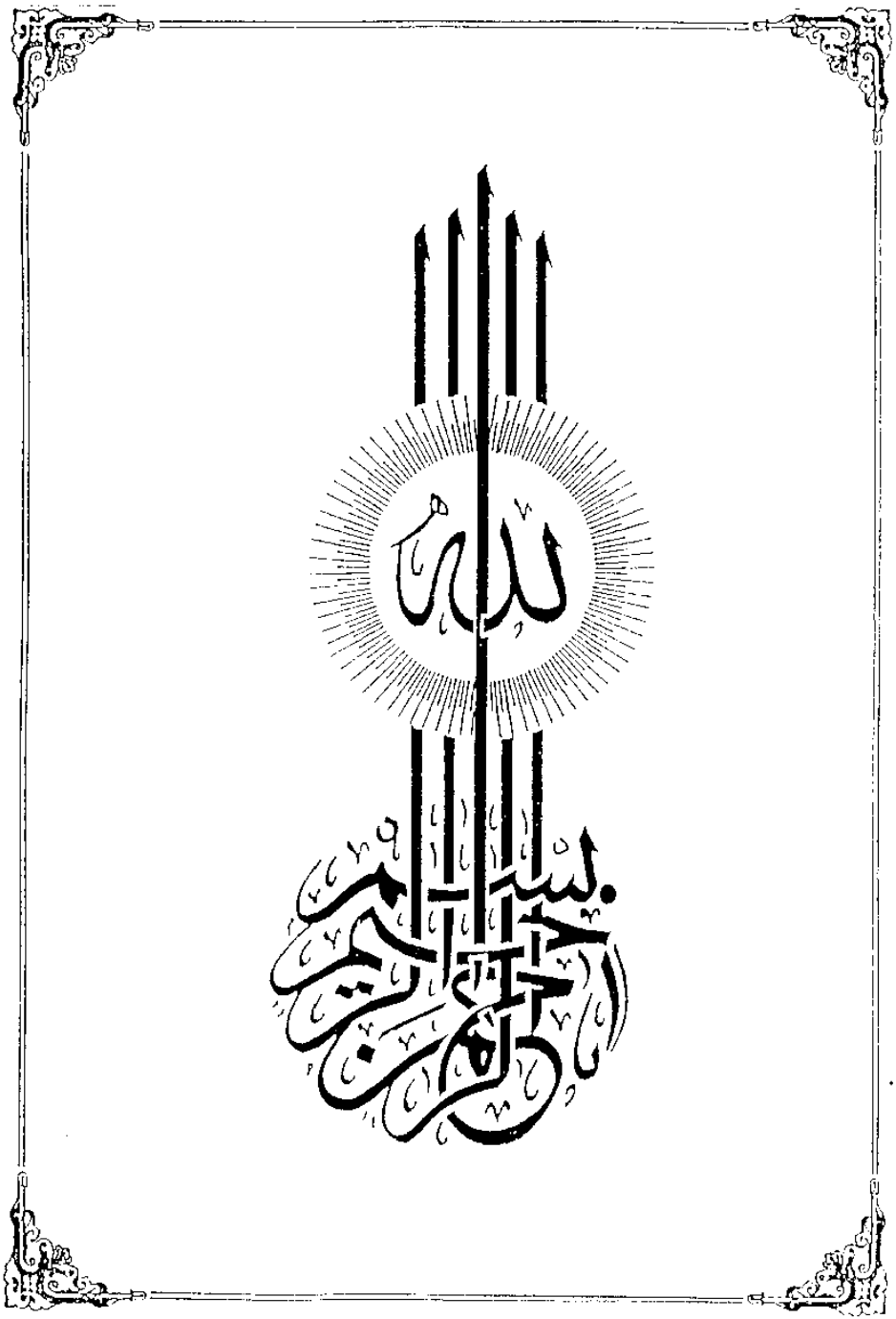

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INDEX

INTRODUCTIONA AND

AIM OF WORK

INTRODUCTION

Portal hypertension is a Common and Serious problem in our children. Its complications particularly haematemesis are responsible for many deaths, also splenomegaly, ascites, malabsorption, septicaemia, and hepatic encephalopathy may contribute to the fatal out come of these children.

Certain types of portal hypertension such as prehepatic and post-Sinusoidal intra-hepatic venous obstruction are predominantly seen in children, also other types can occur.

Liver function tests in these children are commonly fair enough to allow surgical intervension, (Alagille et al., 1979).

This essay aims to study the possible causes of portal hypertension and its bad sequelae in infancy and childhood .Also to define various methods for prevention complications and possible management.

Anatomy and Embryology

HEPATIC CIRCULATION

According to Roberts and Barrowman (1979) the Liver has a double blood supply, a high pressure supply of fully oxygenated blood(mean pressure 90 mm Hg with an Oxygen Content of 19 ml/100 ml blood) from the hepatic artery, and a low pressure supply of less fully-Oxygenated blood (pressure approximately 5-10 mm Hg with an Oxygen content of about 14 ml/100 ml blood), from the valveless portal vein, the two streams hepatic artery and portal vein merge in to Common vessels at the microcirculation . The exchange vessels are permeable, widely dilated Capillary vessels called sinusoids, the pressure in these Sinusoids is at a lower pressure than the mean pressures in most other systemic microcirculation (Brayer, 1963), blood from these sinusoids drains into short and valveless hepatic veins and enters the inferior vena cava.

The Portal System of Veins:

The portal system includes all the veins which drain the blood from the digestive tube (with the exception of the lower part of the anal

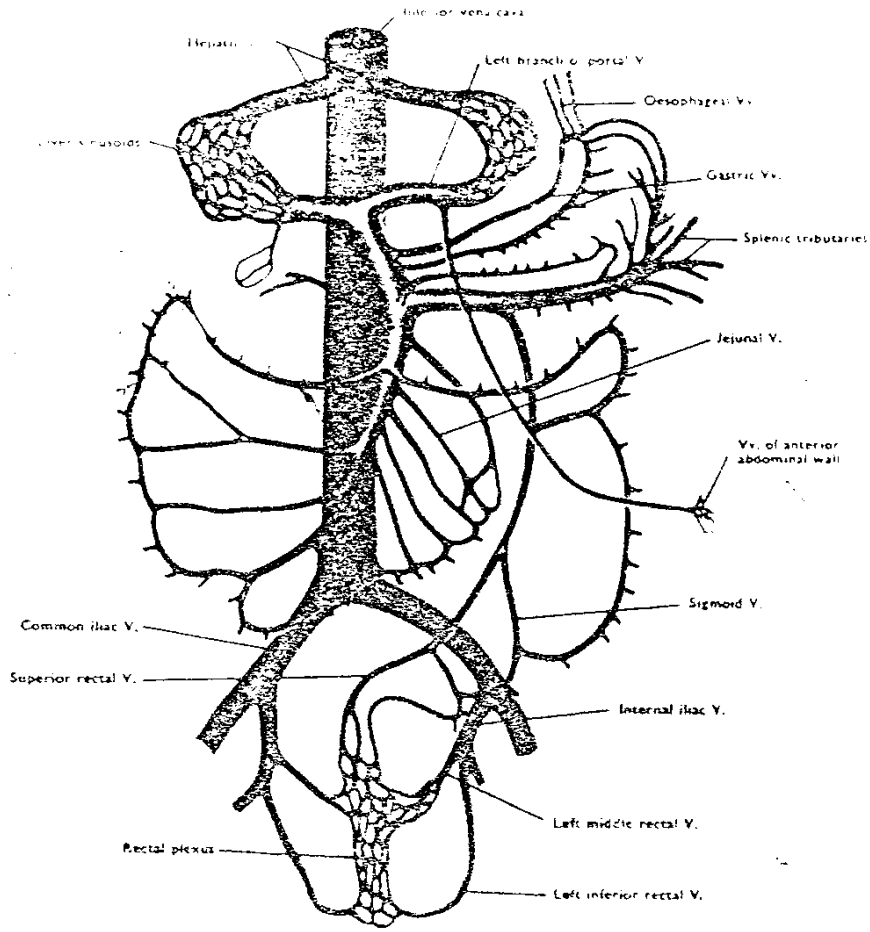

Fig. 1 A diagram of the portal venous system and its anastomosis with systemic venous system. (Chandra etal., 1979)

canal), from the spleen , pancreas and gall bladder, from these viscera the blood is conveyed to the liver by the portal vein (Gray's 1962) . Thus nutrients absorbed from gastro-intestinal tract, as well as hormones (such as insulin and glucagon) released by the pancreas secondary to ingestion of food, are delivered to the liver directly and in high concentration. (Boyer, 1982) .

The Portal Vein :

The Valveless portal vien is an afferent nutrient vessel of the Liver and in this sense is an arterial channel (Rappaport, 1975).

Embryology of the Portal Vein :

The Portal vein derived from the Omphalomesenteric vien which brings blood from the yolk sac and the intestine to the liver, (Rappaport 1975). The Omphalic portion of the vein regresses with the disappearance of the yolk sac.

With growth of the intestine, the mesenteric portion persists and form tributaries of the portal vein(Rappaport 1975).

Anatomy of the Portal Vein :

The portal vein is composed of two portions .

.-A- Extrahepatic portion of the portal vein .

.-B- Intrahepatic portion of the portal Vein .

-A- Extrahepatic portion of the portal vein

The portal vein is formed at the level of the second Lumbar vertebra by the junction of the superior mesenteric and splenic veins, the union of these veins takes place in front of the inferior vena cava and behind the neck of the pancreas (Douglass et al., 1950).

The portal vein extends slightly to the right of the middle line for a distance of S.S- 8 cm to the porta hepatis (Sherlock 1981). It runs in the hepato-duodenal ligament in a plane dorsal to the bile duct and hepatic artery and divides in to two lobar branches before entering the portal fissure (Rappaport 1975).

The right lobar branch is short and thick and receives the cystic vein, the left lobar branch is longer and thin and is joined in front by the paraumblical veins and by a fibrous cord named ligamentum

teres which represents the obliterated left umbilical vein . It is connected to the inferior vena cava by a second fibrous cord termed the ligamentum venosum. The left branch gives branches to the quadrate and caudate lobes before entering the liver at the left end of the porta hepatis (Gray's 1962).

Tributaries of the extrahepatic portion of the portal vein:

1- Splenic vein
2- Superior mesenteric vein .
3- Left gastric vein .
4- Right gastric vein .
5- Paraumblical vein .
6- cystic vein .

-B- Intrahepatic portion of the portal vein .

The liver can be divided into segments, according to the distribution of the major or efferent vascular and biliary branches, . These branches have been named by Bilbey, (1960) after the segments of the liver they supply. Each segment depends on its major vessel for supply. There is no anastomosis between macroscopic

branches, but larger intercommunications at the level of sinusoids (Rappaport 1975) .

The Hepatic Sinusoids :

The hepatic sinusoid has no smooth muscle Coat, but is distinguished from a conventional capillary by several features. It is generally of greater calibre and is lined by two types of cells , an endothelial cell and the phagocytic Kupffer cell. Numerous gaps appear in the endothelial wall when viewed by Transmission electron microscopy (Wiss, 1970). It seems likely that these mainly represent open fenestrae of various sizes in the endothelial cells. The endothelial cell has numerous pinocytotic vesicls. The phagocytic kupffer cells are often situated at bifurcation of sinusoids with pseudopodia actually bridging the vessel. These act as an efficient filter of any bacteria that may gain enterance to the portal venous blood in the intestine. The Kupffer cells as they stretch across the sinusoids, will impose a resistance to flow, the extend of which has not been measured (Jones and Schmucker , 1977).

Anomalies of the Portal Vein :

-(1) Aplasia or Hypoplasia of the Portal vein .

It has been reported by a number of investigators (Hasia and Gellis,(1955) , Marks 1973).

The hypoplasia or atresia may involve the entire length of the vessel or be limited to the point of entrance in to the liver or may occur just proximal to the division in to two branches .

(2) Cavernous transformation of the portal vein .

Is a condition in which the vein is replaced by a spongy trabeculated venous lake with extension in to gastroduodenal ligament .

It has been reported to be one of the causes of portal hypertension in children (Myer and Robinson 1973).

Haemorrhage is the most common manifestation but some children present with asymptomatic splenomegaly (Berdon et al, 1975) . Pancytopenia of varying degree occurs in the majority of the cases (Voorhees et al, 1965).