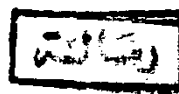


CONGENITAL ANOMALIES OF BILIARY TRACT

E S S A Y

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INTRODUCTION AND AIM OF THE WORK

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- Congenital anomalies of biliary tree are a real challenge for paediatrician and surgeon as well.
- Congenital anomalies of biliary tree are well known it may manifest itself early in life as in cases of congenital biliary atresia.
- The delay in the discovery of this serious congenital anomalies may endanger the patient-life.
- In spite of the fact that infection and haemolytic jaundice play a primary role in the aetiology of neonatal jaundice in our country. Yet early surgical intervention can be of great value in dealing with these cases of congenital obstructive jaundice. Thanks to the recent lines of investigations, early detection of these anomalies can be achieved in the majority of patients, but still some cases passed undiagnosed except late.
- This through a light on the value of surgical exploration as a diagnostic procedure in the cases of doubtful diagnosis.
- The aim of this work is to study the congenital anomalies of biliary tree and to evaluate the recent methods used in its diagnosis and management.

EMBROLOGY & ANATOMY OF THE LIVER

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The liver, the largest gland in the body, lies under the diaphragm mainly on the right side, occupying the right hypochondrial & epigastric regions of the abdomen & extending into the left hypochondrium, sometimes as far as the left midclavicular line... in the newborn infant the liver edge is usually less than 2 cm., below the costal margin in the right midclavicular line. The lower border may be normally palpable about 1 cm. below the costal margin throughout childhood.

During the 1st year of life the liver is relatively large constituting 1/18 of the body weight while in adults it weights one forteith of the body weight.

EMBRYOLOGY OF THE LIVER:

The primordium of the liver is a ventral out growth from the cranial tip of the duodenum at an angle between the foregut & the yolk sac (begining of the fourth week / 17 somites). This diverticulum divides into two portions, a cranial bud which gives rise to the hepatic parenchyma & bile ducts, & a caudal portion from which the gall bladder & cystic duct are derived, these are the endodermal components of the gland.

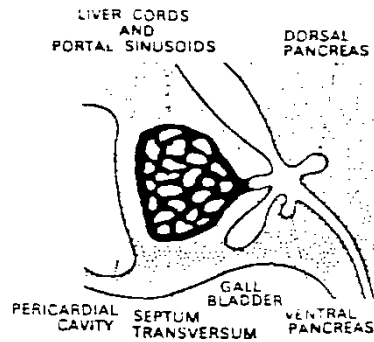
The hepatic diverticulum grows into the septur

transversum at a time when the mesenchymal cells of the latter are engaged in vascularizing activity.

This vascular mesenchyme is invaded by cords of endodermal hepatic cells with the result that the latter are intimately supplied with a system of blood vessels characteristic of the liver. These vessels comprise the hepatic artery, hepatic vein & the portal vein, the latter 2. Constitute a unique dual venous supply found in no other organ. There is also a system of dilated capillaries (hepatic sinusoids) lined with reticuloendothelial (Kupffer's) cells.

The hepatic cords are arranged in a spongework with the sinusoids closely applied to the individual hepatic cells.

This makes possible continuous flow of venous blood from the proximal portion of the portal system into the distal hepatic portion of the sinusoids circulating around the hepatic parenchyma before the blood (loaded with food) enters the heart via the hepatic vein, the lymphatics are also derived from the vascular mesenchyme of the septum transversum. The liver is divided into structural units, the hepatic lobules. The lobules increase in number as function of sinusoids, when the hepatic cords elongate beyond the functional limit of the sinusoids between (supporting)



The human liver at 5 mm., seen in lateral sectional view. (After Arey).

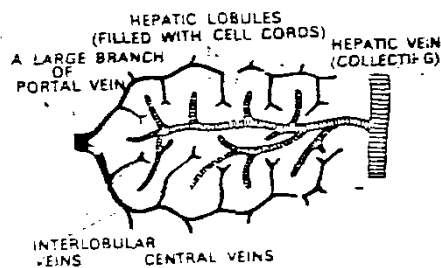
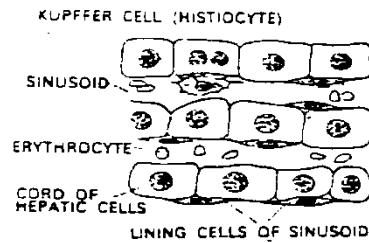


Diagram to show the arrangement of the branches of the portal vein (black) and hepatic vein (blank) and how they encircle the hepatic cords in a lobule. (Modified from Bloom.).

the cords, the central vein divides & the resulting two branches sinusoids assume the place of a new central vein, supporting the two daughter hepatic lobules & so on, the hepatic lobules are separated by portal canals each of which contains derivatives of the hepatic artery, portal vein & bile duct, the hepatic cells begin to secrete typical bile in the fifth month, the bile leaves the hepatic cells via a system of tiny channels or canaliculi which permeate between the individual cells & reach the lumen of interlobular bile ducts, the latter eventually empty into a bile duct from which the bile flows into the gall bladder where it is stored. Thus the hepatic cells have an extensive exposure to both hepatic sinusoids & bile canaliculi, while these developments are taking place, the mesenchymal cells of the septum not involved in the vascularization process, give rise to a capsule & form a fibrous stroma. The connective tissue elements of the liver are, therefore of splanchnopleuric mesodermal origin. The liver is a haemopoietic organ for the foetal period. The blood cells are actively differentiated between the hepatic cords & the sinusoidal lining, later some of these cells become transformed into the cells of Kupffer, which form a part of the reticulo-endothelial system & are noted for their phagocytic properties.



Drawing of a section to show the arrangement of the hepatic cords and the sinusoids. The cells of these cords constitute the parenchyma of liver, which occupies the blank spaces between the incoming portal vein and outgoing hepatic vein in Fig. (2).

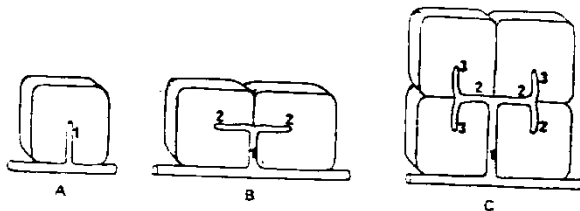


Diagram illustrating the mechanism by which the number of hepatic lobules is increased.

ANATOMY:

In official anatomical terminology, the liver has two main surfaces, diaphragmatic & visceral, the first is subdivided into four parts, commonly also called surfaces & named, anterior, superior, posterior right the visceral surface is sometimes called inferior, the anterior & visceral surface meet at an acute angle like the sharp edge of a wedge to form the lower border. The junctions where other surfaces meet each other are rounded & indistinct, & the demarcation between the posterior and the inferior surfaces which are those of greatest interest in connection with the extrahepatic biliary apparatus, is particularly ill-defined.

Broadly, the bare area is a part of the posterior surface, while the porta-hepatis & the gall bladder bed belong to the visceral surface.

In the embalmed cadaver the hardened liver shows impression which are formed by structures that abut against it but which are not obvious in the living organ.

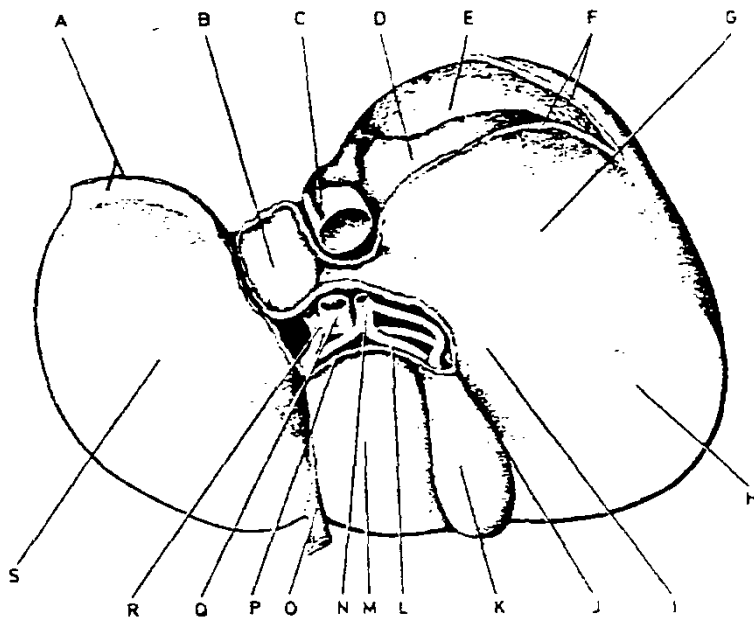
The superior, anterior, posterior & right surfaces can be considered as one curved area related to the diaphragm, with a small part lying in the infracostal

angle & so covered by the upper part of the anterior abdominal wall. Liver biopsies are obtained via the right surface, which extends from the seventh to eleventh rib & is related in its lower third to ribs & diaphragm, in its middle third to ribs diaphragm & pleura & in its upper third to ribs, diaphragm, pleura & lung, the posterior surface contains the bare area & its associated ligaments, the fossa or groove for the inferior vena cava, the caudate lobe & process, the fessure for the ligamentum venosum & impression for the oesophogus & part of the stomach.

The visceral or inferior surface contains the portahepatis, impression for right kidney & suprarenal gland, descending (second) part of the duodenum & right colic flexure, the gall bladder, the quadrate lobe, the fissure for the ligamentum teres & a gastric impression.

On the basis of blood supply & biliary drainage the liver can be divided into a number of segments. So far four segments are recognized in official anatomical nomenclature, left lateral & left medial, right anterior & right pasterior.

Although more elaborate subdivision have been proposed with upper & lower parts to each segment (Healey & Se.roy, 1953) the left lateral segment



The posterior and the inferior (visceral) surfaces of the liver, indicating peritoneal reflections and the impressions due to adjacent viscera. A = Left triangular ligament; B = Caudate lobe; C = Inferior vena cava; D = suprarenal impression; E = bare area; F = superior and inferior layers of coronary ligament; G = renal impression; H = Colic impression; I = Duodenal impression; J = Cystic duct; K = Gall bladder; L = right hepatic duct; M = Quadrate lobe; N = Common hepatic duct; O = Ligamentum teres; P = Part of lesser omentum in porta hepatis; Q = Portal vein; R = Hepatic artery; S = Gastric impression.

After Lord Smith of Marlow