CYSTIC LIVER DISEASES

Essay

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(General Surgery)

By

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LIST OF ABREVIATIONS

AD-PLD	Autosomal Dominant Polycystic Liver Disease
AD-PKD	Autosomal Dominant Polycystic Kidney Disease
CFT	Complement Fixation Test
CPK	Creatine Phosphokinase
CT	Computed Tomography
ECOG	Eastern Cooperative Oncology Group
ELISA	Enzyme Llinked Immunosorbent Assay
ERCP	Endoscopic Retrograde Cholangiopancreatography
HIDA	Hydroxyiminodiacetic-acid
IEP	Immunoelectrophoresis
IVC	Inferior Vena Cava
MRI	Magnetic Resonance Imaging
PLD	polycystic Liver Disease
US	Ultrasound

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AIM OF THE ESSAY

The aim of the essay is to shed some light on modern trends in the management of cystic liver diseases.

INTRODUCTION

Cystic diseases of the liver include simple cysts, multiple cysts arising in the setting of polycystic liver disease, parasitic or hydatid (echinococcal) cysts, cystic tumors, and abscesses ***(Parks and Oniscu, 2007).***

The precise frequency of liver cysts is not known because most do not cause symptoms, but liver cysts have been estimated to occur 5% of the population. No more than 10-15% of these patients have symptoms that bring the cyst to clinical attention ***(Everson et al., 2004).***

The cause of simple liver cysts is not known, but they are believed to be congenital in origin. Adult polycystic liver disease (AD-PCLD) is congenital and is usually associated with autosomal dominant polycystic kidney disease (AD-PKD) ***(Juran and Lazaridis, 2006).*** Liver tumors with central necrosis visualized on imaging studies are often misdiagnosed as liver cysts. True intrahepatic neoplasic cysts are rare ***(Thomas et al., 2005).*** Hydatid cysts are caused by infestation with the parasite Echinococcus granulosus ***(Diker et al., 2007)***. Hepatic abscesses can be amebic or bacterial in origin ***(Parks and Oniscu, 2007).***

Hepatic cysts are usually asymptomatic and found as an incidental finding on imaging or at the time of laparotomy. Pain, fever, and symptoms and signs of complication as portal hypertension, liver cell failure, rupture,

infection and hemorrhage in cysts are the main clinical presentation that varies according to the type of cyst ***(Parks and Oniscu, 2007).***

The evaluation of a patient with cystic liver disease involves laboratory work up as liver function test, complete blood picture, Casoni's test, and tumor markers. A number of options are available for imaging and highly sensitive. Computed tomography scan is also highly sensitive and is easier to interpret, particularly for treatment planning. MRI, nuclear medicine scanning, and hepatic angiography have a limited role in the evaluation of hepatic cysts ***(Power et al., 2007).***

The management of liver cysts is either medical, surgical or both. Each management modality should be tailored according to the type of cyst, presence of complications, and the clinical condition of the patient ***(Parks and Oniscu, 2007).***

Embryology of the liver

The liver arises in the middle of 3rd week of intrauterine life as a diverticulum from the ventral surface of the duodenal foregut close to its junction with the midgut where the latter is continuous with the yolk sac ***(Williams et al., 2002).***

Development of glandular tissue:

The hepatic diverticulum is lined with endoderm, grows ventrally and cranially into the septum transversum, its tip diverges into 2 soild hepatic buds of cells; the future right and left hepatic lobes of the liver ***(Williams et al., 2002).***

Development of Sinusoids:

The hepatic buds develop into epithelial sheets which branch and anastomose to from a close meshwork. The intervals of the meshwork become filled with blood sinusoids, on section, the organ has the appearance of a vascular sponge. These vessels arise in situ possibly under the effect of 2 factors; the influence of endodermal cells of the liver on the potentially angiogenic cells of the mesenchyme of the septum transversum and the dilatation of the pre-existing septal capillaries which are plexiform tributaries of the transseptal vitello-umbilical veins ***(Williams et al., 2002).***

The development of stroma and haemopoietic tissue:

By the continued growth of the liver, its connective tissue stroma and phagocytic sinusoid-lining cells are derived from the included mesenchmal

cells of the septum transversum. Haemopoiesis which begins during the 6th week of intrauterine life is responsible for the large size of liver between the 7th and 9th week of development (10% of total weight of the foetus) ***(Moore, 1999).***

The development of biliary system:

The formation of intrahepatic ducts is dependant upon contact between the developing embryonic liver mass and a preformed extrahepatic duct system. The original diverticulum from the duodenum forms the bile duct, and from its distal part, the cystic duct and the gall bladder arise as an outgrowth, solid at first but later on, become canalized ***(Williams et al., 2002).***

The bile duct opens into the ventral wall of the duodenum, later on, after gut rotation, it migrates to the left across the dorsal surface of the duodenum to the position which it occupies in the adults on the medial border, this occurs at the 7th week. The bile formation begins during the 12th week, but the bile enters the duodenum after the 13th week giving the meconium its green color ***(Moore, 1999).***

The development of hepatic ligaments:

As the liver enlarges, it projects into the abdominal cavity from the caudal surface of the septum transversum. During this process, mesenchyme of the septum is drawn out, and cavitated craniodorasally to form coronary; right, left triangular ligaments and the lesser omentum ***(Williams et al., 2002).***

The hepatic lobulation:

At the 3rd month, the right and left lobes are nearly equal, later on, the relative development of left lobe is less active, which undergoes some degeneration and become smaller than the right. Toward the end of the fetal period, and mostly after birth, these primary lobules of liver are subdivided into smaller secondary units through the connective tissue invasion at early childhood, there is clear demarcation of about 500.000 lobules ***(Williams et al., 2002).***

Anatomy of the Liver

The liver is situated at the right hypochondrium and the epigastrium and extending into the left hypochondrium as far as the left lateral line, wedge shaped with the base to the right and the apex to the left ***(Ralph, 1998).***

It is the largest gland in the body, weighing from 1400 to 1800 gm in man and 1200 to 1400 in woman ***(Lindner, 2000).***

The liver has 2 main aspects **(Fig. 1),** diaphragmatic, which is related to the diaphragm and visceral aspect which is related to the abdominal viscera the diaphragmatic aspect is divisible into 4 surface, superior, which is related to the right copula and left lung. At the right lobe, it reaches to the 4^{th} right intercostals space. At the left lobe, it reaches to the 5^{th} left intercostals space (both levels on full expiration).

The anterior surface is related mainly to diaphragm, costal cartilages, partially to the anterior abdominal wall. The right (lateral) surface is related to the right lateral undersurface of the diphragm from the 7^{th} to the 11^{th} ribs.The posterior surface is related to the inferior vena cava and oesophagus. The inferior surface is nearly the visceral aspect and related to the pylorus, the duodenum, gall bladder, right colon, hepatic flexure, right

third of the transverse colon, right kidney and the right surprarenal gland ***(Williams et al., 2002).***

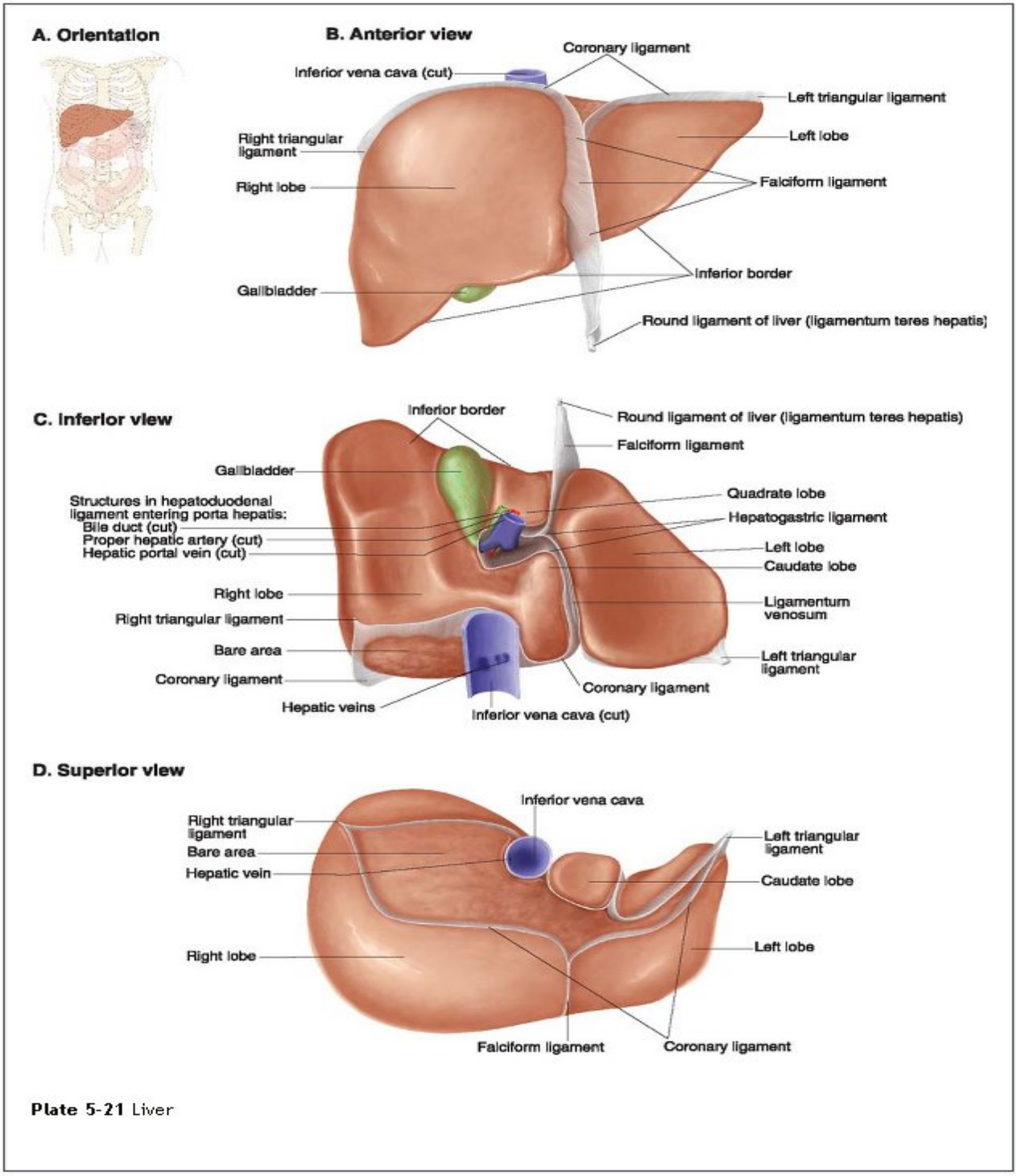

Fig. 1: Anatomy of the liver (Lippincott and Wilkins, 2008).