

## Review Article

# Surgical significance of variations in anatomy in the biliary region

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### ABSTRACT

Variations in the anatomy of the gallbladder, the bile ducts, and the arteries that supply them and the liver are important to the surgeon, because failure to recognize them can cause iatrogenic injury to the biliary tract. A surgeon should be always be careful while operating in this area. In addition these anomalies are associated with a range of other congenital anomalies, including biliary atresia and cardiovascular or other gastrointestinal malformations, biliary lithiasis, choledochal cyst, anomalous pancreaticobiliary junction etc, so a look out for other anomalies should be carried out simultaneously.

**Keywords:** Gall bladder, Cystic fossa, Calots triangle, Monyhans hump, Extrahepatic, Atresia, Double Gall bladder, Triple Gall bladder, Agenesis, Cystic duct, Carolis disease

### NORMAL ANATOMY

The gallbladder is a flask-shaped, blind-ending diverticulum attached to the common bile duct by the cystic duct. It usually lies attached to the inferior surface of the right lobe of the liver. In the adult the gallbladder is between 7 and 10 cm long with a capacity of 30-50 ml. It usually lies in the Cystic fossa. The gallbladder is described as having a fundus, body and neck. The neck lies at the medial end close to the porta hepatis, and almost always has a mesentery; this mesentery usually contains the cystic artery. The mucosa at the medial end of the neck is obliquely ridged, forming a spiral groove continuous with the spiral valve of the cystic duct. At its lateral end the neck widens out to form the body of the gallbladder and this widening is often referred to as 'Hartmann's pouch'.

The cystic duct drains the gallbladder into the common bile duct. It is between 3 and 4 cm long, passes posteriorly to

the left from the neck of the gallbladder, and joins the common hepatic duct to form the common bile duct. The mucosa of the cystic duct bears 5-12 crescentic folds, continuous with those in the neck of the gallbladder.

**Hepatic bile ducts:** The main right and left hepatic ducts emerge from the liver and unite near the right end of the porta hepatis as the common hepatic duct. This descends about 3 cm before being joined on its right at an acute angle by the cystic duct to form the common bile duct. The common hepatic duct lies to the right of the hepatic artery and anterior to the portal vein in the free edge of the lesser omentum.

**Common bile duct:** The common bile duct is formed near the porta hepatis, by the junction of the cystic and common hepatic ducts. It is usually between 6 and 8 cm long. It descends posteriorly and slightly to the left, anterior to the epiploic foramen, in the right border of the lesser omentum.

Hepatopancreatic ampulla (of Vater) as it lies medial to the second part of the duodenum; the common bile duct approaches the right end of the pancreatic duct. The ducts enter the duodenal wall together, and usually unite to form the hepatopancreatic ampulla.

## ANOMALIES

**Absence of the gallbladder** is rare and apparently has a genetic predisposition, because several family members may be affected. Some are born with other congenital anomalies, including biliary atresia and cardiovascular or other gastrointestinal malformations. They often die in infancy. Others are asymptomatic throughout life. Slightly more than half have symptoms suggestive of biliary tract disease; in these patients, dilatation of the bile duct system or choledocholithiasis is a frequent finding. Agenesis of the gallbladder is rarely diagnosed preoperatively in patients with symptoms of biliary tract disease, because the sonogram is interpreted as showing a small, contracted gallbladder, and oral cholecystography and scintigraphy show nonvisualization of the gallbladder, all of which suggest the presence of gallbladder disease. A thorough search for the gallbladder in possible ectopic locations should be made during operation. If none is found, an operative cholangiogram is indicated.

**An ectopic gallbladder** is also rare finding for surgeons. An intrahepatic location on the right side makes cholecystectomy hazardous, and a cholecystostomy with removal of the stones is the preferred treatment of cholelithiasis in patients with this anomaly. A Left-sided gallbladder is located to the left of the falciform ligament and is usually partially embedded in the substance of the left lobe of the liver. The cystic duct may drain into the left hepatic duct or the common hepatic duct. In retro displacement of the gallbladder, the organ is located beneath the posterior and inferior surfaces of the liver.

The Gallbladder may have abnormal positions: Intrahepatic, rudimentary, have anomalous forms, or be duplicated. Isolated congenital absence of the gallbladder is very rare, with a reported incidence of 0.03%. Before the diagnosis is made, the presence of an intrahepatic bladder or anomalous position must be ruled out.

The gallbladder may be almost completely buried within the liver surface, having no peritoneal covering intraparenchymal pattern or it may hang from a short mesentery formed by the two layers of peritoneum separated only by connective tissue and a few small vessels (mesenteric pattern).

**Intrahepatic gallbladder** is one of the ectopic locations of the gallbladder. The gallbladder is usually intrahepatic during its embryologic period and become gallbladder, Cystic Fossa, Calots Triangle, Monyhans Hump, Extrahepatic, Atresia, Double Gall Bladder, Triple Gall Bladder, Agenesis, Cystic Duct, Carolis Disease extra

hepatic later on in its development. In adults approximately 60% of intrahepatic gallbladders are associated with gallstones. Intrahepatic gallbladders can be readily imaged by either ultrasonography or CT scan.<sup>1-4</sup>

Intrahepatic gallbladders have a subcapsular location along the anterior inferior right lobe of the liver, occasionally a small portion of the intrahepatic gallbladder may protrude from the liver. An intrahepatic gallbladder results from a developmental anomaly or a congenital arrest which prevents the gallbladder from moving from its intrahepatic position in the 2nd month of gestation to its normal superficial location. The condition may be suspected, if the cholecystogram or USG reveals a gallbladder in an unusually high location.

An intrahepatic gallbladder has usually impaired function because it does not empty completely. This may result in gallstone formation due to stasis. Most intrahepatic gallbladders are only partially embedded within the hepatic parenchyma and then can usually be easily identified at the time of cholecystectomy. Those that are completely buried within the liver may be a challenge for the general surgeon.

A completely embedded gallbladder is best approached by first identifying the cystic duct where it joins the common hepatic duct and then following the cystic duct back to the gallbladder.<sup>5-8</sup>

Torsion may occur in a floating gallbladder, in which the organ is completely peritonealized and free from the liver or attached to the liver by a long mesentery.

**Double and Triple gallbladders** have been reported, the latter being extremely rare. Double gallbladders may share a common cystic duct and be completely separated, or they may be divided by a septum. When they do not share a common outlet, the cystic ducts of double or triple gallbladders open separately into the common bile duct or, less commonly, into the right hepatic duct.

TriPLICATION of the gallbladder is a rare congenital anomaly of the biliary tract; there are only nine reported cases to date.

**Duplication of the gallbladder** with two separate cavities and two separate cystic ducts has an incidence of about one in every 4000 persons. Duplication of the gallbladder is a rare anomaly of the biliary tract, occurring at a rate of 0.25/1,000 in one autopsy series. Several classifications have been proposed according to anatomic or embryologic development of the gallbladder. Boyden classified double gallbladder in bilobed gallbladder and in true duplicated gallbladder, with two types according to the connection of the cystic ducts. Gross classified double gallbladder into six types, designated A to F, and Harlaftis et al. classified double gallbladder into two groups.<sup>9</sup>

This condition tends to lead to biliary complications, such as cholelithiasis and acute cholecystitis of both gallbladders. The clinical features are usually Right Upper Quadrant pain and tenderness and sometimes jaundice

**Extrahepatic biliary atresia** is the leading cause of obstructive jaundice during infancy, whereas anomalies of the extrahepatic biliary tree rarely cause symptoms during adulthood.<sup>10-13</sup>

Atresia is present in one per 14000 live births, and affects male and females equally. The extrahepatic bile ducts are progressively destroyed by an inflammatory process which starts around the time of birth. Intrahepatic changes also occur and eventually result in biliary cirrhosis and portal hypertension. The untreated child dies before the age of 3 years of liver failure or haemorrhage.

The inflammatory destruction of the bile ducts has been classified into three main types:

- Type I - atresia restricted to the common bile duct;
- Type II - atresia of the common hepatic duct;
- Type III - atresia of the right and left hepatic ducts

Basically considered as a disease of “C”

- Congenital, autosomal recessive disorder.
- Communicating cavernous ectasia of intrahepatic biliary ducts.
- Cystic structures (Multiple) converging towards porta hepatis
- Central dot sign on CT Scan
- Cholangiography most diagnostic
- Congenital hepatic fibrosis and cystic (Poly cystic) kidney disease are associations

#### **Anomalies of Cystic duct**

The cystic duct may be long, and increases the need for its complete dissection. The duct may pass behind the common hepatic duct to enter on its posterior wall or on its left lateral aspect. In chronic cholecystitis, the gallbladder may be small and shrunken, and the cystic duct may be absent or extremely short; in this circumstance, the common bile duct may easily be mistaken for the cystic duct as dissection proceeds from the gallbladder fundus toward the cystic duct. A very long cystic duct may enter the common bile duct a variable distance from the sphincter of Oddi and may be fused with the common duct, in which case the two ducts should not be separated because they share a common wall. Finally, an accessory duct from the liver may enter the cystic duct or the common hepatic duct.

The cystic duct may have several important variations in its anatomy. Rarely, the cystic duct lies along the right edge of the lesser omentum to the level of the duodenum

before the junction is formed, but in these cases the cystic and common bile ducts are usually closely adherent. The cystic duct occasionally drains into the right hepatic duct in which case it may be elongated, lying anterior or posterior to the common hepatic duct, and joins the right hepatic duct on its left border. Rarely, the duct is double or even absent in which case the gallbladder drains directly into the common bile duct.

One or more accessory hepatic ducts occasionally emerge from segment V of the liver and join either the right hepatic duct, the common hepatic duct, the common bile duct, the cystic duct, or the gallbladder directly.

These variations in cystic duct anatomy are of considerable importance during surgical excision of the gallbladder. Ligation or clip occlusion of the cystic duct must be performed at an adequate distance from the common bile duct to prevent angulation or damage to it. Accessory ducts must not be confused with the right hepatic or common hepatic ducts.

**Double common bile duct (DCBD)** is a rare congenital anomaly in which two common bile ducts exist. One usually has normal drainage into the major duodenal papilla and the other usually named accessory common bile duct (ACBD) opens in different parts of upper gastrointestinal tract (stomach, duodenum, ductus pancreaticus or septum). This anomaly is of great importance since it is often associated with biliary lithiasis, choledochal cyst, anomalous pancreaticobiliary junction and upper gastrointestinal tract malignancies.<sup>14-17</sup>

Variations in the anatomy of the hepatic and cystic arteries that are of surgical significance are important for a surgeon. The cystic artery that supplies the gallbladder is usually a branch of the right hepatic artery (>90% of the time). The course of the cystic artery may vary, but it nearly always is found within the hepatocystic triangle, the area bound by the cystic duct, common hepatic duct, and the liver margin “Triangle of Calot”. When the cystic artery reaches the neck of the gallbladder, it divides into anterior and posterior divisions.

In about 5% of cases, there are two right hepatic arteries, one from the common hepatic artery and the other from the superior mesenteric artery. In about 20% of patients, the right hepatic artery comes off the superior mesenteric artery. The right hepatic artery may course anterior to the common duct. The right hepatic artery may be vulnerable during surgical procedures, in particular when it runs parallel to the cystic duct or in the mesentery of the gallbladder. The cystic artery arises from the right hepatic artery in about 90% of cases, but may arise from the left hepatic, common hepatic, gastroduodenal, or superior mesenteric arteries.

A bend in the course of the right hepatic artery, throwing it into the configuration of a caterpillar hump or Moniyhans Hump invites injury unless it is carefully

dissected free. A very short cystic artery also puts the hepatic artery at risk. Occasionally, the right hepatic artery courses anterior to the common bile duct.

**Choledochal Cysts:** Choledochal cysts are congenital cystic dilatations of the extrahepatic and/or intrahepatic biliary tree. They are rare, The incidence is between 1:100,000 and 1:150,000 in populations of Western countries- but are more commonly seen in populations of Eastern countries. Choledochal cysts affect females three to eight times more often than males. Although frequently diagnosed in infancy or childhood, as many as one half of the patients have reached adulthood when diagnosed.

The cause is unknown. Cysts of the biliary duct system are uncommon. About 80% are diagnosed during childhood, and the remainder become apparent in adulthood. Originally, they were described as cystic dilatations of the extrahepatic duct system. Subsequently this classification was extended to include the frequent association with cystic dilatation of the duct system within the liver, a condition described in 1958 and now known as Caroli's disease.

The etiology is uncertain, but some evidence suggests that fusiform dilatation results from distal obstruction and destruction of the proximal bile duct epithelium by pancreatic juice, because the pancreatic duct enters proximal to the ampulla in most of these patients, and distal obstruction and damage to the epithelium cause cysts in puppies. Classification of choledochal cysts.

- Type I, fusiform or cystic dilations of the extrahepatic biliary tree, is the most common type, making up >50% of the choledochal cysts.
- Type II, saccular diverticulum of an extrahepatic bile duct. Rare, <5% of choledochal cysts.
- Type III, bile duct dilatation within the duodenal wall (cholechoceles), makes up about 5% of choledochal cysts.
- Type IVa and IVb, multiple cysts, make up 5–10% of choledochal cysts. Type IVa affects both extrahepatic and intrahepatic bile ducts.
- Type IVb cysts affect the extrahepatic bile ducts only. Type V, intrahepatic biliary cysts, is very rare and makes up 1% of choledochal cysts.

The classic symptoms are abdominal pain, jaundice, and an abdominal mass, but not all children have a mass. In both children and adults, cholangitis is frequent, probably because of bile stasis and colonization with bacteria. Most cysts can be detected by ultrasonography or by radionuclide scanning, but the definitive diagnosis requires cholangiography. Choledochal cysts should be treated by complete excision and Roux-en-Y hepaticojejunostomy whenever possible. The long-term results of procedures in which the cyst remains in situ are not good because of recurrent pancreatitis in about one third of patients and the development of carcinoma in the

cyst in about one fourth of them. Recent evidence suggests that excision of the extrahepatic portion of the cyst alone eliminates the risk of subsequent cholangiocarcinoma in patients who have both intrahepatic and extrahepatic cystic disease, perhaps because the reflux of pancreatic juice is prevented.

Extrahepatic anomalies are present in 10-25% of patients and include cardiovascular defects, polysplenia, preduodenal or absent portal vein, malrotation, situs inversus, and bowel atresias.<sup>17,18</sup> A rare fetal type of biliary atresia has also been described in which congenital malformations occur more commonly and cholestatic jaundice is present from birth. A bile duct remnant can not be identified at the time of exploratory laparotomy in patients with this variant of biliary atresia.<sup>19</sup>

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