Anatomy and Embryology of the Biliary Tract

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KEYWORDS

• Gallbladder • Biliary tree • Portal triad • Anatomy • Embryology

KEY POINTS

- Variation in the anatomy of the extrahepatic biliary tree and its associated vasculature should be anticipated. When aberrant anatomy is encountered, other aberrancies should be expected.
- The embryologic development of the extrahepatic biliary tract is complex and incompletely understood; however, several important factors in cell signaling have been defined in recent years.
- Biliary atresia is an uncommon but serious cause of perinatal jaundice and requires operative intervention, usually a Kasai portoenterostomy. Liver transplant is often ultimately required.
- The symptoms of choledochal cysts may be nonspecific, but diagnosis is important in the face of increased risk of cholangiocarcinoma inherent to these patients.
- The replaced right hepatic artery is a common aberrancy of the hepatic vasculature and is found posterolateral in the portal triad. The replaced left hepatic artery can be found in the gastrohepatic ligament.
- Ducts of Luschka, perhaps better termed *subvesical ducts*, are an important cause of postcholecystectomy bile leak, a complication that may be avoided by cautious, shallow dissection of the gallbladder from the fossa.

INTRODUCTION

Working knowledge of extrahepatic biliary anatomy is of paramount importance to the general surgeon. The laparoscopic cholecystectomy is one of the most common surgical procedures in the United States. In surgical training, it is the procedure whereby learners often cut their teeth in the laparoscopic arena, first with the privilege of peeling the gallbladder from its fossa and later by dissecting out the cystic structures. The variation of the anatomy can be staggering. Depending on the disease process, the

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setting of inflammation can significantly impair visualization and distort the usual locations of the regional structures. Congenital malformations are also a source of anatomic variation and can confuse or surprise the surgeon at the time of surgical exploration. Misunderstanding and underestimation of the anatomy can result in misdiagnosis and serious injury to the biliary tree in the operative setting. Although biliary injury is uncommon, its potential complications carry a high morbidity. In this article, the authors review the embryologic development of the extrahepatic biliary tract and gallbladder as well as its variable anatomy.

EMBRYOLOGY

General Biliary Embryology

Understanding of the biliary tract begins with the appreciation of its embryologic development. Beginning in the fourth week of gestation, the liver bud arises from the distal extent of the foregut. As the liver parenchyma develops, the cells between it and the foregut proliferate, forming the precursor to the bile duct.[1] Between the fourth and fifth weeks of gestation, the gallbladder primordium buds off the caudal extent of the bile duct giving rise to the gallbladder and cystic duct. This bud lies in close proximity to the ventral pancreatic bud. The shared stalk rotates posteriorly and medially to join the dorsal pancreatic bud (**Fig. 1**). The ventral pancreatic bud gives rise to the uncinate process; its duct, the duct of Wirsung, typically joins with the common bile duct (CBD). This confluence occurs at the ampulla of Vater, and they drain into the duodenum via the major papilla. Usually, the duct draining the dorsal pancreatic bud will fuse with the duct draining the ventral pancreatic bud. This duct, the duct of Santorini, may fail to fuse (known as *pancreas divisum*) and/or drain directly into the duodenum at the minor papilla.

The extrahepatic biliary tree develops in close concert with the hepatic artery. Further details of the development of the extrahepatic biliary tract remain nebulous. It was initially thought that the biliary tract lumen passed through a phase in which the lumen was obliterated by proliferating endothelial cells, and failure to recanalize resulted in biliary atresia in neonates, similar to the pathogenesis of duodenal atresia. This belief has been refuted by studies in human embryos showing that the lumen never obliterates during maturation.[2] The process of how the intrahepatic and extrahepatic biliary networks anastomose is not well understood, but they seem to be in continuity throughout development.

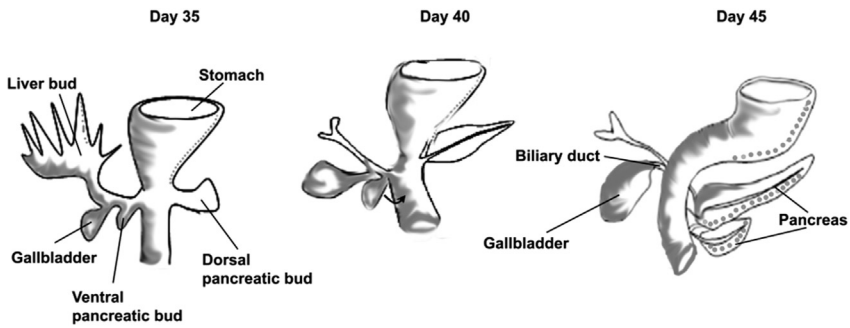

Fig. 1. Embryologic development of the biliary tree and pancreas. (*From* Sahu S, Joglekar MV, Yang SNY, et al. Cell sources for treating diabetes. In: Gholamrezanezhad A, editor. Stem Cells in Clinic and Research, 2011. InTech, http://dx.doi.org/10.5772/24174. Available at: http://www.intechopen.com/books/stem-cells-in-clinic-and-research/cell-sources-for-treating-diabetes.)

Cell Signaling in Biliary Development

The development of the liver and biliary tract is governed by many of the major pathways in cell signaling. These pathways include Notch, Wnt, sonic hedgehog, and transforming growth factor β.[3] Not surprisingly, these pathways have been implicated in the pathogenesis of biliary and pancreatic malignancies.[4–6] Although the development of intrahepatic bile ducts is fairly well understood, there seem to be distinctly different mechanisms regulating the development of the extrahepatic biliary tract. Current literature suggests that the development of the extrahepatic biliary tree is more closely related to the development of the duodenum and the pancreas.[7] Evidence of this includes the expression of Pdx1 in both biliary and pancreatic progenitor cells but not in the liver progenitor cells in the murine model.[8] Of particular importance are the transcription factors hepatic nuclear factor (HNF) 1β,[9] HNF6,[10] Sox17, and Hes1,[8] which when absent predispose to malformation of the extrahepatic biliary tree and gallbladder agenesis. Much of the difficulty in studying the embryologic development of the extrahepatic biliary tract stems from the essential nature of the cell signaling pathways regulating it. When these pathways are disturbed, it is often lethal to the embryo.

CONGENITAL DISORDERS OF THE BILIARY TRACT

Biliary Atresia

The pathogenesis of biliary atresia is a complicated process with multiple factors influencing development. The incidence of biliary atresia varies by region of the world and ranges from 1 in 5000 in Asian countries to 1 in 19,000 in European countries.[11] Around 20% of patients with biliary atresia also suffer from an additional congenital abnormality, including splenic abnormalities (most common), venous malformations, and syndromes driven by chromosomal abnormalities. The factors thought to influence the development of biliary atresia in the prenatal period include genetic dysregulation, immune dysfunction, and inflammation. Patients present with unresolving perinatal jaundice, cholestasis (pale stool and a direct hyperbilirubinemia), and progressive liver dysfunction. In this context, ultrasound showing no dilation of the bile ducts is suggestive of biliary atresia. Liver biopsy is usually necessary for diagnosis. Histologic examination of the bile ducts reveals "ductular reaction, bile plugs within bile ductules, portal tract edema, and portal fibrosis."[12] The natural history of biliary atresia is progression to cirrhosis and death by 2 years of age. The first-line treatment of biliary atresia is usually the Kasai portoenterostomy. The outcome is determined by the quality of biliary drainage. Regardless of drainage, cirrhosis will often progress over time, and liver transplant becomes necessary.

Choledochal Cysts

Choledochal cystic disease is another congenital condition whose pathophysiology remains incompletely understood. The incidence of choledochal cysts varies by region, occurring in about 1 per 1000 in Asia but only 1 per 100,000 to 150,000 in the Western world. Females are affected more often than males. The most commonly accepted theory for pathogenesis is pancreaticobiliary maljunction where the CBD and pancreatic duct share a long common channel. Pancreaticobiliary maljunction leads to the reflux of pancreatic enzymes up into the biliary tree. Subsequent inflammation and dilation occur. However, this theory does not completely account for other characteristics of the disease, including antenatal findings of dilation when pancreatic enzymes are not being produced in significant quantities.[13] The five types of choledochal cysts are depicted in **Fig. 2**. The most common presentation is fusiform dilation of the extrahepatic ducts, sometimes including the cystic duct (type I) representing 80% to 90% of cases.

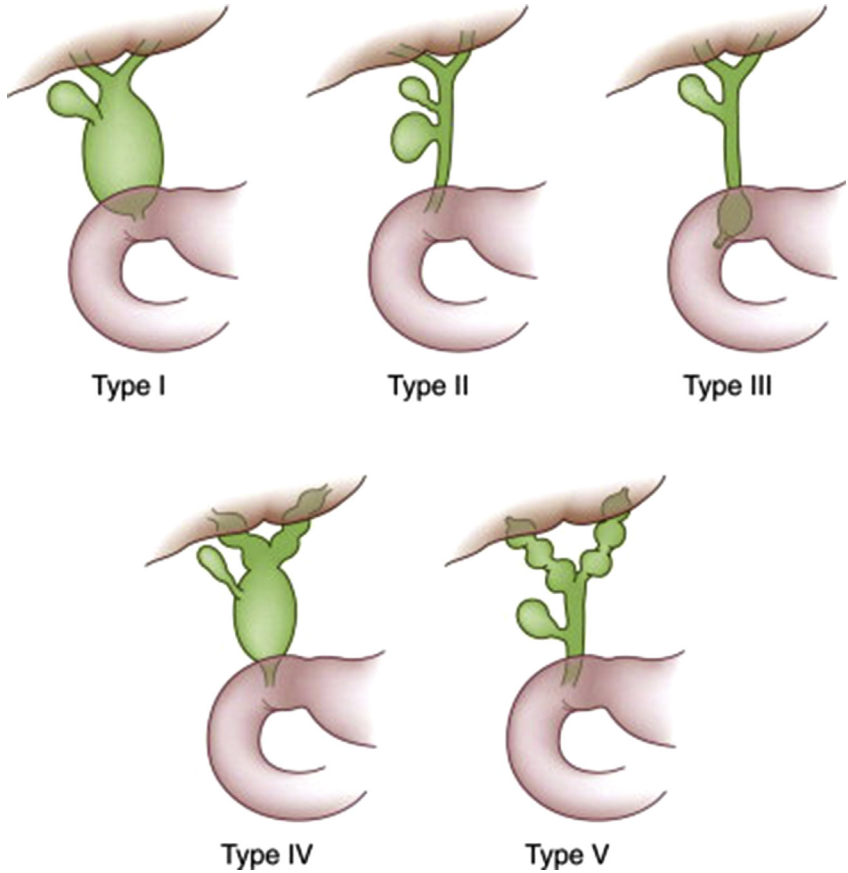

Fig. 2. Todani classification of choledochal cysts. (*From* Sabiston DC, Townsend CM. Sabiston textbook of surgery: the biological basis of modern surgical practice. 19th edition. Philadelphia: Elsevier Saunders; 2012; with permission.)

Presentation is variable. Patients usually present at childhood with symptoms that may classically include right upper quadrant pain, jaundice, and a palpable mass in the right upper quadrant. Patients may develop pancreatitis or cholangitis. Diagnosis is based on imaging, usually with either computed tomography (CT) or ultrasonography. Cholangiography is often necessary for surgical planning. Although the pathogenesis is not well defined, the complications that can result are. These complications include choledocholithiasis, cholecystitis, cholestasis, cirrhosis, and, most seriously, an increased risk of cholangiocarcinoma with age. The treatment of choledochal cystic disease is total cyst resection and biliary drainage, usually with a Roux-en-Y hepaticojejunostomy. Unfortunately, studies suggest that an increased risk for cancer persists despite resection, with up to 5% going on to develop cholangiocarcinoma.[14] Further details of choledochal cystic disease are discussed elsewhere in this issue.

Gallbladder Agenesis

As mentioned earlier, several signaling pathways and transcription factors have been shown to be of critical importance in the development of the gallbladder. It is estimated that congenital absence of the gallbladder occurs in 10 to 65 per 100,000

live births. This approximation may underestimate the true incidence because the malformation may go undetected.[15] Females are more likely to be diagnosed, but autopsy studies show an equal incidence between the sexes. Other congenital malformations or variants may be associated. Typically, there is little consequence of gallbladder agenesis; however, some patients develop symptoms of biliary colic.[16] Diagnosis is suggested by the absence of the gallbladder on right-upper-quadrant ultrasound. Further studies are usually required to confirm this and may include CT, magnetic resonance cholangiopancreatography, or the more invasive endoscopic retrograde cholangiopancreatography (ERCP) or endoscopic ultrasound. Cholescintigraphy in these patients may be misleading because nonvisualization of the cystic duct could lead the surgeon to diagnose cholecystitis, subjecting patients to an unnecessary procedure. Cases were often diagnosed intraoperatively after thorough exploration had failed to reveal the gallbladder. This situation occurs less commonly as a result of advances in imaging technology.[15] If at exploration the gallbladder is not easily visualized, intraoperative ultrasound can be used to avoid more aggressive exploration. Because sphincter of Oddi dysfunction (SOD) has been postulated to be one of the causes for biliary colic in these patients, endoscopic sphincterotomy may be helpful for symptomatic patients. This procedure is only pursued after medical management, such as with smooth muscle relaxants, has failed.

BILIARY ANATOMY

Classic Extrahepatic Biliary Anatomy

Perhaps the most consistent feature of biliary anatomy is its inconsistency. Aberrant anatomy should be expected and sought during any biliary surgery. Classically, the right and left hepatic ducts exit the liver and join to form the common hepatic duct, as seen in **Fig. 3**. The left hepatic duct courses from the base of the umbilical fissure along the inferior border of segment IV of the left lobe before joining the short right hepatic duct just below the infundibulum of the gallbladder. The longer length generally seen with the left hepatic duct allows for a more sufficient target for operative biliary decompression or bypass in cases of obstruction or for reconstruction in the face of malignancy. The close relationship between the confluence of the left and right hepatic ducts with the undersurface of the liver hilum emphasizes the need for extensive hepatic resection seen with hilar cholangiocarcinoma. Also, the short length of the right and left hepatic ducts provides a tumor in this location with ready access to intrahepatic secondary biliary radicals, often preventing curative resection.

The cystic duct may be of variable length and typically joins the common hepatic duct to form the CBD. The CBD courses down the hepatoduodenal ligament anterior to the portal vein and lateral to the hepatic artery. The CBD courses inferiorly, posterior to the first portion of the duodenum, then posterior to the pancreas in a groove, often covered by a thin layer of pancreas.[17] Finally, it enters into the second portion of the duodenum either alone or after joining the pancreatic duct.

The length of the CBD varies between 7 and 11 cm in length and has an internal diameter of up to 8 mm at a normal physiologic pressure.[18] Lining the lumen is a columnar epithelium that contains mucus-secreting cells. The main arteries supplying the CBD course along its lateral and medial walls and originate from the gastroduodenal and right hepatic arteries. This arterial anatomy is clinically relevant in iatrogenic injury of the CBD because compromise of this vascular network can lead to stenosis.

The hepatic artery is intimately associated with the biliary tree, situated medial to the CBD in the hepatoduodenal ligament. In the classic description, the celiac axis branches off of the aorta after it passes through the diaphragm. The celiac axis

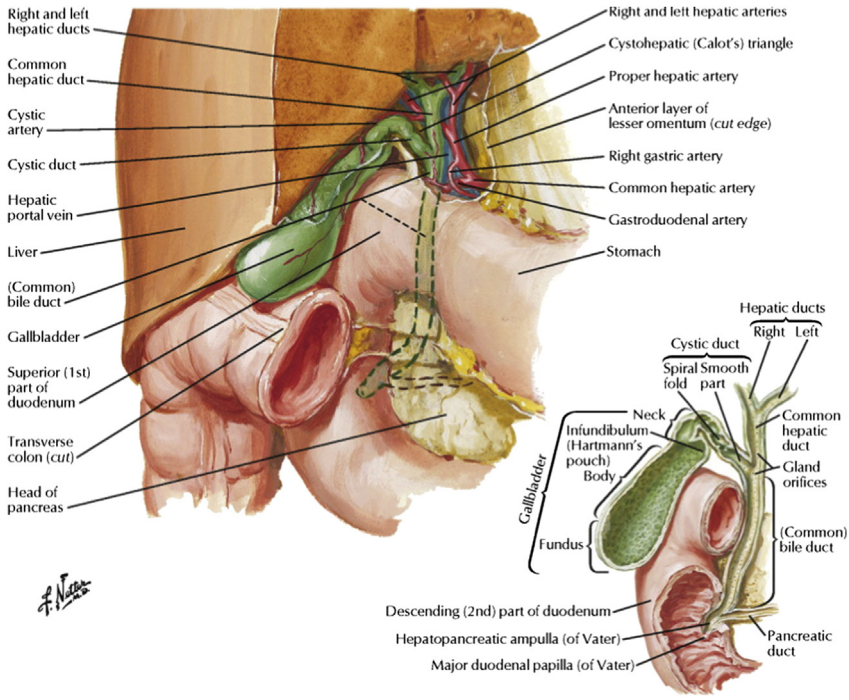

Fig. 3. Classic anatomy of the extrahepatic biliary ducts. (Netter illustration from www.netterimages.com. © Elsevier Inc. All rights reserved.)

trifurcates into the left gastric, splenic, and common hepatic arteries (Fig. 4). The common hepatic artery gives rise to the gastroduodenal and right gastric arteries before becoming the hepatic artery proper, which ascends in the hepatoduodenal ligament. The hepatic artery proper then bifurcates at a variable level into the left and right hepatic arteries before entering the liver. The left hepatic artery continues along the medial aspect of the hepatoduodenal ligament to enter the left liver through the

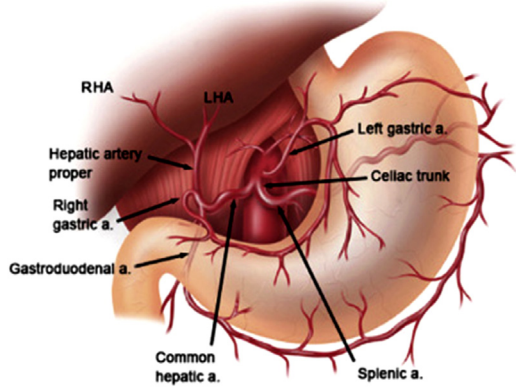

Fig. 4. Classic anatomy of the celiac axis. LHA, left hepatic artery; RHA, right hepatic artery. (*From* Geller DA, Goss JA, Tsung A. Liver. In: Brunicardi FC, Andersen DK, Billiar TR, et al, editors. Schwartz's principles of surgery. 9th edition. New York: McGraw-Hill Publishing; 2010; with permission.)

umbilical fissure. The right hepatic artery traverses from medial to lateral in the hepatoduodenal ligament, typically passing behind the common hepatic duct to enter the right liver. The cystic artery supplying the gallbladder usually branches off of the right hepatic artery. Thorough understanding of the relationships between these structures is imperative for general and hepatobiliary surgeons, particularly aberrant anatomy as described later.

The portal vein lies posterior to the CBD and the hepatic artery in the porta hepatis. It forms from the union of the superior mesenteric vein and the splenic vein, which receives venous blood from the inferior mesenteric vein. The left gastric (coronary) vein draining the lesser curve of the stomach empties into the portal vein near its origin. Similar to the hepatic artery proper, the portal vein branches bifurcate before entering the liver. The extrahepatic portal vein shows the least variation of the portal structures with the most common aberrancy being separate takeoffs of the anterior and posterior right portal branches from the main portal vein. Very little variation is seen in the left portal vein before entering the liver. The left, right, or both portal veins will provide blood supply to the caudate lobe.

Variations in Extrahepatic Biliary Anatomy

The right and left hepatic ducts run a short course outside of the liver parenchyma before forming the common hepatic duct. Rarely, the right and left ducts join within the liver. Alternatively, they may course separately and join lower in the hepatoduodenal ligament (Fig. 5).

Of particular importance is the first order branching of the right and left hepatic ducts within the liver. In a recent report based on radiographic imaging, there were atypical branching patterns of the right hepatic duct in 14% of patients, and there were atypical branching patterns of the left hepatic duct in 8%.[19] As shown in Fig. 6, the right anterior and posterior segmental branches can occur in many conformations, with the most worrisome being type A4 in which the right posterior segmental duct drains into the cystic duct. Ligating this duct may cause cholestasis in the

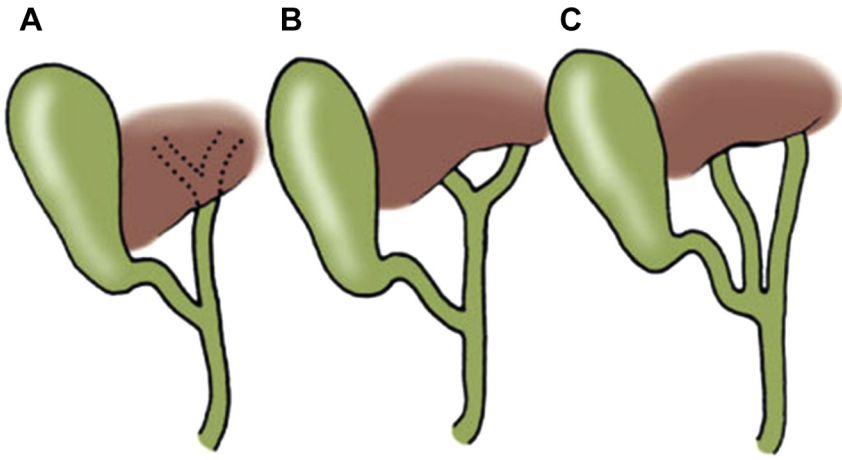

Fig. 5. Variation in confluence of right and left hepatic ducts. Intrahepatic (*A*), extrahepatic/typical (*B*), and low (*C*) confluence of the right and left hepatic ducts. (*From* Skandalakis JE, Branum GD, Colborn GL. Extrahepatic biliary tract and gallbladder. In: Skandalakis JE, Colburn GL, Weidman TA, editors. Skandalakis' Surgical Anatomy. Athens (Greece): Paschalidis Medical Publications, Ltd; 2004; with permission.)

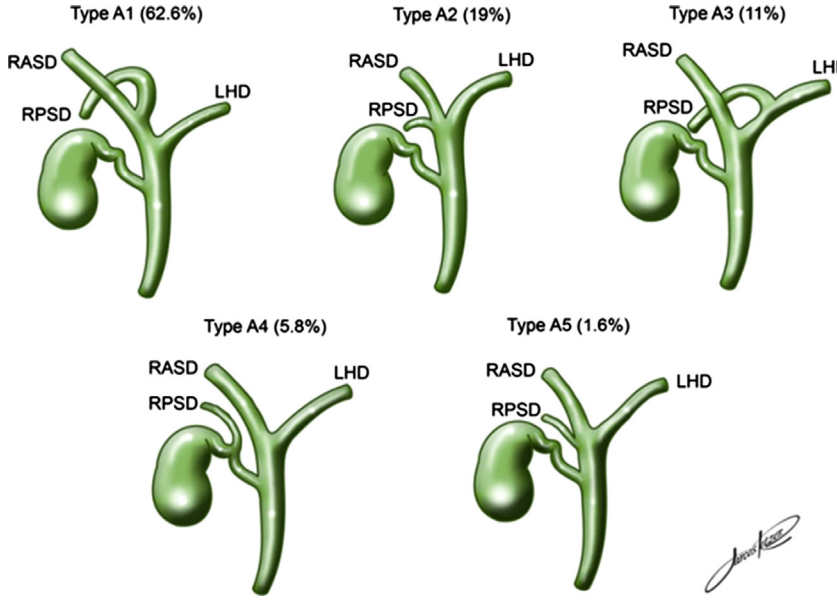

Fig. 6. Variations in sectoral drainage to right hepatic duct. LDH, left hepatic duct; RASD, right anterior hepatic duct; RPSD, right posterior hepatic duct. (*From* Chaib E, Kanas AF, Galvão FH, et al. Bile duct confluence: anatomic variations and its classification. Surg Radiol Anat 2013. [Epub ahead of print]; with permission.)

segment from which it drains. Given the wide variability in the conformation of these structures, intraoperative cholangiography can be immensely helpful in elucidating the anatomy and should be considered when unusual anatomy is encountered.

Variations in Hepatic Artery Anatomy

Only about 70% of patients have the classic hepatic arterial anatomy whereby the hepatic artery proper bifurcates to form the right and left hepatic arteries. Accessory right and left hepatic arteries may be found in addition to the usual right and left hepatic arteries (**Fig. 7**E–G) or the right or left hepatic arteries may be replaced, meaning they originate from another source entirely. Accessory arteries will usually supply a discreet segment of the liver; therefore, some argue that the nomenclature of the accessory hepatic artery is misleading. Rarely, the common hepatic artery derives completely from the superior mesenteric artery (SMA); this is known as the completely replaced common hepatic artery (see **Fig. 7**A).

The right hepatic artery usually courses posterior to the hepatic duct before entering the liver. In approximately one-fourth of patients, the right hepatic artery will lie anterior to the duct (see **Fig. 7**H). In 10%, the right hepatic artery will cross posterior to the portal vein (not pictured).[20] Nearly 20% of patients have a replaced right hepatic artery[21]; the most common origin of a replaced right hepatic artery is the SMA (see **Fig. 7**C). The replaced right hepatic artery can be found coursing upward posterior to the pancreas and portal vein. Approaching the level of the gallbladder, the right hepatic artery gives rise to the cystic artery, which can either course anteriorly or posteriorly to the hepatic duct. The replaced right hepatic artery then follows the usual course of the right hepatic artery into the liver hilum.[21] If a replaced right hepatic artery is ligated, the surgeon is obliged to perform a cholecystectomy because flow to the cystic artery is likely compromised. Ligation of the replaced right hepatic artery may

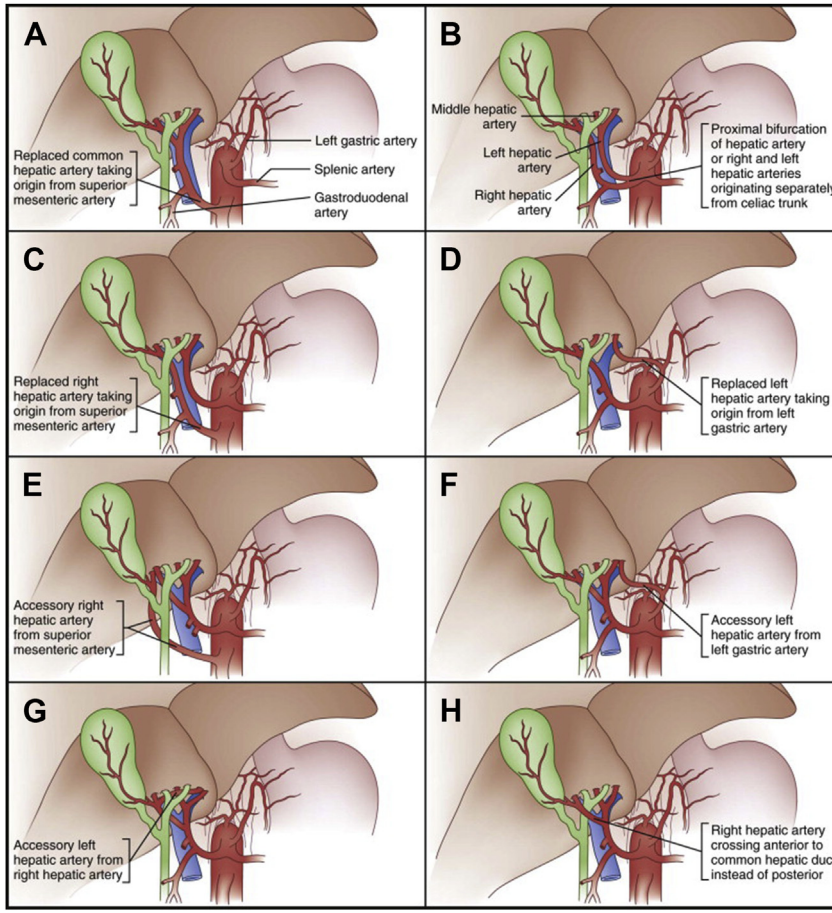

Fig. 7. Variations in hepatic arterial anatomy. (*From* Sabiston DC, Townsend CM. Sabiston textbook of surgery: the biological basis of modern surgical practice. 19th edition. Philadelphia: Elsevier Saunders; 2012; with permission.)

also be deleterious to a biliary enteric anastomosis because the loss of the blood supply to the bile duct predisposes the anastomosis to ischemia and leak.[22]

In approximately 15% of patients, a replaced left hepatic artery will be encountered (see **Fig. 7**D). In these patients, the left hepatic artery will most likely originate from the left gastric artery, course in the gastrohepatic ligament, and enter the liver at the hilum at the ligamentum teres. Although the replaced left hepatic artery is usually of little consequence in hepatic, biliary, and pancreatic surgery, it is relevant in gastric operations that require division of the gastrohepatic ligament. CT angiography can be of assistance in planning major surgeries in the region by delineating vascular aberrancies.

THE GALLBLADDER

Gallbladder Anatomy

The gallbladder is a muscular sac situated beneath the liver. Bile flowing from the liver drains to the CBD. The resting tone in the sphincter of Oddi prevents the flow of bile

into the duodenum and allows the bile to fill the duct with subsequent retrograde filling of the cystic duct and gallbladder. There, the bile is concentrated by the gallbladder epithelium, which contains channels that actively transport sodium chloride. Water follows, thereby concentrating the bile. The typical capacity of the gallbladder is 30 mL but it can distend to hold up to 300 mL of fluid, particularly in the face of chronic distal obstruction. The wall of the gallbladder is composed of the visceral peritoneum (on areas not in direct contact with the liver), subserosa, muscularis, lamina propria, and columnar epithelium. The parts of the gallbladder are named as seen in **Fig. 8**. They are the fundus, body, infundibulum (Hartman pouch), and the neck.

The neck drains into the cystic duct. The lumen of the cystic duct is characterized by mucosal folds called the *spiral valves of Heister*. The cystic duct can run a very short course draining into the right hepatic duct or it can course alongside the common hepatic for a distance with insertion just above the pancreas. Congenital absence of the gallbladder is discussed earlier. Importantly, the gallbladder can be intrahepatic. This possibility should be entertained when working up gallbladder agenesis or when the gallbladder is not visualized in biliary surgery. Other rare anomalies of the gallbladder and cystic duct have been described, including duplication of the gallbladder as well as the left-sided gallbladder (draining into the left hepatic duct or common hepatic duct). These anomalies are exceedingly rare.

The blood supply to the gallbladder is from the cystic artery, which is usually a branch off of the right hepatic artery. Not surprisingly, there is significant variation in the course of the cystic artery. Rarely, it may branch from the left hepatic artery or hepatic artery proper, running anteriorly to the hepatic duct on its course to the gallbladder. It may arise from a replaced right hepatic artery from the SMA as mentioned earlier. **Fig. 9** shows the various conformations as well as their prevalence.

Venous drainage of the gallbladder includes veins that follow along the cystic and hepatic ducts to drain into the liver via the portal system as well as veins that drain directly from the gallbladder into the liver. Lymphatic vessels in the gallbladder are located in the subserosal layer and drain to the Calot node and lymph nodes along the porta hepatis. Lymphatic drainage can also course directly into the liver along

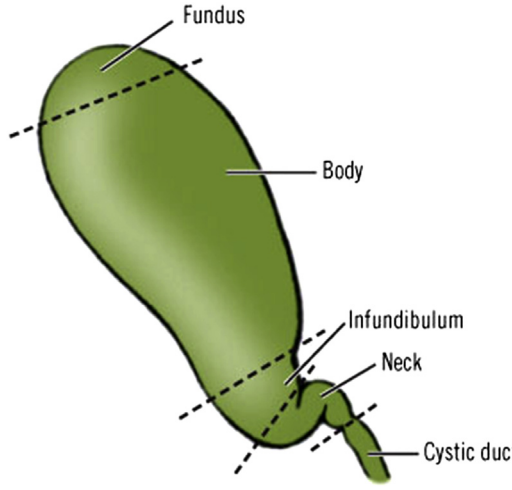

Fig. 8. The gallbladder. (*From* Skandalakis JE, Branum GD, Colborn GL. Extrahepatic biliary tract and gallbladder. In: Skandalakis JE, Colburn GL, Weidman TA, editors. Skandalakis' Surgical Anatomy. Athens (Greece): Paschalidis Medical Publications, Ltd; 2004; with permission.)

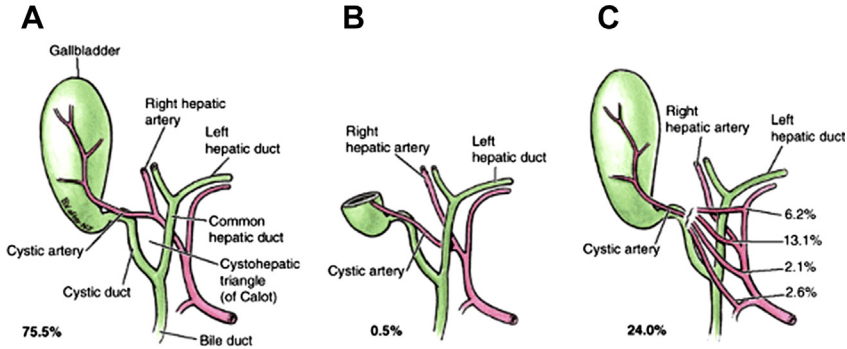

Fig. 9. Variations in cystic artery anatomy. (*A*) Most common configuration of the cystic artery, branching from right hepatic artery. (*B*) Less common, the cystic artery is seen branching from the right hepatic artery prior to crossing posterior to the common hepatic duct. (*C*) The cystic artery is seen branch from various arteries and courses anterior to the common hepatic duct. (*From* Agur AMR, Grant JCB. Grant's atlas of anatomy. 11th edition. Philadelphia: Lippincott Williams & Wilkins; 2005; with permission.)

segments V and IVB before reaching lymph nodes within the hepatoduodenal ligament. Hence, radical resection for gallbladder cancer routinely includes these segments of liver.

Contraction of the gallbladder is under the regulatory control of multiple signals. The major positive mediators of contraction are cholecystokinin (CCK) and parasympathetic innervation. CCK is a hormone secreted by the epithelium in the duodenum in response to intraluminal nutrients. CCK secretion results in postprandial gallbladder contraction to move bile into the duodenum for digestion. The hepatic branch of the vagus nerve supplies parasympathetic innervation, which also promotes contraction. Like the rest of the intestinal tract, the gallbladder is innervated by the enteric nervous system, promoting coordination with the migratory motor complex. There are also multiple modulators of gallbladder contraction. The gallbladder receives innervation by the sympathetic system via the celiac plexus. Sympathetic stimulation promotes relaxation of the gallbladder smooth muscle. Recent literature also supports that components of bile itself dampen gallbladder contractions through G-protein coupled receptors.[23] Stasis of bile in the gallbladder is thought to contribute to the formation of cholelithiasis.

Ducts of Luschka

One feared complication of the laparoscopic cholecystectomy is bile leak. Although the more common location for leak is the cystic duct stump, leak from a duct of Luschka is the second most common culprit. There is great controversy over the term *duct of Luschka*, and many researchers prefer the use of the term *subvesical bile duct*. In a recent systematic review of the literature, Schnelldorfer and colleagues[24] categorized subvesical bile ducts into 4 subtypes based on anatomic characteristics:

- Accessory segmental subvesical bile duct: an intrahepatic duct running along gallbladder fossa draining a segment of the liver that is drained by another intrahepatic duct
- Segmental subvesical bile duct: an intrahepatic duct running along gallbladder fossa draining a discreet segment of the liver

- Aberrant subvesical bile ducts: a network of ducts that end blindly in the connective tissue surrounding the gallbladder but that are in continuity with hepatic ducts, suggesting embryologic origin
- Hepaticocholecystic bile duct: a duct that drains from the liver directly into the gallbladder

Prevalence of these subtypes is not known; however, accessory segmental and segmental subvesical bile ducts are likely common, whereas aberrant subvesical bile ducts and hepaticocholecystic bile ducts are rare.[24] Injury occurs during the removal of the gallbladder from the fossa when the plane of dissection is too deep into the liver bed. Theoretically, the leak can be identified by direct visualization of the gallbladder fossa with identification of bilious drainage. More often, however, these leaks are identified in the postoperative period. Imaging may be helpful in defining the type of duct that has been injured; however, the main principle of treatment is drainage of the bile. In patients whose bile leak fails to improve, endoscopic sphincterotomy and stenting may be necessary to decrease luminal biliary pressures.[25]

AMPULLARY ANATOMY AND PHYSIOLOGY

Much like the rest of the biliary tract, the ampulla of Vater has variable anatomy. Classically, the CBD is described as coursing along the pancreatic groove, curving, then traversing the wall of the duodenum obliquely. The CBD joins with the pancreatic duct to form a short common channel that drains into the duodenum at the major papilla (**Figs. 10**B and **11**). This anatomy is present in about 60% of patients. Most other patients will have ducts that remain separate through the wall of the duodenum but share an opening at the papilla, the so-called double barrel (see **Fig. 10**A). Rarely, the ducts empty into the duodenum separately.[26] The sphincter of Oddi surrounds the ducts in the wall of the duodenum.

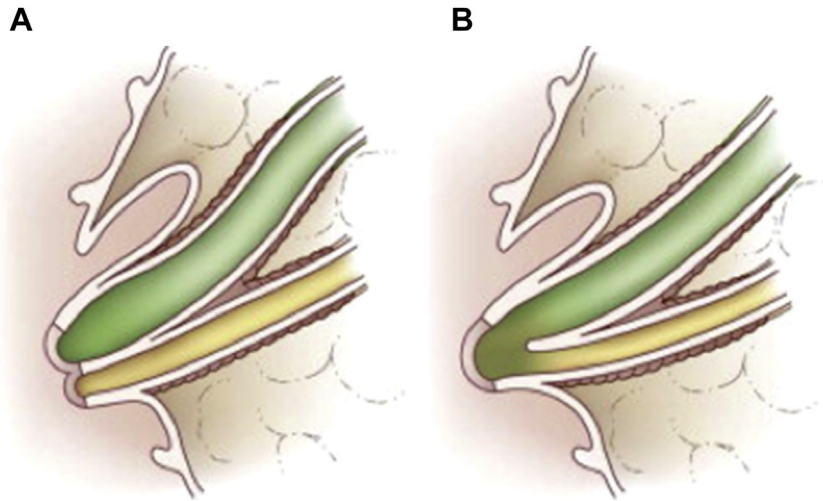

Fig. 10. Variable anatomy of biliary drainage into the duodenum. (*A*) Double barrel opening of CBD and pancreatic duct. (*B*) Common channel shared by CBD and pancreatic duct. (*From* Sabiston DC, Townsend CM. Sabiston textbook of surgery: the biological basis of modern surgical practice. 19th edition. Philadelphia: Elsevier Saunders; 2012; with permission.)

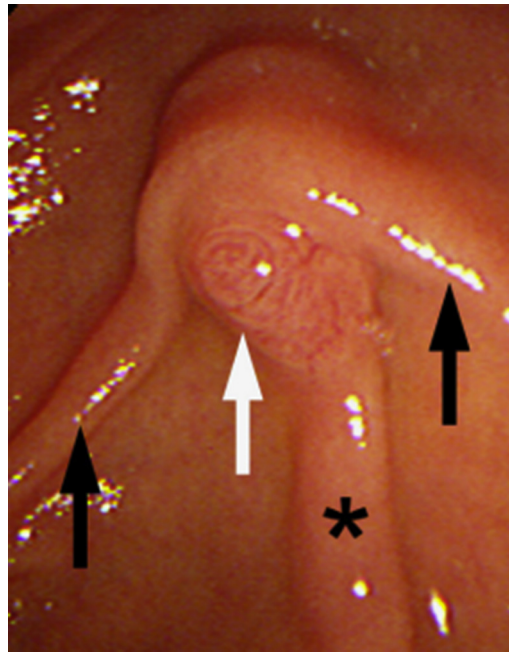

Fig. 11. Major papilla (*white arrow*), transverse mucosal folds (*black arrows*), longitudinal mucosal fold (*asterisk*). (*From* Kim TU, Kim S, Lee JW, et al. Ampulla of Vater: comprehensive anatomy, MR imaging of pathologic conditions, and correlation with endoscopy. Eur J Radiol 2008;66(1):48–64.)

The sphincter of Oddi exhibits rhythmic contractions above a basal pressure greater than that in the duodenum. When stimulated by cholecystokinin, the sphincter relaxes and allows the flow of bile into the digestive tract. In addition to CCK, the sphincter is regulated by the autonomic nervous system and the enteric nervous system. As the migratory motor complex causes contraction in the duodenum, the tone of the sphincter likewise increases.[27] When the CBD and pancreatic duct share a long common channel, the sphincter's contraction fails to occlude both ducts; the confluence allows regurgitation of pancreatic fluid into the biliary tree, a proposed mechanism for the formation of choledochal cystic disease as mentioned earlier. Regurgitation is also thought to lead to pancreatitis in some patients.

SOD

In patients with abdominal pain characterized by biliary colic in the absence of gallbladder pathology (eg, cholelithiasis and biliary dyskinesia) or in patients with recurrent idiopathic pancreatitis, the diagnosis of SOD should be entertained. Patients presenting with biliary pain may exhibit abnormal liver function testing (defined as greater than twice normal on 2 occasions) and/or may have a dilated CBD. The 2 main causes are stenosis and physiologic dysfunction of the sphincter.[28] SOD can be classified as one of 3 types according to the Milwaukee classification:

Type I: biliary colic, elevated liver enzymes, bile duct dilation, and delayed drainage of contrast from duct on ERCP

Type II: biliary colic and 1 or 2 of elevated liver enzymes, bile duct dilation, or delayed drainage of contrast on ERCP
Type III: biliary colic only

Manometry can be useful and is often necessary to make the diagnosis. A basal pressure of greater than 40 mm Hg meets the criteria for SOD.[28] Manometry carries a significant risk of pancreatitis. Especially for those with type II or type III SOD, diagnosis can be challenging; many treatment options have limited data to support their use. Medical treatment, such as with calcium channel blockers or nitrates, may or may not be helpful; most options carry limiting side-effect profiles. According to a 2001 Cochrane Review, sphincterotomy is most likely to be helpful if manometry reveals elevated biliary basal pressures.[29]

SUMMARY

The anatomy of the biliary tract exhibits a wide degree of variation. The embryologic development of the biliary tract is highly complicated. It is better understood today than even just 5 years ago, but significant gaps in knowledge still exist. Therefore, the cause of congenital abnormalities, including biliary atresia and choledochal cystic disease, remains poorly understood and will continue to require surgical intervention.

Knowledge of anatomic variation is important in the operative setting. When the usual appearance of structures is not encountered, it can be tempting to fit abnormal findings within the paradigm of what is normal. This practice can lead to errors and injury. Intraoperative cholangiography can be helpful in interpreting the anatomy and should be used liberally. Similarly, the variation in hepatic vasculature can represent a challenge, and inadvertent ligation or injury can lead to poor outcomes. Recognition of variable anatomy is crucial in avoiding complication and achieving optimal outcomes for patients. When one anatomic variant is encountered, the surgeon must be on the lookout for others.

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