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Embryology

It is important to know that the anatomy of the biliary tract is intimately associated with both the liver and the pancreas. These three organs are closely associated with each other during embryologic development. At the 3-mm stage, three buds can be recognized (Fig. 79.1). The cranial bud develops into two lobes of the liver, whereas the caudal bud becomes the gallbladder and extrahepatic biliary tree. The ventral pancreas, which eventually becomes the pancreatic head and uncinate process, also develops from the caudal bud. At the 7-mm stage, the liver and hepatic ducts have formed. The gallbladder, cystic duct, and ventral pancreas have arisen from the common duct. By the 12-mm stage, the ventral and dorsal buds of the pancreas form the complete pancreas. Within the 8th week of gestation, a completely open lumen has formed in the gallbladder, bile ducts, and pancreatic ducts.

Anatomy

The anatomy of the biliary tract can be divided into various segments: the intrahepatic ducts, the extrahepatic ducts, the gallbladder and cystic duct, and the sphincter of Oddi.

The portal vein, hepatic artery, and biliary tree follow roughly parallel courses and bifurcate just before entering the liver. This major bifurcation

divides the liver into left and right lobes. According to Couinaud's classification, the caudate lobe is segment I; segments II, III, and IV are on the left; and segments V, VI, VII, and VIII are on the right (Couinaud 1957).

Intrahepatic Ducts

The left hepatic duct drains the left side of the liver (segments II through IV) and joins the right hepatic duct to form the common hepatic duct. The right hepatic duct drains segments V through VIII and is formed from the union of the right posterior and right anterior segmental ducts. The right posterior segmental duct is formed by the confluence of ducts draining segments VI and VII. The union of the ducts draining segments V and VIII forms the right anterior segmental duct. The right posterior segmental duct has an almost horizontal course, whereas the right anterior segmental duct tends to have a more vertical orientation (Figs. 79.2 and 79.3). The biliary drainage of the caudate lobe (segment I) is variable. However, in majority of the individuals, the caudate lobe drains into both the right and left hepatic ducts (Mortele and Ros 2001).

Extrahepatic Ducts

Most patients have a bifurcation where the right and left hepatic ducts join to form the common hepatic duct. In some patients, three hepatic ducts join to form the common hepatic duct. Usually, the hepatic ducts meet just outside of the liver parenchyma, with the

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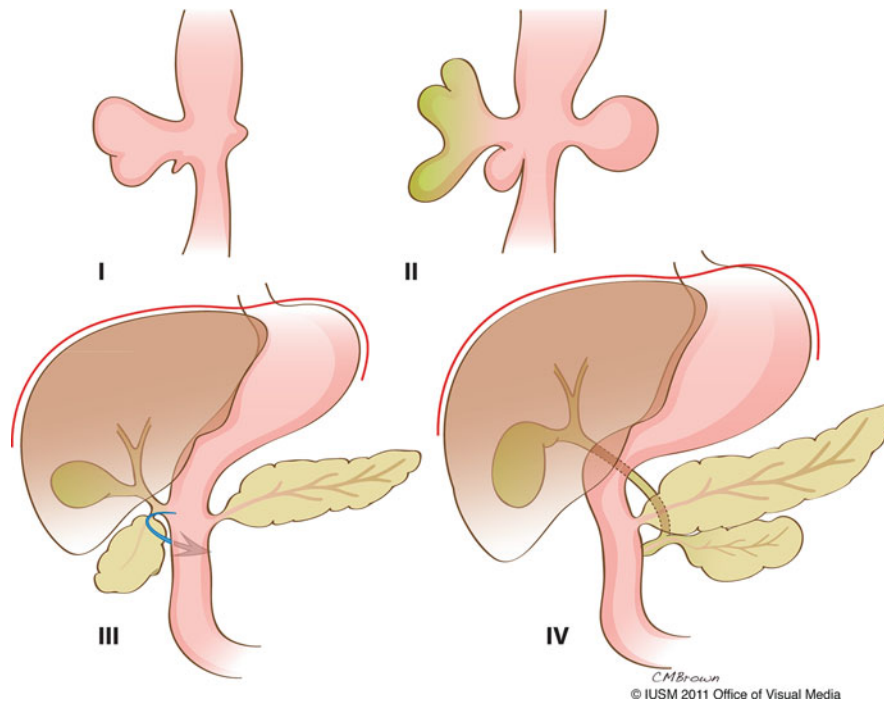
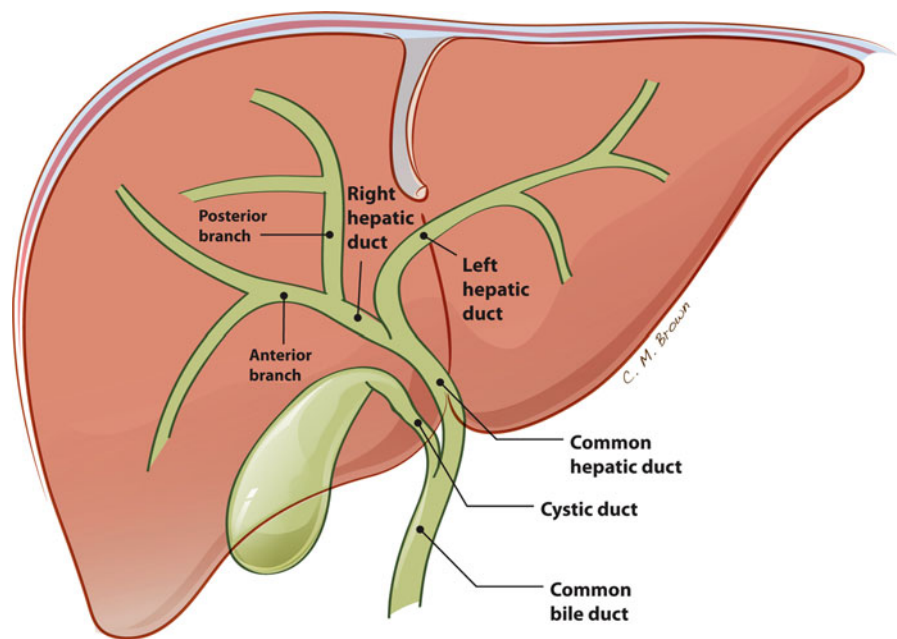


Fig. 79.1 Embryologic development of the biliary ducts and gallbladder. Anatomy of the gallbladder and biliary ducts is closely associated with the liver and pancreas. Three buds can be identified at the 3-mm stage (*I*) and 5-mm stage (*II*). At the 7-mm stage (*III*) the liver and hepatic ducts have formed. The ventral bud rotates with the gut, passing behind the duodenum

from the right to left, and eventually fuses with the dorsal bud by about the 7th week of gestation. Failure of fusion of the ventral and dorsal duct systems occurs in approximately 10% of individuals, resulting in pancreas divisum. By the 12-mm stage (*IV*), the ventral and dorsal buds of the pancreas form the complete pancreas

Fig. 79.2 Biliary ducts and gallbladder. The right hepatic duct is formed from the union of the right posterior and right anterior segmental ducts. The right posterior segmental duct has an almost horizontal course, whereas the right anterior segmental duct tends to have a more vertical orientation. The right and left hepatic ducts join to form the common hepatic duct. This is the most common form of right hepatic ductal configuration. The union of the cystic and common hepatic ducts forms the common bile duct



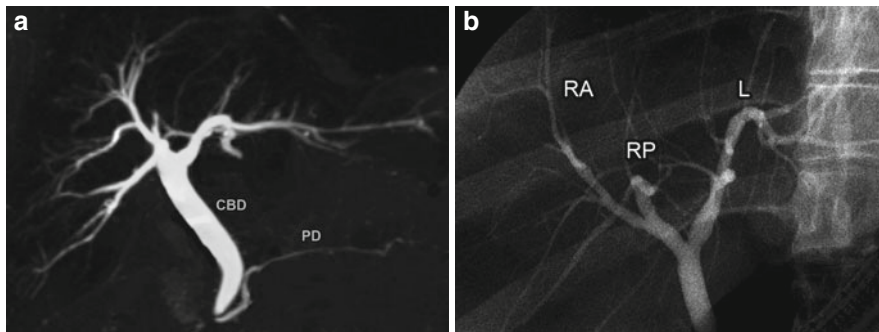


Fig. 79.3 Intrahepatic and extrahepatic biliary ducts. (a) Magnetic resonance cholangiopancreatogram (MRCP) is the best non-invasive imaging technique to evaluate the biliary tree and the pancreatic duct. This maximum intensity projection (MIP) image in coronal projection demonstrates the intra and extrahepatic

biliary tree and pancreatic duct (*CBD* Common bile duct, *PD* pancreatic duct). (b) Endoscopic retrograde pancreatocholangiogram (ERCP) image of the intrahepatic ducts demonstrating the normal intrahepatic ductal anatomy (*RA* right anterior segment, *RP* right posterior segment, *L* left hepatic duct)

cystic duct entering 2–3 cm distally. The union of the cystic and common hepatic ducts forms the common bile duct. It varies in length from 5–15 cm, depending on the actual position of the ductal union. The average diameter is about 6 mm. The common bile duct can be divided into four portions or segments: supra-duodenal, retro-duodenal, intrapancreatic, and intramural. The lower third, or intrapancreatic portion, of the common bile duct traverses the posterior aspect of the pancreas to enter the second portion of the duodenum, where the pancreatic duct usually joins it. The intramural or intraduodenal portion of the common bile duct passes obliquely through the duodenal wall to enter the duodenum at the papilla of Vater. The lower portion of the common bile duct and the terminal portion of the pancreatic duct are enveloped and regulated by a complex sphincter, the sphincter of Oddi.

Gallbladder and Cystic Duct

The gallbladder is a pear-shaped organ that lies on the inferior surface of the liver at the junction of the left and right hepatic lobes between Couinaud's segments IV and V. The gallbladder varies from 7 to 10 cm in length and from 2.5 to 3.5 cm in width. The gallbladder's volume varies considerably, being large during fasting states and small after eating. The gallbladder has been divided into four areas: the fundus, body, infundibulum, and neck. Hartmann's pouch is an asymmetrical bulge of the infundibulum that lies

close to the gallbladder's neck (Frierson 1989). It projects inferiorly and posteriorly toward the duodenum. Gallstones frequently lodge in the Hartman's pouch and if the gallbladder is inflamed, this pouch may adhere to the cystic duct.

Rokitansky-Aschoff sinuses are invaginations of epithelium into the lamina propria, muscle, and subserosal connective tissue of the gallbladder. These sinuses are present in about 40% of normal gallbladders and are present in abundance in almost all inflamed gallbladders.

The cystic duct arises from the gallbladder and joins the common hepatic duct to form the common bile duct. The length of the cystic duct is variable, averaging between 2 and 4 cm. The cystic duct usually courses downward in the hepatoduodenal ligament to join the lateral aspect of the supra-duodenal portion of the common hepatic duct at an acute angle. The cystic duct contains a variable number of mucosal folds, similar to those found in the neck of the gallbladder. Although referred to as valves of Heister, these spiral folds do not have a valvular function.

Sphincter of Oddi

The sphincter mechanism is subdivided into several sections and has separate sphincters for the distal bile duct, the pancreatic duct, and the ampulla. In more than 90% of the population, the common channel, where the biliary and pancreatic ducts join, is less than 1.0 cm in length and lies within the ampulla.

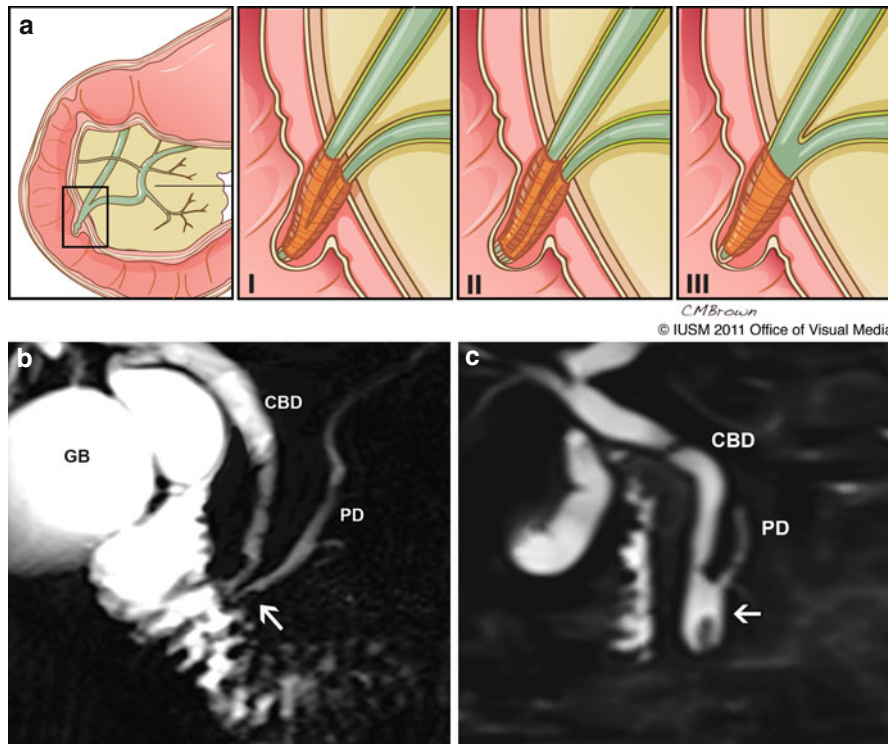


Fig. 79.4 Sphincter of Oddi. (a) Illustration of the sphincter of Oddi variations. *I* common bile duct and pancreatic duct join together and are encircled by the sphincter, *II* common bile duct and pancreatic duct encircled by the sphincter, however, do not join, *III* anomalous junction of pancreaticobiliary ductal system (AJPBDS). There is a long common channel of the biliary and pancreatic duct that is not entirely encircled by the sphincter, therefore, bi-directional reflux of bile or pancreatic excretions can occur. Accordingly, various pathological conditions such as

cholangitis, gallstones, biliary tract or gallbladder cancer, pancreatitis, and pancreatic calculi can develop. (b) Coronal MRCP image shows the common bile duct (CBD) and pancreatic duct (PD) having a separate channel within the sphincter (arrow). (c) Coronal MRCP image shows a long common channel (arrow) that is not covered by the sphincter. This condition results in free communication between the common bile duct (CBD) and the pancreatic duct (PD). Filling defect within the common channel is a calculus

In the rare situation in which the common channel is longer than 1.0 cm or the biliary and pancreatic ducts open separately into the duodenum, pathologic biliary or pancreatic problems are likely to develop (Fig. 79.4).

Anomalous junction of pancreaticobiliary ductal system (AJPBDS) is a congenital anomaly defined as a union of the pancreatic and biliary ducts that is located outside the duodenal wall. Because of the lack of the sphincter muscle (sphincter of Oddi) at the union, two-directional regurgitations may occur. The anomalous junction is linked with several complications including cholangitis, pancreatitis, and biliary and pancreatic calculi formation. Prophylactic surgical interventions are recommended for increased risk of biliary tract or gallbladder cancer (Miyazaki et al. 2008).

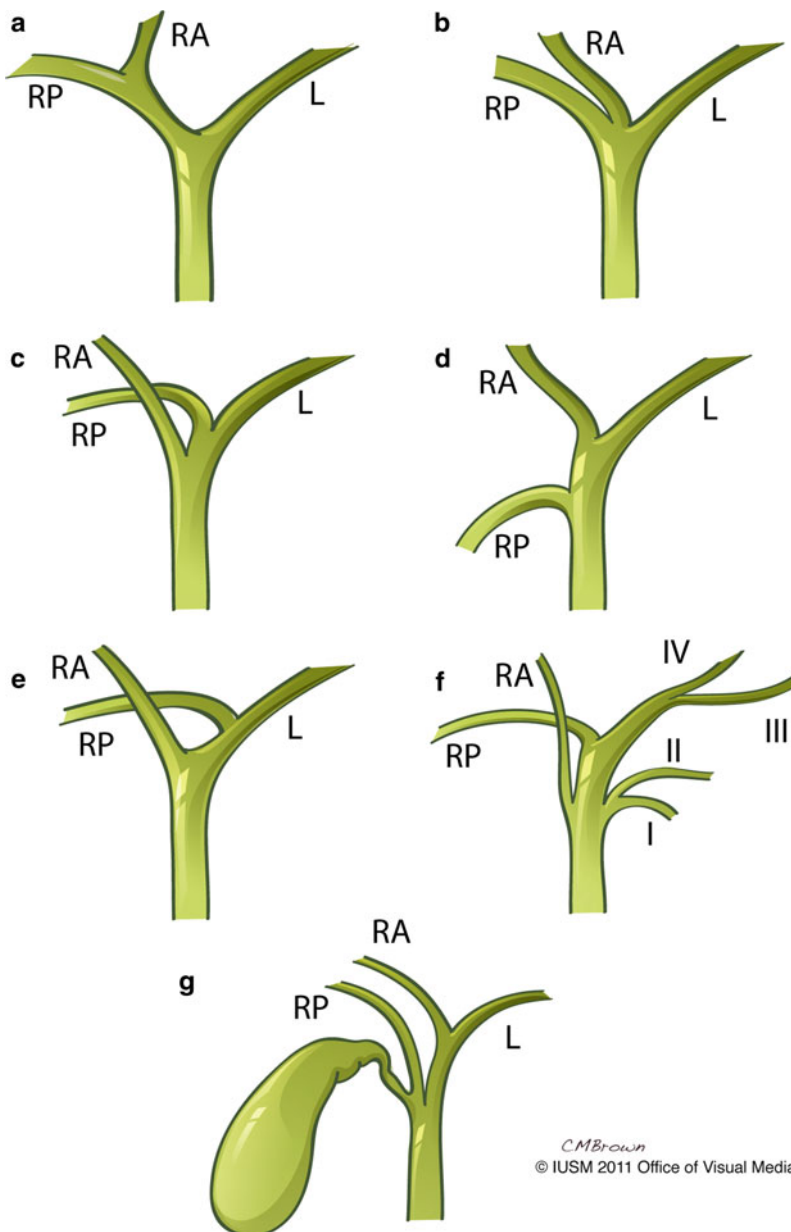
Vascular

The blood supply to the right and left hepatic ducts and upper portion of the common hepatic duct arises from the cystic artery and the right and left hepatic arteries. The cystic artery usually arises as a single branch from the right hepatic artery. The venous drainage from the hepatic ducts and hepatic surface of the gallbladder is through small vessels that empty into branches of the hepatic veins within the liver. A small venous trunk ascending parallel to the portal vein receives veins draining the gallbladder and bile duct.

Lymphatic Drainage

Lymphatic vessels from the hepatic ducts and upper common bile duct drain into the hepatic lymph nodes. Lymph from the lower bile duct drains into the lower hepatic nodes as well as the upper

Fig. 79.5 Variations of the intrahepatic bile ducts. (a) The right anterior and right posterior intrahepatic ducts join, and the right hepatic duct unites with the left hepatic duct to form the common hepatic duct; 57–68%. (b) The right anterior, right posterior, and left hepatic ducts unite to form the common hepatic duct; 12–18%. (c) The right posterior and left hepatic ducts join to form the common hepatic duct, and the right anterior duct joins below the union; 8–20%. (d) The right posterior duct joins the common hepatic duct below the union of the right anterior and the left hepatic ducts; 4–7%. (e) The right posterior duct joins the left hepatic duct distal from the bifurcation; 6%. (f) Multiple segmental branches join the left hepatic duct before or at the junction with the right hepatic ducts; 2%. (g) The cystic duct joins at the union of all the ducts or with one of the right hepatic ducts; 1.5–3%



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pancreatic lymph nodes. Lymphatic vessels from the gallbladder and cystic duct drain primarily into the hepatic nodes.

Anomalies

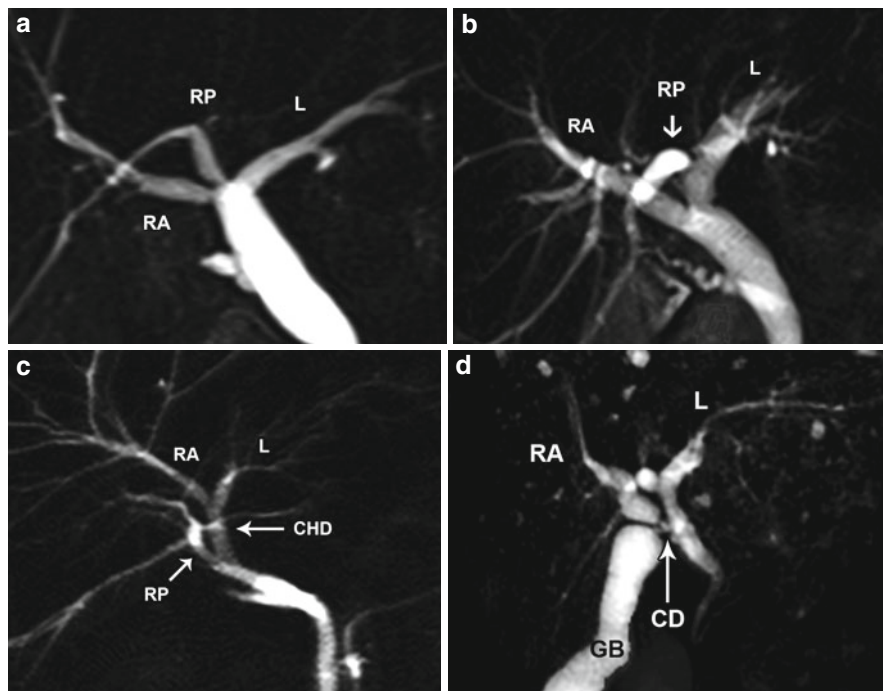
The anatomy of the extrahepatic biliary tree is highly variable. A thorough knowledge of this variable anatomy is important because failure to recognize the

frequent anatomic variations may result in significant ductal injury. Anomalies of the extrahepatic biliary tree may involve the hepatic ducts, common bile duct, or cystic duct.

Biliary Ducts

Bile duct injuries can occur after either open or laparoscopic cholecystectomies (Fig. 79.5). Variant biliary

Fig. 79.6 Variations of the intrahepatic bile ducts. (a) Coronal MRCP image shows the right anterior (*RA*), right posterior (*RP*), and left (*L*) hepatic ducts join to form a trifurcation. (b) Coronal MRCP image shows the right posterior duct (*RP*) joining the left hepatic duct (*L*). (*RA* right anterior). (c) Coronal MRCP image shows the right posterior duct (*RP*) joining the common hepatic duct (*CHD*). (*RA* right anterior, *L* Left hepatic duct). (d) Coronal MRCP image shows the right anterior duct (*RA*) joining the cystic duct (*CD*). (*CBD* common bile duct, *L* Left hepatic duct)



anatomy is one of the factors that may contribute to the occurrence of bile duct injury after laparoscopic cholecystectomy.

In 57–68% of patients, the right anterior and right posterior intrahepatic ducts join, and the right hepatic duct unites with the left hepatic duct to form the common hepatic duct (Yoshida et al. 1996). The most common anatomic variants in the branching of the biliary ducts involve the right posterior duct and its fusion with the right anterior or left hepatic duct (Gazelle et al. 1994). In 12–18% of patients, the right anterior, right posterior, and left hepatic ducts unite to form the common hepatic duct. In 8–20% of patients, the right posterior and left hepatic ducts join to form the common hepatic duct, and the right anterior duct joins below the union. In 4–7% of patients, the right posterior duct joins the common hepatic duct below the union of the right anterior and the left hepatic ducts. In 1.5–3% of patients, the cystic duct joins at the union of all the ducts or with one of the right hepatic ducts (Fig. 79.6).

Accessory hepatic ducts may emerge from the liver to join the right hepatic duct, common hepatic

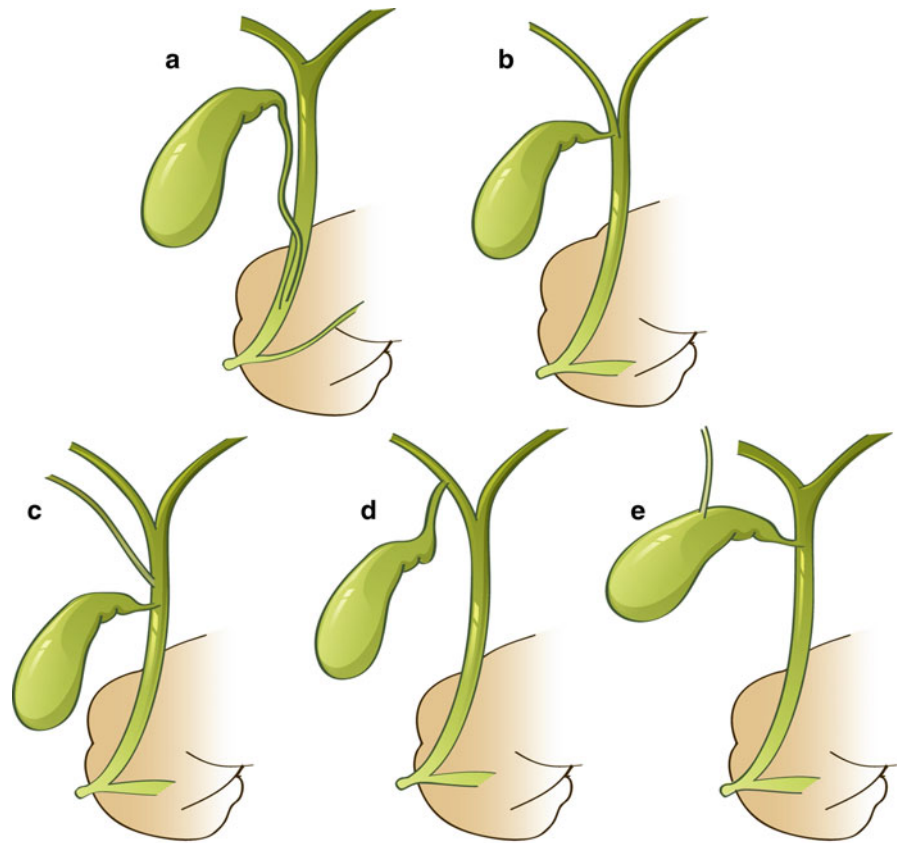
duct, cystic duct, common bile duct, or gallbladder. These ducts are present in approximately 10% of individuals.

Several types of common bile duct malposition or duplications of the common bile duct are reported; however, these are rare anomalies.

Cystic Duct

The cystic duct may run parallel to the common hepatic duct for a variable distance, or it may run spiral anterior or posterior to the common hepatic duct to form a left-sided union (Fig. 79.7). The cystic duct may be normal in length or may course downward in the hepatoduodenal ligament for a considerable distance before forming a low union with the common hepatic duct. The cystic duct may join the right hepatic duct or a right segmental duct. Less often, the cystic duct, right hepatic duct, and left hepatic duct may join at the same level to form a trifurcation (Fig. 79.8) (Benson and Page 1976).

Fig. 79.7 Cystic duct variations. (a) Long cystic duct with low fusion with the common hepatic duct. (b) High fusion of cystic duct with common hepatic duct (trifurcation). (c) Accessory hepatic duct. (d) Cystic duct draining into the right hepatic duct. (e) Cholecystohepatic duct



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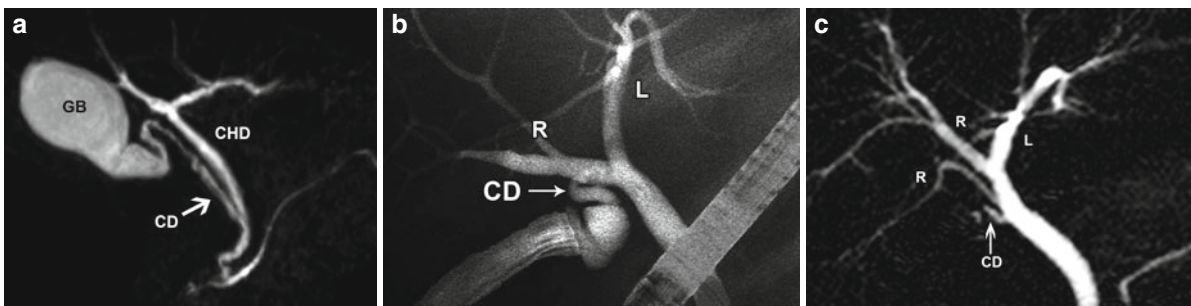
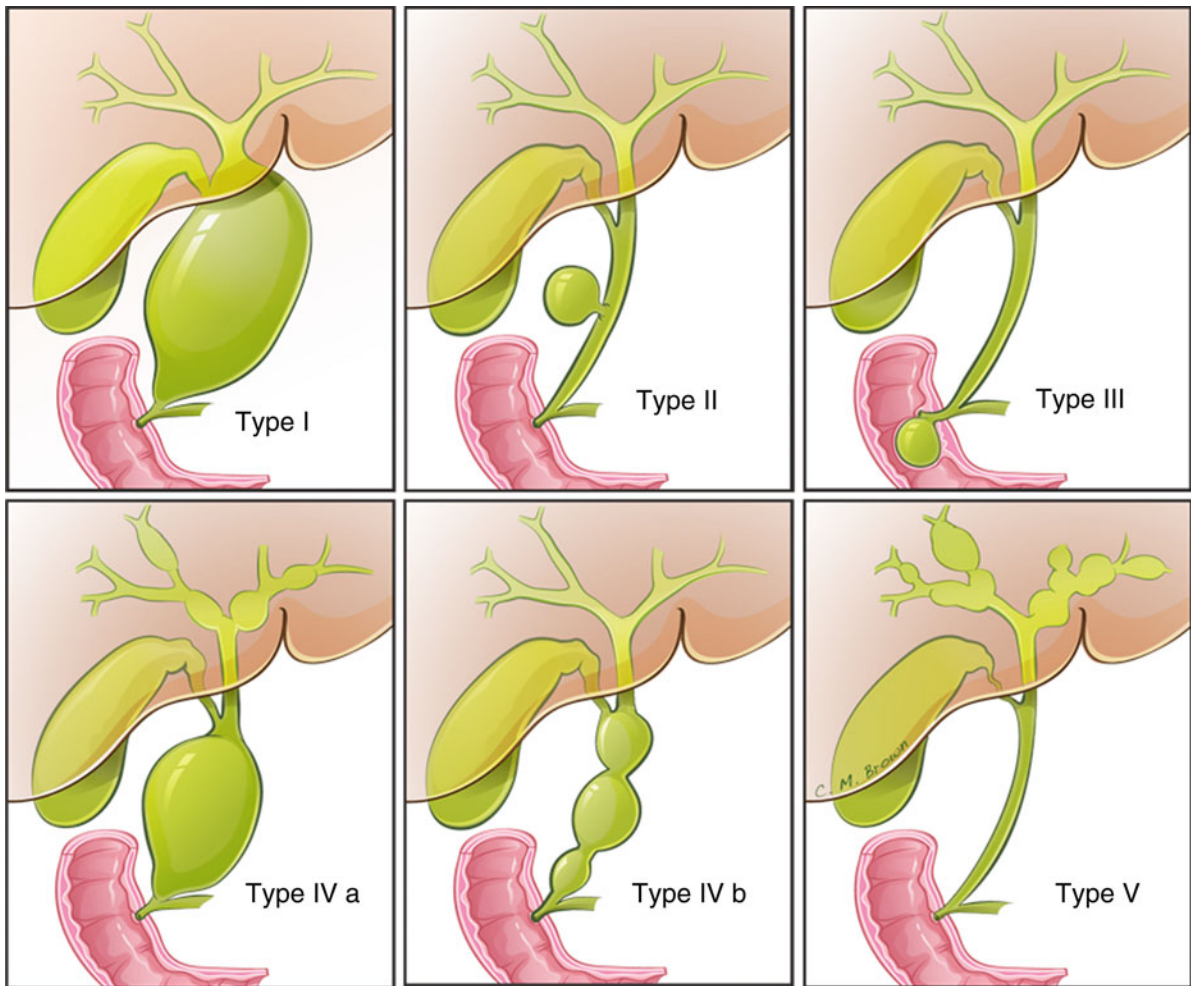


Fig. 79.8 Ductal anomalies. (a) MRCP image shows that the cystic duct (CD) is unusually long, following a parallel course to the common hepatic duct (CHD) until the insertion. (b) High insertion of the cystic duct. Endoscopic retrograde cholangiopancreatogram (ERCP) image shows the cystic duct (CD) joining the right (R) and left (L) hepatic ducts at the level of junction

forming a trifurcation. (c) Accessory right hepatic duct. Coronal MRCP image shows two right hepatic ducts (R). The dominant right hepatic duct joining the left hepatic duct (L) to form the common hepatic duct, the second duct draining into the common hepatic duct proximal to the insertion of the cystic duct remnant (CD)



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Fig. 79.9 Choledochal cysts are rare congenital biliary tract anomalies characterized by dilatation of all or part of the extra or intrahepatic biliary tree. There are five subtypes as defined by Todani. Type I can be subdivided into type Ia (diffuse) cysts, which involve the entire extrahepatic biliary ducts (shown here); type Ib (focal) cysts, which involve only a focal segment of the extrahepatic biliary ducts; and type Ic (fusiform) cysts, which involve only the common bile duct. Type II cysts are true diverticulum of the extrahepatic bile duct. Type III cysts, also

known as choledochoceles, are focal dilatation of the intraduodenal segment of the distal CBD. Type IV choledochal cysts can be subdivided into type IVa cysts, which involve both the extrahepatic biliary ducts and the intrahepatic bile ducts (shown here); and type IVb cysts, which involve only the extrahepatic biliary ducts with multiple saccular dilatations. Type V Choledochal Cyst (Caroli Disease) show intrahepatic saccular or fusiform-dilated cystic structures of varying sizes that communicate with the biliary tree

Choledochal Cyst

Choledochal cysts are rare congenital biliary tract anomalies characterized by dilatation of all or part of the extra or intrahepatic biliary tree. Choledochal cysts are most common in adult females, who complain about biliary tract symptomatology or perhaps

pancreatitis. A right upper quadrant mass with pain and jaundice in infants suggests a choledochal cyst. There are five subtypes as defined by Todani (Todani et al. 1977) (Figs. 79.9 and 79.10).

Type I choledochal cysts are confined to the extrahepatic biliary ducts. They can be further subdivided

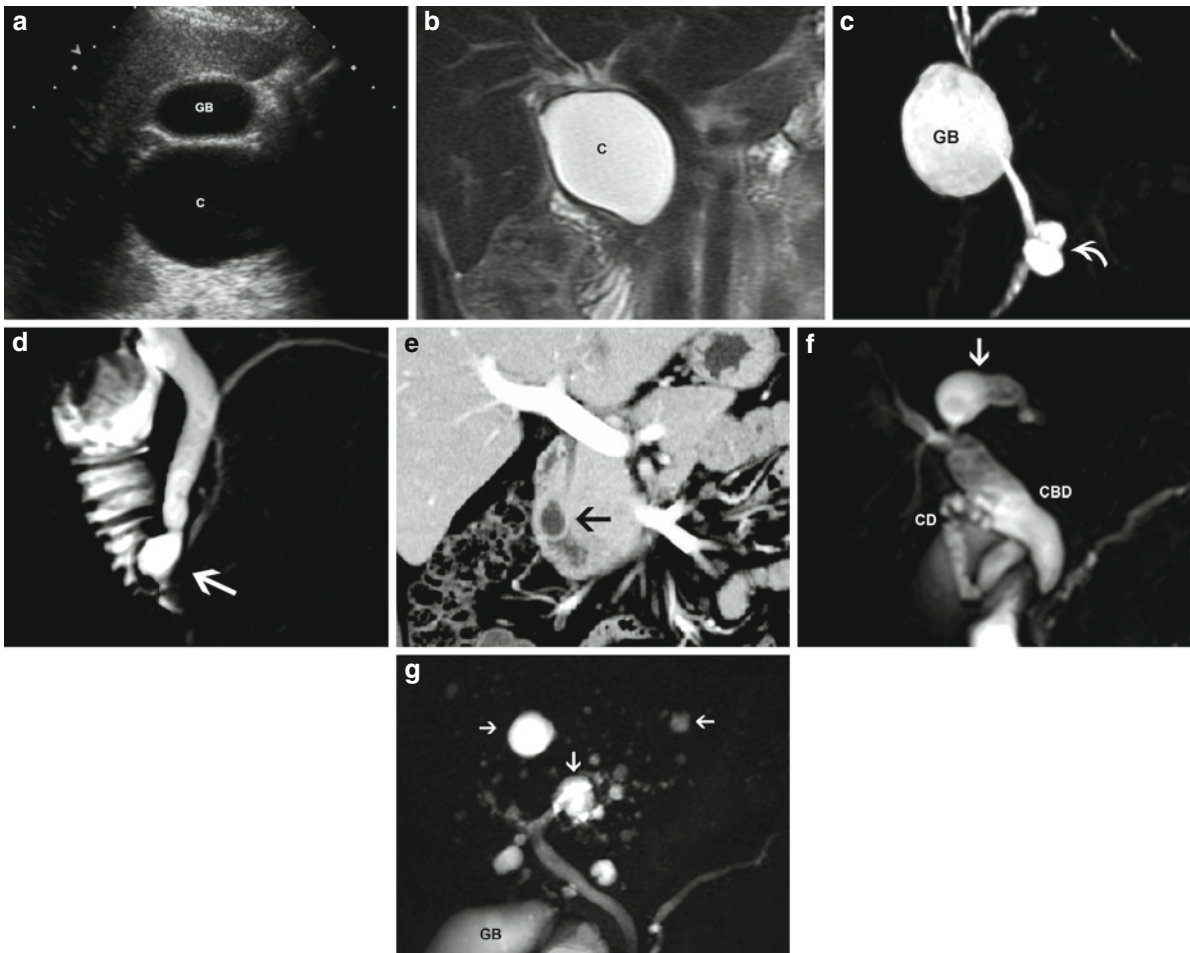


Fig. 79.10 Choledochal cysts. (a) Type I. Ultrasound image of the right upper quadrant in transverse projection demonstrates an anechoic cystic structure (C) located inferior to the gallbladder (GB). (b) Type I. T2-weighted magnetic resonance image (MRI) in coronal projection demonstrates a simple fluid-filled cyst (C) in the right upper quadrant. (c) Type II. Coronal MRCP image shows diverticulum (arrow) of the common bile duct. (GB gallbladder). (d) Type III (choledochocele). Coronal MRCP image shows saccular dilatation (arrow) at the intraduodenal

portion of the common bile duct. (e) Type III (choledochocele). Coronal reformatted contrast-enhanced CT image demonstrating a cyst in the intraduodenal segment (arrow) of the common bile duct protruding into the duodenal lumen. (f) Type IVa. Coronal MRCP image demonstrates fusiform dilatation of the intrahepatic ducts (arrow) and common bile duct (CBD). (CD cystic duct). (g) Type V (Caroli disease). Coronal MRCP image shows innumerable intrahepatic cysts (arrows). (GB gallbladder)

into type Ia (diffuse) cysts, which involve the entire extrahepatic biliary ducts; type Ib (focal) cysts, which involve only a focal segment of the extrahepatic biliary ducts; and type Ic (fusiform) cysts, which involve only the common bile duct. Type I cysts result from an anomalous pancreaticobiliary union characterized by formation of a long, frequently ectatic common channel. Associated complications include biliary

obstruction, biliary cirrhosis, cyst rupture, portal vein thrombosis, hepatic abscess, and malignancy (Miyazaki et al. 2008). Type II choledochal cyst represents true diverticula of the extrahepatic biliary ducts. Type III choledochal cyst (or choledochocele) represents ectasia of an intramural common bile duct segment. These lesions are not embryologically related to the cystic dilatations of the other choledochal cysts.

Choledochoceles usually manifest in adults. The most common signs and symptoms are recurrent abdominal pain and jaundice. Cholangitis, pancreatitis, or nausea and vomiting from duodenal obstruction may also be seen. There is a high associated prevalence of choledocholithiasis. Type IV choledochal cysts are, by definition, multiple and can have both intrahepatic and extrahepatic components. They can be further subdivided into type IVa cysts, which involve both the extrahepatic biliary ducts and the intrahepatic bile ducts, and type IVb cysts, which involve only the extrahepatic biliary ducts with multiple saccular dilatations. Type V choledochal cyst (Caroli disease) is a rare congenital cystic dilatation of the intrahepatic bile ducts. It is an autosomal recessive disorder that results from the arrest of or a derangement in the normal embryologic remodeling of ducts, which in turn causes varying degrees of destructive inflammation and segmental dilatation. Imaging studies show intrahepatic saccular or fusiform dilated cystic structures of varying sizes that communicate with the biliary tree. Caroli disease usually manifests in children or young adults with recurrent bouts of cholangitis due to bile stasis, biliary calculi, recurrent attacks of right upper quadrant pain, fever, and, more rarely, jaundice.

Gallbladder

The gallbladder anomalies may be divided into three groups based on formation, number, and position (See Table 79.1).

Phrygian cap is the most common of the gallbladder anomalies reported to be present in up to 18% of patients with a functioning gallbladder (Figs. 79.11 and 79.12) (Boyden 1935). An infolding of a septum between the body and the fundus creates the Phrygian cap deformity. Phrygian cap has no clinical significance and occurs in individuals of all ages and, more commonly, in women. *Bilobed gallbladder* is a rare anomaly of formation, consisting of a completely divided gallbladder drained by a common cystic duct. Alterations in the contour of the gallbladder may result in a dumbbell or *hourglass gallbladder* deformity. In adults, this abnormality usually results from chronic cholecystitis and should be removed. Congenital *diverticulum* of the gallbladder is rare. Diverticula may occur in any part of the gallbladder and may vary

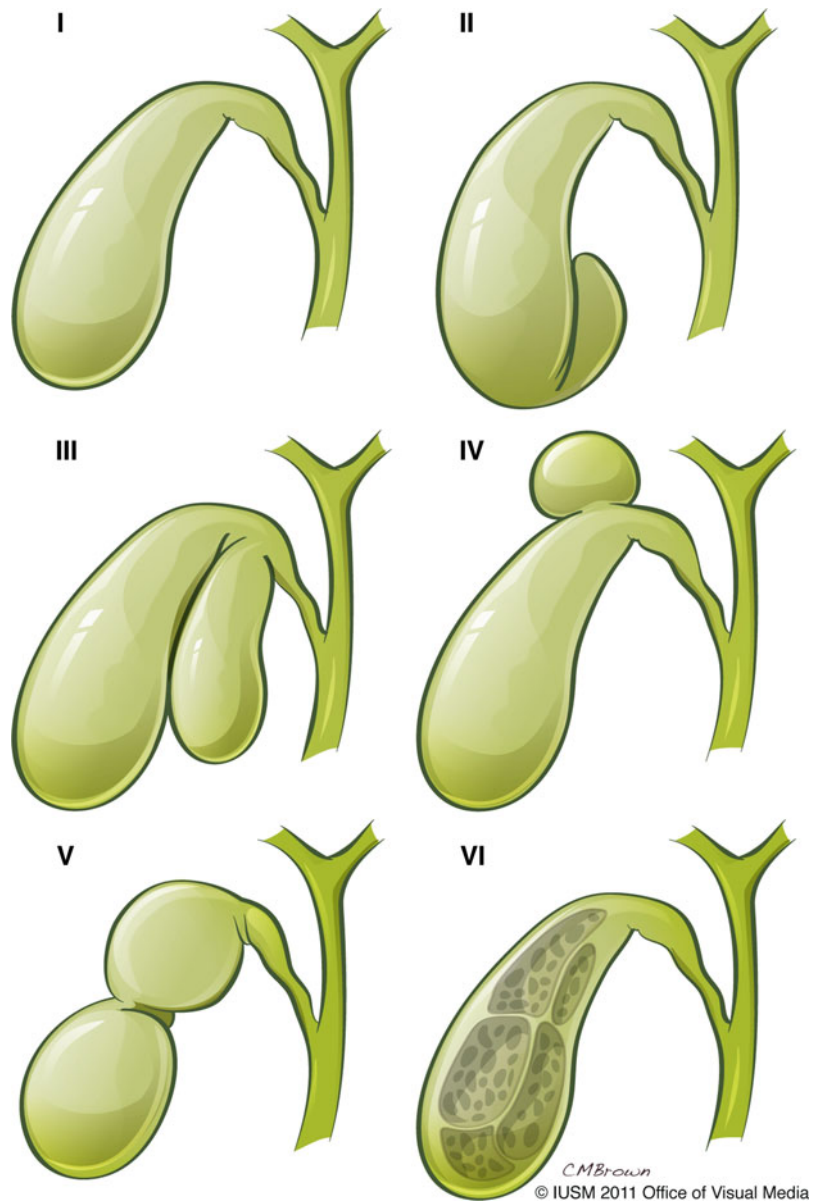
Table 79.1 Anomalies of the gallbladder

Formation
Phrygian cap
Bilobed gallbladder
Hourglass gallbladder
Diverticulum of the gallbladder
Septated gallbladder
Rudimentary gallbladder
Number
Absence of the gallbladder (agenesis)
Duplication of the gallbladder
Position
Floating gallbladder
Intrahepatic gallbladder
Left-sided gallbladder
Transverse gallbladder
Retrodisplaced gallbladder

greatly in size from 0.5 to 9 cm in diameter. In infants and children, a *rudimentary gallbladder* is believed to be due to congenital hypoplasia. In an elderly person, this situation may be the result of fibrosis from cholecystitis. More than 200 cases of *absence of the gallbladder* (agenesis) have been reported. Most cases are associated with other biliary abnormalities. *Duplicated gallbladder* has two separate cavities, each drained by its own cystic duct and sometimes supplied by its own cystic artery. Duplication of the gallbladder is clinically unimportant and generally requires no treatment.

Rarely, a gallbladder may be found in an abnormal location. This type of gallbladder requires no treatment unless it causes symptoms. Five different conditions are recognized: *floating*, *intrahepatic*, *left-sided*, *transverse*, and *retrodisplaced*. A floating gallbladder has been reported to occur in approximately 5% of persons. In this condition, the gallbladder is completely surrounded by peritoneum and is attached to the under-surface of the cystic fossa by the peritoneal reflection from the liver. Floating gallbladder may undergo torsion and require surgical intervention. An intrahepatic gallbladder is partially or completely embedded within the substance of the liver. In adults, approximately 60% of intrahepatic gallbladders are associated with gallstones.

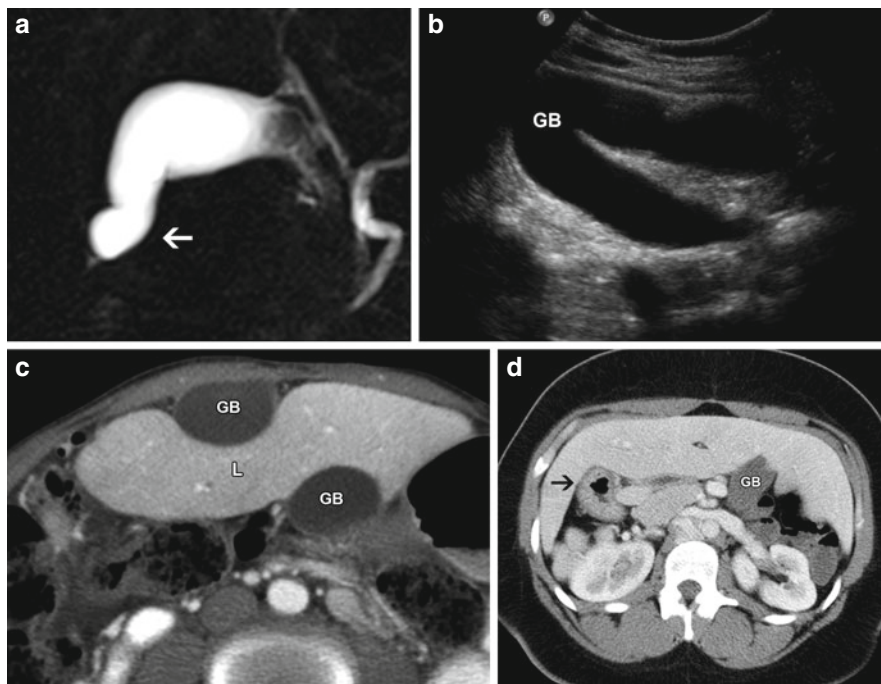
Fig. 79.11 Gallbladder formation anomalies. *I* normal, *II* phrygian cap, *III* bilobed gallbladder, *IV* gallbladder diverticulum, *V* hourglass gallbladder, *VI* septated gallbladder



Pearls to Remember

- Biliary tract, gallbladder, liver, and pancreas are closely associated with each other during embryologic development.
- It is important to know the variations in the biliary ducts because failure to recognize anatomic variations may result in significant complications such as bile leak following gallbladder or liver surgeries.
- Choledochal cysts are associated with several complications that can manifest clinically from infantile to adult ages. Type I choledochal cysts are associated with increased risk of developing malignancy.

Fig. 79.12 Gallbladder anomalies. (a) Phrygian cap. Coronal MRCP image shows deformity (*arrow*) in the fundus of the gallbladder. (b) Bilobed gallbladder. Ultrasound image demonstrates a divided lumen of the gallbladder (*GB*). (c) Duplicated gallbladder. Axial contrast-enhanced CT shows two gallbladders (*GB*) located anterior and posterior to the left lobe of the liver (*L*). (d) Left-sided gallbladder. Axial contrast-enhanced CT image of the 27-year-old patient with heterotaxy syndrome shows gallbladder (*GB*) located on the left side of the abdomen. *Arrow* points to the lumen of the stomach, which is located on the right side



- Anomalous junction of pancreaticobiliary ductal system (AJPBDS) increases the risk of developing biliary or gallbladder malignancy and associated with several other complications.

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