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Case Report

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Congenital Common Bile Duct Agenesis: An Extremely Rare Anomaly - Two Case Reports and Review of Literature

Louis F Chai[1], Alexander E Trebelev[2], Gary S Xiao[3*]

Abstract

Variations of the hepatico-pancreatic-biliary system are frequently vascular, but biliary duct deviations can occur.1 In the extra-hepatic biliary system, anomalies occur during foregut development and include accessory ducts, anomalous insertions, or agenesis. Though anomalies may be clinically silent, discovery usually occurs in symptomatic patients resulting in imaging or intraoperatively during exploration. The rarest of anomalies is common bile duct agenesis, resulting in formation of a cholecystohepatic duct, gallbladder interposition, or perhaps most appropriately, a hepaticocystic duct.2-3 We present here two cases discovered intraoperatively and an updated review on this anomaly.

Keywords: Hepaticocystic duct; Common bile duct agenesis; Extra-hepatic biliary tree; Cholecystectomy

Introduction

Biliary tree abnormalities and variations on the anatomy have been described in many forums, with studies showing incidences as low as 0.58% and as high as 47.2% of patients studied [1-4]. These variations are of particular importance during surgical interventions as ligation or excision of improperly identified structures around the area can cause significant morbidity and mortality when undiagnosed. These anomalies can occur anywhere along the biliary tree, but the least commonly seen is the agenesis of the common bile duct, resulting in the formation of a hepaticocystic duct. In this variation, the hepatic ducts, either individually or conjoined, directly enter in various locations of the gallbladder and the cystic duct directly flows into the duodenum [5]. This poses particular concern during cholecystectomies should the interposition be unnoticed, resulting in persistent proximal leakage of bile from the channels.

Case 1

The patient is a 32-year-old Asian male who presented as an outpatient for evaluation of repeated epigastric and RUQ abdominal discomfort. The patient had no significant past medical or surgical history. Original imaging from several years prior showed a small stone in the gallbladder and he had been having intermittent abdominal pain for 2 years. Work-up of the symptoms at presentation included a RUQ ultrasound that revealed a 1.9cm calcified stone in the fundus of the gallbladder, causing localized wall thickening, and a 3mm channel that was believed to be the common bile duct. The remainder of the exam was insignificant for any other pathology in the visualized hepato-pancreatico-biliary system (Figure 1). The patient was scheduled for elective laparoscopic cholecystectomy given the significant increase in size of the

Affiliation:

[1]Department of Surgery, Rutgers Robert Wood Johnson Medical School, Robert Wood Johnson Place, New Brunswick, NJ 08901

[2]Quantum Imaging & Therapeutic Associates, 629 D Lowther Road Lewisberry, PA 17339

[3]Transplant and Hepato-Pancreato-Biliary Surgery, Tower Health Transplant Institute, Reading Hospital. 301 South Seventh Avenue, Suite 320, West Reading, PA 19611

***Corresponding author:**
Gary Xiao, MD, FACS. Transplant and Hepato-Pancreato-Biliary Surgery, Tower Health Transplant Institute, Reading Hospital, 301 South Seventh Avenue, Suite 320, West Reading, PA 19611, USA

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gallstone and with the diagnosis of symptomatic cholelithiasis. The patient initially underwent the conventional 4-port cholecystectomy without any complication. After careful dissection in the area of the gallbladder neck, Calot's Triangle was exposed, the critical view of safety was obtained, and what was thought to be the cystic duct was identified. Clips were placed proximal to the gallbladder neck and further distal along the cystic duct. The cystic artery was similarly clipped and both isolated structures were divided between the clips. The gallbladder was then dissected off of the liver bed without any complication until transection of the tissues around the scarred and fibrosed fundus to completely remove the gallbladder revealed a bile leak from a 4 to 5mm duct-like structure in this area. Given the leak, a clip was temporarily placed on the structure and the gall bladder was removed to allow for further examination of the anatomy and to perform an intraoperative laparoscopic cholangiogram. The clips were removed from the remnant cystic duct stump and the cholangiogram catheter was placed into the cystic duct. When contrast was injected into the duct, it went directly into the duodenum. The cholangiogram was then repeated after withdrawing the catheter to be certain that the tip was not too deep within the common bile duct. The results were the same and no proximal reflux was seen into a common bile duct even after morphine was given to contract the sphincter of Oddi. At this time, with unclear anatomy, the procedure was converted from a laparoscopic to open case in order to properly explore the porta hepatis via a right subcostal Kocher incision. The portal vein and hepatic artery were identified with no visualized common bile duct. A biliary probe was used on the proximal and distal stumps from the transected ductal structures with the proximal stump reaching the left and right intrahepatic ducts and the distal duct tracing directly into the duodenum. A biliary catheter was then inserted into the proximal duct to perform an intra-operative cholangiogram, which showed a duct stump 5 millimeters below the bifurcation of the right and left intra-hepatic ducts and a normal intra-hepatic biliary tree. This indicated that the most likely anatomy was a hepaticocystic duct secondary to agenesis of the common bile duct. The distance between the proximal and distal stump measured approximately 3cm. A retrocolic, Roux-en-Y hepaticojejunostomy was performed to repair the transected proximal hepatic duct. The patient remained stable throughout the entirety of the case and was managed on the surgical floor post-operatively. The patient recovered well, was able to tolerate a regular diet, and was discharged to home with normal liver function. On subsequent outpatient follow-up, the patient was fully recovered from the surgery and returned to his daily activities. He continues to be well four years post-operatively.

Case 2

A 29-year-old male presented to an outside hospital for evaluation for abdominal pain and was diagnosed with acute cholecystitis. The patient was taken to the operating room nine days prior to presentation at our hospital for a laparoscopic cholecystectomy. Though the dissection was difficult, the case proceeded uneventfully and the patient was subsequently discharged post-operatively. The patient returned to the outside hospital on post-operative day 7 with high fevers and had a collection seen on ultrasonography, which was then identified as a bile duct leak on HIDA scan. An ERCP was performed at this time, but the biliary duct could not be cannulated due to an obstruction. The patient was brought back to the operating at this point for an exploratory laparotomy. After exploration, the surgeons identified a bile leak, but could not identify the proximal bile duct. Intra-operative cholangiogram was performed through the distal duct, which revealed flow into the duodenum, but the proximal duct could not be identified. Given the complexity of the anatomy and the case, the patient was transferred to us for further surgical management. On arrival, the intraoperative cholangiogram from the outside hospital was reviewed and a diagnosis of congenital agenesis of the common bile duct as suspected as flow through the ligated cystic duct went directly into the duodenum. At this time, Interventional Radiology (IR) was consulted to perform at PTC to review the biliary anatomy and create a road map for further repair. However, only the right hepatic system could be catheterized as the left hepatic duct was too small (Figure 2a). The cholangiogram through the right PTC showed a large bile leak, but no extra-hepatic ducts could be identified at all. Given these findings, the patient was then taken to the operating room for exploratory laparotomy. Upon examining the surgical site, the distal bile duct was significantly dilated to 9-10 millimeters in diameter. Proximally, the right PTC catheter placed by IR was emerging from the intra-hepatic right biliary system and further exploration did not reveal any intra-hepatic biliary ducts from the left or any extra-

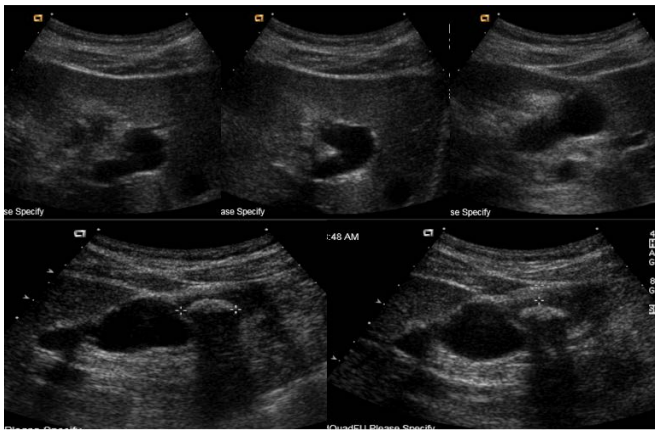

Figure 1: Pre-operative right upper quadrant ultrasound for Case 1 revealing dilated biliary ducts with a large gallstone within the gall bladder. No other abnormalities were detected on imaging.

Chai LF, et al., J Surg Res 2026
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hepatic ducts. A complete portahepatis lymphadenectomy and dissection with skeletonized hepatic artery and portal vein was performed in an attempt to localize the left intra-hepatic duct and right extra-hepatic duct to no avail. It was suspected that the patient had variant anatomy with common bile duct agenesis with the left hepatic duct connecting to the right hepatic duct high in the liver parenchyma. Given this, continuity was restored to the biliary tree via a hepaticoportal enterotomy, or Kasai procedure. The tissue around the porta hepatis and the right PTC wrapped around an enterotomy in a two layer anastomosis. The patient was managed for the first three post-operative days in the intensive care unit before being transferred to the surgical floors where his recovery continued before ultimately being discharged. Subsequent follow-up appointments revealed adequate patency of the right-sided biliary catheters and ultimately, dilation of the left sided biliary system to permit cannulation and drainage into the intact hepaticoportal enterostomy (Figure 2b and 2c). Clinically, the patient remains stable and with unremarkable laboratory results 18 months from surgery.

Discussion

The biliary tree has embryological roots as the most caudal portion of the foregut and arises from what is known as the hepatic diverticulum. This divides into two parts, the liver

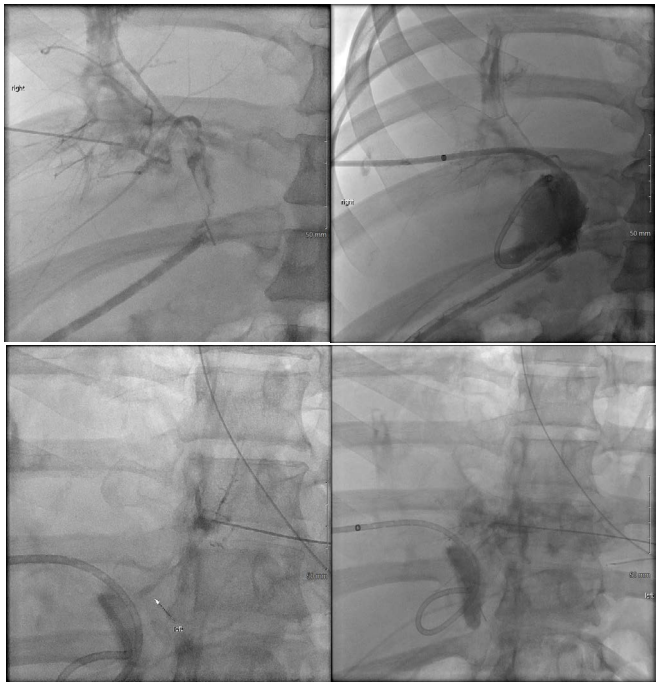

Figure 2a: Initial cholangiography on presentation on 5/14/2017 for Case 2. From top left clockwise: cholangiography showing intact right hepatic duct system with distal extravasation of contrast at ligated common hepatic duct, insertion of percutaneous trans-hepatic catheter with flow into the jejunal limb, collapsed and inaccessible distal left hepatic duct system, and collapsed and inaccessible proximal left hepatic duct system.

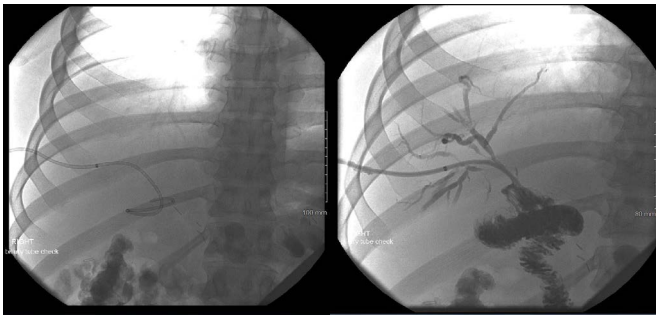

Figure 2b: Drain check performed on 6/14/2017 for Case 2. Left image with intact catheter in the right hepatic system and right image showing flow into the jejunal limb. Attempts were made to access the left hepatic system again, but was inaccessible once again.

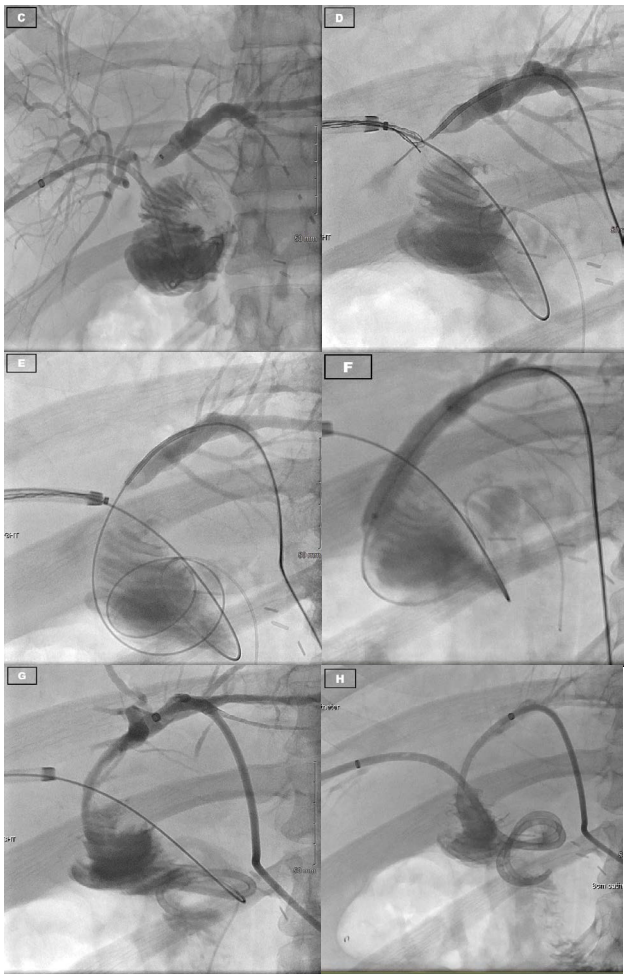

Figure 2c: Follow up cholangiography several months after surgery for Case 2. Pictures as labeled: A - Dilated left hepatic duct system, B - catheter passing through severely stenotic region of left hepatic duct, unable to float into common hepatic duct, C - right hepatic duct cholangiogram with flow into jejunal limb, D - Removal of right PTC catheter over wire, insertion of left hepatic duct wire, snare of left hepatic duct wire through right hepatic duct, E - left hepatic duct wire dragged through common hepatic duct into duodenum via right hepatic duct snare, F - balloon dilation of stenotic area of left hepatic duct, G - insertion of PTC catheter into left hepatic duct, H - reinsertion of right PTC catheter.

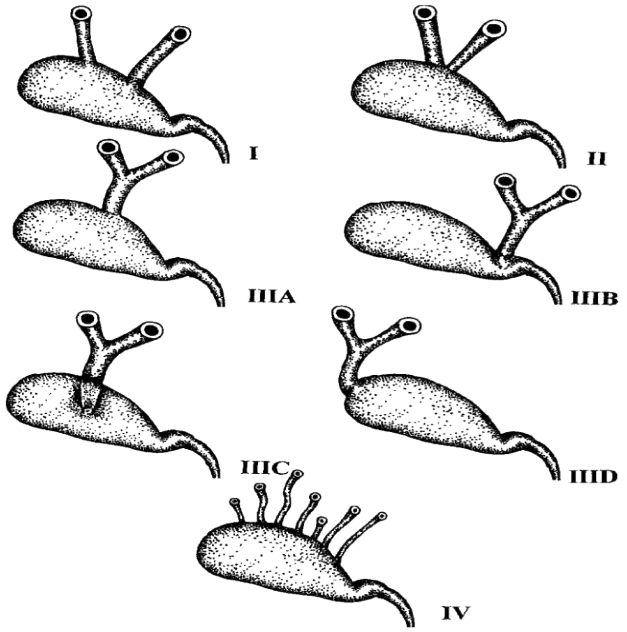

Figure 3: Hepaticocystic duct variants as first described by Losanoff et al.

and gallbladder, with the cells in between developing into the cystic and bile ducts to form the fully developed biliary tree that enters the duodenum [6]. The variations in anatomy arise during this stage of development and can arise in any of the involved structures. In terms of the hepaticocystic duct, it is believed that the aberrancy occurs secondary to delayed separation of the hepatic and cystic tissues, persistence of fetal communications, or failure of the cells of the common bile duct to proliferate or recannalize [1,5]. Thus, the gallbladder and cystic duct are interpositioned between the hepatic ducts and the duodenum as the only common channel for biliary outflow. With regards to biliary tree aberrancies, the hepaticocystic duct appears to be the rarest of all anomalies, which can include duplicated structures, anomalous insertions, and congenital agenesis. In a review of the literature, there have only been 25 reports of varying detail of this developmental abnormality in the world as far as we are aware (Table 1) [7-24]. Of these cases, multiple variants have been described with different insertions of the hepatic ducts into the gallbladder with the common link between all of them being the agenesis of the CBD and the solitary outflow of the entire biliary track being provided by the cystic duct into the duodenum [25-31]. This classification system was first described by Losanoff et al. and consists of four types (Figure 3). In Type I, each hepatic duct inserts into the gallbladder separately. Type II results in the joining of the hepatic ducts when they enter the gallbladder. Type III has multiple variants based on where the insertion of the common hepatic duct enters the gallbladder. Type IIIa inserts in the superior portion of the gallbladder wall, IIIb into the neck of the gallbladder, IIIc into the posterior wall of the gallbladder, and IIId into the gallbladder fundus. The type IV variant has multiple individual ducts draining the liver inserting into the wall of the gallbladder [3]. In our first patient, we observed the Type IIId variant of the hepaticocystic duct whereas the second did not appear to match any of Losanoff's classifications. Presentation of extra-hepatic biliary tree abnormalities can be hard to diagnose and distinguish pre-operatively from normal anatomy. Often, pathology presents with symptoms of RUQ pain, jaundice, nausea, vomiting or fevers with laboratory findings of leukocytosis or elevated liver enzymes, all of which may be due to cholelithiasis, choledocholithiasis, cholangitis, or hepatic pathology [3,5,15,16,20-24]. Work-up may include RUQ ultrasounds, computed tomography (CT), magnetic resonance imaging (MRI), or radioactive nucleotide studies such as the HIDA scan. Of these, abnormalities would most likely be discovered on HIDA as flow of the nucleotide is observed through the biliary tree, though CT or MRI may also reveal anatomic variations. However, workup is usually limited to RUQ ultrasound in the correct clinical setting for gallbladder disease before operative management may be indicated, which is what occurred in our two cases [17,19]. Thus, diagnoses of biliary tree aberrancies are generally not obtained until operative exploration or intraoperative cholangiogram for unclear anatomy [3,20,25]. Our first patient's anatomy was initially noted when there was persistent bile leak noted from a duct near the fundus of the gallbladder and subsequent porta hepatis exploration and intraoperative cholangiogram. Imaging highlighting the proximal left and right hepatic duct and the transected cystic duct leading to the duodenum from Case 2 is shown in Figure 2a-2c. Consequences of an unrecognized biliary duct abnormality could be catastrophic, leading to significant morbidity and mortality. With anomalous hepatic ducts inserting at various locations on the gallbladder, ligation is essentially guaranteed while performing a cholecystectomy, especially if the surgeon is unaware of their presence [26]. While small caliber ducts may be ligated without any complications, accessory ducts with large lumens can cause significant leakage of bile, infection, or poor nutrient absorption. Severe consequences include formation of sub-phrenic abscesses, cirrhosis, external biliary fistulas, frank peritonitis, and even death [4,6,21,27]. When hepaticocystic ducts are diagnosed, various management strategies can be employed depending on when duct anatomy is discovered. When identified prior to removal of the gallbladder, a planned partial cholecystectomy could be performed with a choledochoplasty to preserve the biliary outflow into the duodenum through to remnant cystic duct [15,19-20,22]. If the cholecystectomy has already been completed, leaving behind a one or more proximal hepatic duct stumps and the distal cystic duct stump, primary anastomosis can be performed or a Roux-en-Y hepaticojejunostomy can be used to achieve adequate elimination of bile [5,15,19,21]. Primary anastomosis should only be attempted if is not a significant

Chai LF, et al., J Surg Res 2026
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defect resulting from the cholecystectomy, if there will be anastamotic tension, or if there will not be high risk of biliary stricture [16,19,21]. The distance between our hepatic stump and cystic duct stump after cholecystectomy in Case 1 was approximately 3cm and had too large of a defect to perform a primary anastomosis. No discernible left hepatic duct could be found radiographically or on surgical exploration in Case 2. Thus, we opted for a Roux-en-Y hepaticojejunostomy to re-establish continuity to the biliary tree in Case 1 and a hepaticoporto enterostomy in Case 2 given the anatomical remnants post-cholecystectomy in each of the patients. The patients did not experience any complications from the surgeries and both remain well post-operatively.

Table 1: Known reported cases of common bile duct agenesis and hepaticocystic ducts [1,2,5,7-24, 28-31].

Case Report	Symptoms	Variant	Diagnosis	Management
Abeysuriya et. al.	N/A	IIIb	Autopsy	N/A
Aristotle et. al.	N/A	IIIb	Autopsy	N/A
Chiavola et. al.	Unknown	Unknown	Unknown	Unknown
Crucknell HH.	Unknown	IIIa	Unknown	Unknown
Elhamel A.	RUQ pain, jaundice	I	Open exploration, intraoperative cholangiogram	Partial cholecystectomy, choledochoplasty
Harada et al.	Epigastric pain	IIIb	Open exploration, intraoperative cholangiogram	Partial cholecystectomy, choledochoplasty
Hashmonai et al.	Asymptomatic	IIId	Open exploration, post-operative cholangiogram	Partial cholecystectomy, choledochoplasty
Jackson et al.	Epigastric pain, jaundice	I	Open exploration, post-operative cholangiogram	Partial cholecystectomy, choledochoplasty
Kaushik et al.	Symptomatic cholelithiasis	I or IIIb? (separate entrance into neck)	Open exploration, intraoperative cholangiogram	Partial cholecystectomy
Krishna et al.	Epigastric pain	IIIb	Open exploration, intraoperative cholangiogram	Roux-en-Y hepaticojejunostomy
Losanoff et al.	RUQ pain	IIId	Open exploration	Roux-en-Y hepaticojejunostomy
Markle GB	RUQ pain, nausea, vomiting	III?	Open exploration	Roux-en-Y hepaticojejunostomy
Mittal et al.	Abdominal pain	IIIa or IIIc	Open exploration, intraoperative cholangiogram	Roux-en-Y hepaticojejunostomy
Moosman et al.	Unknown	I	Unknown	Unknown
Nikolov et al.	Unknown	I	Unknown	Unknown
Nygren et al.	Unknown	IIIb	Unknown	Unknown
Olsha et al.	Unknown	IIIa	Unknown	Unknown
Rabinovitch et al.	Abdominal pain, nausea, vomiting, fevers	I (variation - R. duct only into GB), separate into duodenum	Open exploration	Partial cholecystectomy, choledochoplasty
Redkar et al.	RUQ pain, jaundice	IIIb	Open exploration, intraoperative cholangiogram	Roux-en-Y hepaticojejunostomy
Schorlemmer et al.	Jaundice	IV	Open exploration, intraoperative cholangiogram	Cholecystoduodenostomy
Shah et al.	Cholangitis	IIIb	Open exploration, intraoperative cholangiogram	Partial cholecystectomy, choledochoplasty
Stokes et al.	RUQ pain, jaundice	IIIc	Open exploration, intraoperative cholangiogram	Biliary tree reconstruction via Y-tube
Walia et al.	Unknown	I	Unknown	Unknown
Williams et al.	RUQ abdominal pain, jaundice	I	Open exploration	Primary anastomosis and dilation
Zimmerman et al.	Unknown	IIIa	Unknown	Primary anastomosis

Conclusion

Aberrancies of the biliary tree are uncommon findings, with a wide variety of incidences quoted by different reviews and case series. However, with cholecystectomies being one of the most common surgical procedures performed, these abnormalities have operative significance and correct identification is essential in preventing complications that are potentially fatal. Though conventional work-up with RUQ ultrasound may not identify such anatomical variations, proper exploration and identification of the critical view of safety intra-operatively during a cholecystectomy may prevent these complications. If there is doubt about the anatomy during the procedure, a cholangiogram should be performed to better delineate structures. This would allow for early identification before the gallbladder resection, potentially sparing the patient from a larger operation to create a Roux-en-Y hepaticojejunostomy for biliary continuity and could instead be managed with primary anastomosis or choledochoplasty after partial cholecystectomy. Though rare, when abberant anatomy is discovered, a hepaticocystic duct should be considered as an anatomical variation for proper diagnosis and management in biliary tree pathology perioperatively.

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