Chapter 24
Laparoscopic Surgery for Congenital Biliary Dilatation in Children

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Abstract The treatment of choice for congenital biliary dilatation is complete excision with Roux-en-Y hepaticojejunostomy, a procedure that is now being performed laparoscopically. We describe our technique (laparoscopic dilated bile duct dissection, distal common bile duct ligation, intrahepatic bile duct and common channel protein plug clearance, and customizing the length of the Roux-en-Y loop) and discuss attendant issues from our wealth of experience of treating this condition using both open and minimally invasive surgery.

In summary, laparoscopic excision of the bile duct and Roux-en-Y hepaticojejunostomy is feasible and safe, associated with lower postoperative morbidity and less blood loss, in the hands of experts. With continued advancement in technology and improvement in surgical skills with experience, it is only a matter of time before minimally invasive surgery becomes the mode of choice for treating congenital biliary dilatation.

Keywords Congenital biliary dilatation · Hepaticojejunostomy · Laparoscopy · Children

24.1 Introduction

Minimally invasive surgery has gained acceptance for treating pediatric congenital biliary dilatation because of comparative advantages over more conventional open surgery, such as, better cosmesis, less requirement for analgesia, a more rapid return to baseline functional status, quicker rehabilitation, and less likelihood of complications secondary to postoperative adhesions. Surgical intervention for congenital biliary dilatation necessitates an exhaustive understanding of anatomic variations centered on the porta hepatis and mastery of skills appropriate for dealing them.

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In 1995, Ferallo et al. first described laparoscopic resection and Roux-en-Y hepaticojejunostomy in a 6-year-old case with congenital biliary dilatation [1]. Although their technique has been modified by the advent of finer instruments and the adoption of novel maneuvers, it is the standard viable option for treating congenital biliary dilatation using minimally invasive surgery [2, 3].

The authors will introduce their laparoscopic technique for dilated bile duct excision. A distinguishing feature of their technique is intraoperative endoscopy of the common channel and intrahepatic bile ducts that is performed routinely to examine for biliary debris/stones and protein plugs that are cleared by irrigation with normal saline to prevent mid- to long-term postoperative cholangitis, pancreatitis, and stone formation [4–6].

24.2 Surgical Technique

24.2.1 Laparoscopic Excision

24.2.1.1 Patient/Port Positioning and Initial Preparation

Under general anesthesia, the patient is placed in the reverse Trendelenburg position. A GelPOINT® mini advanced access platform. (Applied Medical, Rancho Santa Margarita, USA) inserted in a 2 cm umbilical incision is used to introduce a 30° 5 or 10 mm laparoscope into the abdomen. Pneumoperitoneum is established with CO_2 insufflated at a flow rate of 0.5–1.0 L/min at a pressure of 8 mmHg, increasing to 12 mmHg if required. Two additional 5 mm trocars are inserted in the right upper quadrant and left upper quadrant, respectively (Fig. 24.1). A percutaneous stay suture is introduced just below the xiphoid process to snare the falciform ligament and retract/elevate the liver to improve exposure. A pair of Babcock forceps is inserted through the left subcostal port in the anterior axillary line to grasp and elevate the gallbladder to expose the porta hepatis and allow the dilated bile duct to be dissected free from surrounding structures, such as the portal vein and hepatic artery. Usually, there are more adhesions between a cystic bile duct and the portal vein and hepatic artery than with a fusiform bile duct, especially in older children.

In adolescents and adults, adhesions can be very dense and complicate dissection. When adhesions are extremely dense, an additional trocar inserted in the lateral right subcostal area can be used for an assistant to grasp the bile duct and facilitate safe dissection (Fig. 24.2). If the adhesion bile duct is inflamed and there are dense adhesions, the anterior wall of the bile duct can be incised at any time during bile duct dissection to allow the posterior wall of the bile duct to be dissected safely under direct vision.

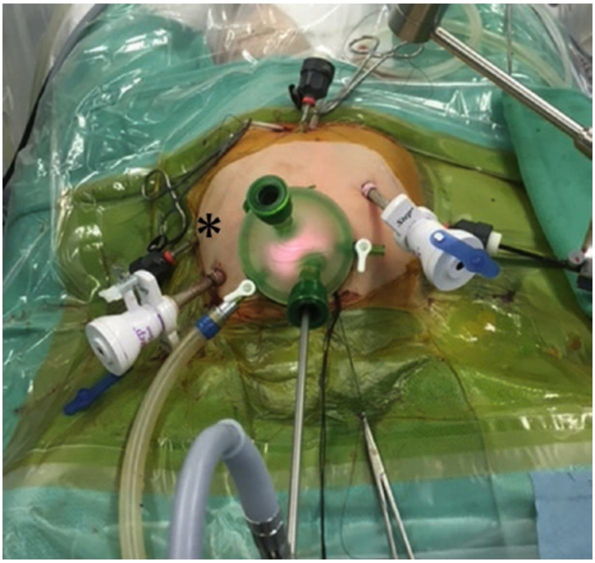

Fig. 24.1 Trocar positions. A 30° 5 or 10 mm laparoscope is introduced through a GelPOINT® mini advanced access platform. (Applied Medical, Rancho Santa Margarita, USA) inserted in a 2 cm umbilical incision. Two additional 5 mm trocars are inserted in the right upper quadrant and left upper quadrant, respectively, as working trocars. The left upper quadrant trocar (left subcostal trocar in the anterior axillary line) is inserted to expose the porta hepatis. An additional 3.9 mm trocar is placed in the left epigastrium for intraoperative endoscopy. Another additional 3.9 mm trocar (*asterisk*) may be placed in the lateral right subcostal area for an assistant to grasp the bile duct to facilitate safe dissection of the bile duct by the surgeon

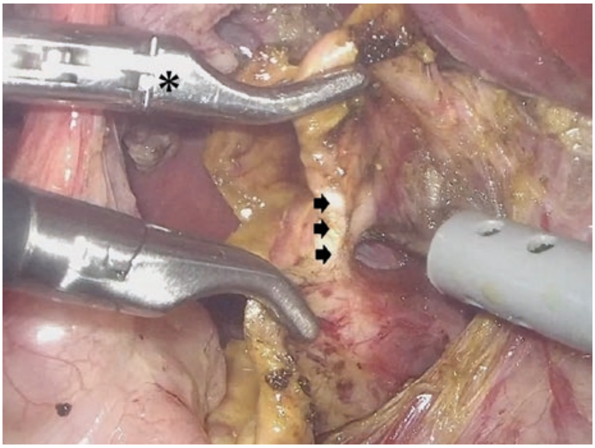

Fig. 24.2 Severely dense adhesions. If the adhesion is inflamed and there are dense adhesions, the anterior wall of the bile duct may be incised at any time during dissection to allow the posterior wall (*arrows*) of the bile duct to be dissected safely under direct vision. An additional trocar (*asterisk*) in the lateral right subcostal area may be placed for an assistant to grasp the bile duct during dissection

24.2.2 Intraoperative Cholangiography

Intraoperative cholangiography is performed if preoperative magnetic resonance cholangiopancreatography is not available or fails to delineate the anatomy of the hepatopancreaticobiliary ducts, especially the anatomy of the pancreaticobiliary junction, and the presence of debris or protein plugs in the intrahepatic bile ducts and common channel. Preoperative magnetic resonance cholangiopancreatography is the investigation of choice and is accurate in the majority of cases.

24.2.3 Intraoperative Endoscopy

For intraoperative endoscopy, an additional 3.9 mm trocar is inserted in the left epigastrium for the introduction of a fine pediatric ureteroscope [7]. We use a pediatric ureteroscope specifically because it allows normal saline to flow continuously through a dedicated side channel, allowing constant visualization and irrigation. While some surgeons suggest that laparoscopic examination is sufficient, we find that a constant flow of saline dilates the lumen to allow safe examination and assists in clearing debris and protein plugs. Without a constant flow of saline, the lumen collapses, greatly compromising both examination and clearing, and side channels on flexible scopes are essentially designed only for flushing and are inadequate for inspection and irrigation.

Intraoperative endoscopy is particularly valuable during excision of fusiform type congenital biliary dilatation to ensure that any wide intrapancreatic choledochus is excised adequately as any remnant may contribute to stone formation that may cause postoperative pancreatitis in the long-term. It is less important in cystic type congenital biliary dilatation, since the intrapancreatic choledochus is short and narrow and the patient does not often present with pancreatitis, probably because there are no debris in the common channel. Intraoperative endoscopy is performed routinely in all congenital biliary dilatation patients unless the ureteroscope cannot be inserted into the intrapancreatic choledochus and common channel from the distal part of the bile duct because they are too narrow.

24.2.4 Complete Excision

The cystic artery is identified and divided. Dissection of the dilated bile duct is initiated by removing the adjacent peritoneum using monopolar electrocautery and a Maryland dissector to establish a plane of dissection, beginning on the anterior/lateral wall and continuing to the distal sides and then to the posterior portion.

The exact level of transection of the distal common bile duct is determined by intraoperative endoscopy when the orifice of the pancreatic duct can be identified with the ureteroscope [8] and with intraoperative cholangiography if the orifice of the pancreatic duct cannot be identified.

During intraoperative endoscopy, the part of the ureteroscope emerging externally from the trocar is held with mosquito forceps and pulled very gently from the pancreatic duct to the level where the distal dissection was ceased under laparoscopic view (Fig. 24.3). Because there is a light source at the tip of the ureteroscope, the laparoscopic surgeon can measure the actual length of the intrapancreatic part of the dilated bile duct from the pancreatic duct orifice to the point where dissection was ceased. If the intrapancreatic part is longer than 5 mm, the distal end of the bile duct is dissected further caudally toward the intrapancreatic duct orifice. This procedure is repeated until the intrapancreatic part is 5 mm or less in length. To prevent erroneous measurement, an exteriorized silk suture is fixed by clamping with a pair of mosquito forceps to ensure constant tension is maintained on the bile duct. Thus, the laparoscopic surgeon can continue to dissect toward the common channel to excise the intrapancreatic part of the bile duct, confident there is no risk for injuring the pancreatic duct because the exact length of the intrapancreatic part of the bile duct is known. Once 5 mm or less in length, the intrapancreatic part is ligated and excised. Once the bile duct has been freed, the distal part is divided as close as pos-

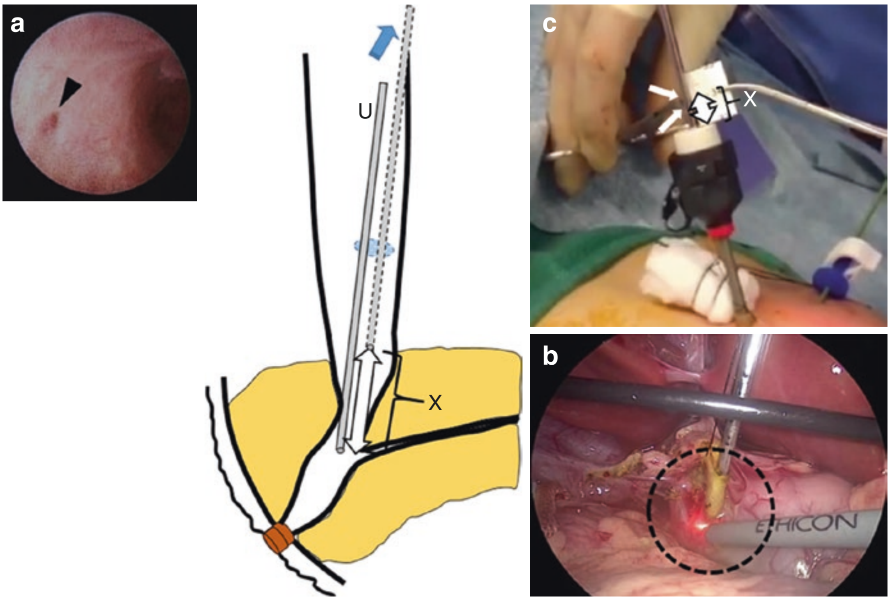

Fig. 24.3 Measuring the intrapancreatic part of the bile duct during laparoscopic excision. After opening the dilated bile duct distally, an ureteroscope (Diagram: U) is inserted to identify the orifice of the pancreatic duct (**a**: *arrowhead*). The part of the ureteroscope emerging externally from the trocar is held with mosquito forceps (**c**: *white arrows*) and pulled very gently (Diagram: *Blue arrow*) from the pancreatic duct to the level where distal dissection was ceased under laparoscopic view (**b**). The laparoscopic surgeon can measure the length X (Diagram and **c**: *double-headed white arrow*) which will be the length of the intrapancreatic part of the congenital biliary dilatation

sible to the pancreaticobiliary junction, and the stump is ligated with an endoloop. When the pancreatic duct orifice cannot be identified, intraoperative cholangiography may be performed by placing an endoscopic metal clip at the distal end of the dissected bile duct to indicate the extent of dissection required further distally, because the clip and the confluence between the common channel, intrapancreatic choledochus, and pancreatic duct can be visualized. If dissection is inadequate, the bile duct can be dissected further distally and intraoperative cholangiography repeated as above until bile duct dissection is considered adequate.

The proximal bile duct is excised leaving 10 mm of common hepatic bile duct for the hepaticojejunostomy. Should the anatomy be more complicated than expected, for example, if there is membranous stenosis in the common hepatic duct, conversion to mini laparotomy for open hepaticojejunostomy should be considered without hesitation.

24.2.5 Extracorporeal Transumbilical Jejunal Roux-en-Y

The ligament of Treitz is identified, and jejunum 15 cm distal to the ligament is exteriorized through the umbilical port site to create the Roux-en-Y jejunal loop extracorporeally. Pneumoperitoneum is interrupted, and the jejunum is divided, and the length of the Roux limb is customized by bringing it up to 1 cm above the xiphoid process on the anterior abdominal wall. Customizing ensures that a Roux limb will grow with the patient and not become tortuous as predetermined lengths of Roux limb (30, 40, or 50 cm) have a tendency to do, leading to stasis and risk for cholangitis. A jejunojejunostomy is performed extracorporeally. The customized Roux limb is approximated to the native jejunum for 8 cm cranially to prevent the contents of the native jejunum from refluxing into the Roux limb. The jejunojejunostomy should fit naturally into the splenic flexure after anastomosis [9]. Finally, an antimesenteric enterotomy is made near the closed end of the Roux limb, and the jejunum returned to the abdominal cavity; the pneumoperitoneum is reestablished; and the jejunal limb is passed through a retrocolic window to lie without tension at the porta hepatis. A scalpel should be used for the enterotomy in the jejunum to prevent thermal injury to the jejunal wall; we never use diathermy with coagulation mode for the enterotomy, since thermal injury can cause scarring [5, 6]. If the enterotomy is made slightly on the anterior side of the jejunum rather than the antimesenteric side, the hepaticojejunostomy is easier, because the mucosa of the posterior wall of the jejunum can be easily identified while performing the anastomosis.

24.2.6 Hepaticojejunostomy

From experience, an additional two ports (3.9/5 mm) are required for hepaticojejunostomy, one lateral right subcostal port and one between the right subcostal and right upper quadrant ports in order to prevent the quality of the anastomosis from deteriorating, especially when the diameter is less than 9 mm. End-to-side hepaticojejunostomy is performed using interrupted 5/0 or 6/0 absorbable sutures with the right upper quadrant port as the needle holder in the right hand, the 5 mm port for the scope, and the 3 mm subcostal port as the needle receiver in the left hand (Fig. 24.4a, b). Both the right and left edge sutures are exteriorized and used as traction sutures during anastomosis of the anterior wall to facilitate accuracy, especially when the hepaticojejunostomy anastomosis diameter is less than 9 mm.

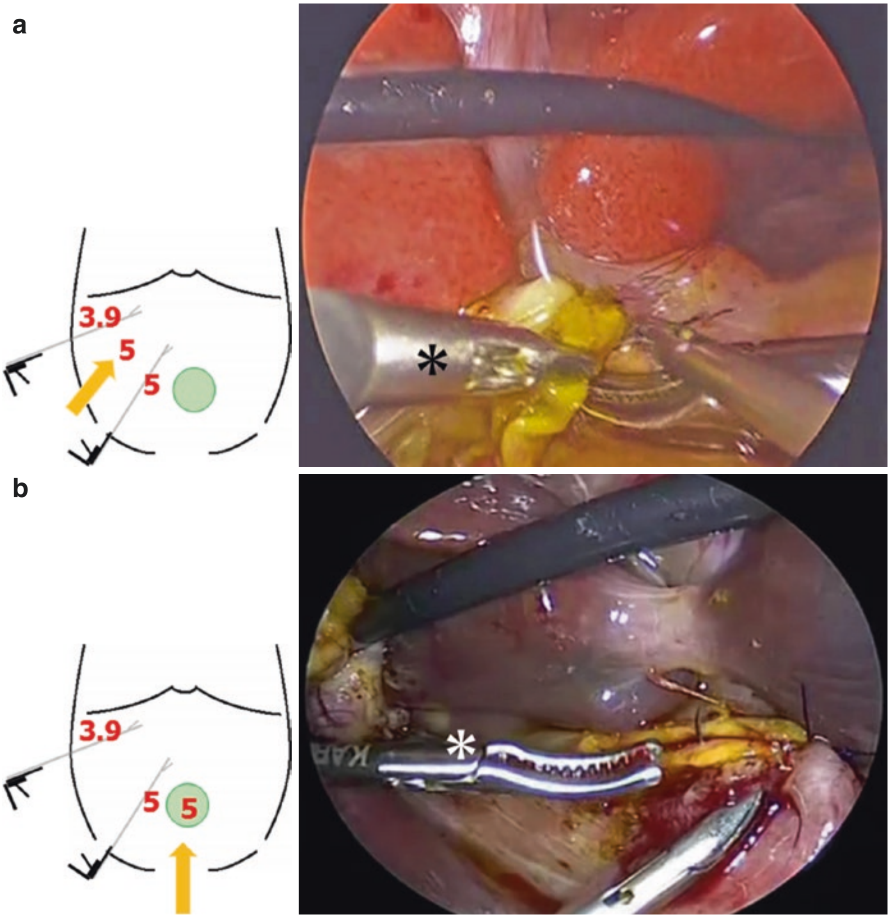

Fig. 24.4 End-to-side hepaticojejunostomy. (**a**) Coaxial ergonomics: hepaticojejunostomy is performed using interrupted 5/0 or 6/0 absorbable sutures with the right upper quadrant port as a needle holder in the right hand, the 5 mm port for the scope (Diagram: *orange arrow*) and the 3.9 mm subcostal port (*asterisk*) as a needle receiver in the left hand. The green circle is the GelPOINT® mini advanced access platform. (**b**) Paraaxial ergonomics: hepaticojejunostomy is performed with the right upper quadrant port as a needle holder in the right hand and the 3.9 mm subcostal port (*asterisk*) as a needle receiver in the left hand. The scope (Diagram: *orange arrow*) has been moved to the GelPOINT® mini advanced access platform.

A tube drain is inserted in the foramen of Winslow. The resected bile duct and gallbladder are extracted through the umbilical port site. The trocars are removed and the wounds closed.

24.2.7 Single-Incision Laparoscopic Excision

First reported in 2012, single-incision laparoscopic surgery for congenital biliary dilatation involves placing all trocars at a single site with a single skin incision instead of multiple skin incisions at separate sites as in conventional laparoscopic surgery [10]. In general, it is more difficult than conventional laparoscopic surgery due to problems related to triangulation and instrument manipulation. To facilitate single-incision laparoscopic surgery, specialized multichannel ports and/or specialized laparoscopic instruments with curved bent tips or rotating mechanisms are used. To date, some 260 cases of successful single-incision laparoscopic surgery for congenital biliary dilatation have been reported [11–13]. Compared with conventional laparoscopic surgery, single-incision laparoscopic surgery would appear to be more efficient based on operative time and postoperative recovery and have better cosmesis [14], although longer follow-up is still needed. Single-incision requires mastery of both laparoscopic and hepaticobiliary surgery to manage potential complications. Such expertise is proportional to experience obtained from performing a large number of conventional laparoscopic hepaticojejunostomies as a team to foster technical proficiency and establish rapport between surgeons and assistants.

24.2.8 Robotic-Assisted Excision

Robotic surgery has been in use for the treatment of congenital biliary dilatation since 2006 [15]. Its advantages include superb visualization and instrument control. The camera provides high magnification with three-dimensional visualization through a stereo endoscope, and the surgeon is assisted by features such as direct control of the visual field, improved dexterity, tremor reduction, motion scaling, and higher degrees of freedom compared with standard laparoscopic instruments. These advantages make dissecting, suturing, and knot-tying easier, and the hepaticojejunostomy anastomosis is facilitated greatly by robotic assistance. For example, the authors recently treated a pediatric case of pancreaticobiliary malunion without dilatation of the common hepatic duct successfully using da Vinci robotic assistance for the hepaticojejunostomy anastomosis with a common hepatic duct only 4 mm in diameter. Such a procedure would have been quite difficult using conventional laparoscopy.

The potential promise of robotic-assisted surgery is that it can encourage surgeons to be more ambitious when planning and performing complex minimally invasive procedures. To date, some 45 pediatric congenital biliary dilatation cases treated by robotic-assisted hepaticojejunostomy have been reported [15–19]. The mean operative time of these cases ranged from 180 to 520 min, significantly longer than for conventional laparoscopic procedures; bile leak was reported in one case. At present, the size of robotic hardware prevents it from being used more generally in pediatric surgery. The hardware is also expensive and maintenance costs are high. With ongoing technical improvements, robotic assistance will enable surgeons to approach the optimal goal of minimally invasive surgery, i.e., atraumatic, scarless treatment.

24.2.9 Laparoscopic Hepaticojejunostomy Versus Laparoscopic Hepaticoduodenostomy

Although hepaticoduodenostomy is an easier, quicker procedure and allows bile to enter the duodenum directly, which is more physiological [20–22], postoperative cholangitis and bile gastritis are known complications with risk for mucosal damage and possible malignant change. Todani et al. [20] reported a patient who underwent bile duct excision and hepaticoduodenostomy at 13 months old and developed hilar bile duct carcinoma 18 years later. Inflammation of the bile duct mucosa was thought to be related to the reflux of duodenal contents (including activated pancreatic enzymes) into the intrahepatic bile ducts though the anastomosis which prompted them to abandoned hepaticoduodenostomy in favor of hepaticojejunostomy. The authors also reported bilious gastritis due to marked duodenogastric bile reflux on upper gastrointestinal endoscopy, and histology of biopsied gastric mucosa showed gastritis [23].

Overall, hepaticojejunostomy is recommended for biliary reconstruction in children requiring congenital biliary dilatation excision, because hepaticoduodenostomy is associated with some degree of duodenal contents reflux into the biliary tree, especially when intrahepatic bile duct dilatation is present. A long-term prospective randomized controlled study is warranted to compare the outcomes of laparoscopic hepaticoduodenostomy and hepaticojejunostomy.

24.2.10 Surgical Outcome

In mid- to long-term follow-up studies published recently [24, 25], experienced laparoscopic surgeons were reported to be able to achieve results similar to open surgery. In a report comparing laparoscopic bile duct excision with open surgery in children [22, 25], the operative time was found to be longer and overall costs higher, but there was significantly less blood loss and duration of hospitalization was shorter. There were no significant differences in the incidences of bile leaks or wound infections. This would appear to suggest that in the hands of skilled laparoscopic surgeons, laparoscopic bile duct excision and Roux-en-Y reconstruction are safe and effective.

24.3 The Authors' Experience

The authors performed 43 laparoscopic congenital biliary dilatation excisions between 2009 and 2017. Cases requiring conversion to open laparotomy ($n = 1$) and minilaparotomy ($n = 2$) were excluded, leaving 40 cases, 32 females and 8 males. Congenital biliary dilatations were fusiform in 21 cases and cystic in 19 cases. Mean age (range) at surgery was 4.8 (0.3–14.1) years, and mean weight at surgery was 17.0 (5.5–47.0) kg. Five patients had intrahepatic bile duct dilatation. There were no intraoperative complications. Estimated mean blood loss was minimal at 15 mL. Hepaticojejunostomy diameters were 6–9 mm in 13/21 fusiform cases and 12/19 cystic cases, more than 10 mm in 8/21 fusiform cases and 7/19 cystic cases.

Intraoperative endoscopy of both the common channel and intrahepatic bile ducts was performed in 25 cases (21 fusiform; 4 cystic); the remaining 15 had intraoperative endoscopy of intrahepatic ducts alone because the ureteroscope could not be inserted into the intrapancreatic choledochus and common channel. Protein plugs were present in the common channel in all 21 fusiform cases (massive in 6, moderate in 12, minimal in 3), successfully cleared by irrigation with normal saline from the side channel of the ureteroscope. Debris were present in all 15 cases who had intraoperative endoscopy of the intrahepatic bile ducts alone (moderate in 6, minimal in 9). There were no debris in the intrahepatic bile ducts of the 25 who had intraoperative endoscopy of both the common channel and intrahepatic bile ducts.

Although all patients are well after a mean follow-up of 4.5 years (range: 6 months to 8.5 years) with cosmetically esthetic wounds, there were three cases of postoperative complications. The first was pancreatitis that developed 8 months postoperatively in a case with massive protein plugs on intraoperative endoscopy, even though all plugs were cleared thoroughly by irrigation. The pancreatitis was treated by conservative medical management, and there have been no further episodes. The cause was attributed to new 3 × 3 mm debris. The second was duodenal obstruction in a cystic case. At exploratory laparoscopy, the third part of the duodenum was found to be compressed by the Roux-en-Y limb that had been inadequately fixed to the colonic mesentery. Once the sutures between the Roux-en-Y limb and colonic mesentery were released laparoscopically, the postoperative recovery was uneventful. The third was anastomotic leak treated by minilaparotomy.

24.4 Conclusion

The authors were the first to report the value of customizing the length of the short Roux loop, performing the hepaticojejunostomy very close to the closed end of the Roux loop blind end, and creating the enterotomy on the anterior side of the Roux loop as part of their routine laparoscopic bile duct excision procedure. Despite extra trocars and longer operative time, postoperative pain is minimized, allowing patients to be discharged earlier. Intraoperative endoscopy is invaluable for reducing mid- to long-term postoperative complications.

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