Biliary Atresia

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Biliary atresia remains one of the most intractable gastrointestinal surgical diseases in infancy. The introduction of liver transplantation (LTx) has revolutionized the protocols for the treatment of this condition, hepatic portoenterostomy (the Kasai procedure) is still the first-line surgical treatment.

The main clinical manifestations of biliary atresia are persistent jaundice, clay-colored feces, and hepatomegaly. Although symptoms do not vary greatly, anatomic findings of the biliary tract vary from case to case. Only about 10–15% of patients with biliary atresia have an extrahepatic bile duct large enough to perform a mucosa-to-mucosa anastomosis to the intestine. These correctable cases can undergo hepaticoenterostomy. The remaining 85–90% of patients do not have a viable bile duct amenable to a conventional anastomosis. For these noncorrectable-type cases, hepatic portoenterostomy should be performed.

41.1 Surgical Treatment: Hepatic Portoenterostomy

Hepatic portoenterostomy was first devised in 1957 as corrective surgery for patients with the noncorrectable type of biliary atresia. The basis of this procedure is that intrahepatic bile ducts are patent in early infancy, and minute intrahepatic bile ducts are present in the cone-shaped fibrous tissue, replacing extrahepatic biliary radicles. In hepatic portojejunostomy, extrahepatic bile ducts, including fibrous remnants at the porta hepatis, are completely removed, and bile drainage is established by the anastomosis of an intestinal conduit to the transected surface at the porta hepatis. Microscopic biliary structures at the liver hilum drain bile into the intestinal conduit, and, in time, an autoapproximation occurs between the intestinal and ductal epithelial elements.

Successful hepatic portoenterostomy depends on early diagnosis and surgery (preferably in the first 30 days after birth), adequate operative technique, prevention of postoperative cholangitis, and precise postoperative management.

41.1.1 Preoperative Care

In addition to routine preoperative care for neonates and young infants undergoing surgery, vitamin K (1–2 mg/kg per day) is usually given for several days before surgery. Packed red blood cells are cross-matched, and preoperative broad-spectrum antibiotics are administered.

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41.1.2 Operative Technique

Surgery is performed under general anesthesia with tracheal intubation. The patient is placed in the supine position. A pillow is placed under the back of the patient for better exposure. Laparotomy is performed through a right subcostal or upper abdominal transverse incision (Fig. 41.1). Some pediatric surgeons recommend mobilizing and completely exposing the liver to enable adequate visualization of the porta hepatis. The authors do not use this technique because of its potential invasiveness; we believe that enough exposure is obtained for Kasai portoenterostomy without this technique.

Liver biopsy is obtained. Then, a catheter is inserted into the small gallbladder to perform cholangiography. After atresia of the extrahepatic bile duct is confirmed by cholangiography, the gallbladder is freed from the liver (Fig. 41.2), and dissection is advanced along with the cystic duct toward the common hepatic duct. Dissection is performed with the help of surgical loupes at 2.5× magnification.

The superficial peritoneum on the hepatoduodenal ligament is opened, and the anatomy of the involved bile duct and hepatic arteries is assessed. Dilated lymphatic channels around the hepatoduodenal ligament should be carefully ligated and divided to avoid excessive lymphatic fluid loss, which occasionally causes intractable postoperative ascites. The common bile duct remnant is carefully dissected, because it is often adherent to the surrounding tissues.

The common bile duct is clamped and divided adjacent to the duodenum. After the common bile duct is severed, it is raised, and the hepatic duct remnant is freed from underlying hepatic arteries and the portal vein (Fig. 41.3). The hepatic duct usually transforms into a cone-shaped fibrous tissue, which is situated cranial to the bifurcation of the portal vein. It continues adjacent to the portal tracts in the liver. Even if a cyst-like structure is present, it should be removed and should not be used for the anastomosis to the intestine.

The separation of fibrous remnants from right and left portal veins is carefully advanced posteriorly. The bifurcation of right and left portal veins must be retracted to obtain proper exposure of the porta hepatis. Several small branches bridging from the portal vein to fibrous remnants are identified and divided between ligatures, facilitating downward displacement of the portal vein (Fig. 41.4). The posterior aspect of fibrous remnants is exposed deep and wide enough behind bilateral main branches of the portal vein. The caudate lobe also should be well visualized behind the bifurcation of the portal vein and caudal to fibrous remnants.

Dissection between the anterior aspect of fibrous remnants and the quadrate lobe of the liver is also sufficiently advanced. The fibrous remnant at the porta hepatis is vertically divided in the middle (Fig. 41.5), taking care not to injure the liver capsule between the quadrate and caudate lobes.

The divided remnants on both sides are independently and completely removed in order without injuring the liver capsule (Fig. 41.6). The transection of fibrous remnants is carefully performed using small, round scissors or a sharp scalpel at the level of the posterior surface of the portal vein. The transected surfaces of the fibrous remnants are located close to the bilateral main branches of the portal vein, and the surrounding liver capsule should be at the same level.

Although some surgeons confirm the presence of microscopically patent ducts at the level of the anastomosis using frozen sections perioperatively, we do not use frozen-section guidance because we always transect the portal bile duct remnants at the same level. Hemorrhage from the superficial incision of the porta hepatis is occasionally considerable. Irrigation with warm saline stops the bleeding, usually within 10 min. Oozing at the porta hepatis is usually controlled by the anastomosis of the posterior row of the portoenterostomy. Ligation or cautery should not be applied because of the possibility of accidental obliteration of small bile ducts that may be opening on the transected surface.

The next step is the construction of a Roux-en-Y loop of the jejunum (Fig. 41.7). A Roux-en-Y anastomosis with the ascending limb is constructed, with an approximate length of 50 cm (or a weight of 10 cm/kg). The end of the gastric limb of the intestine is anastomosed not to the antimesenteric surface but to the lateral surface of the hepatic limb of the intestine. Currently, most surgeons avoid using a cutaneous stoma because of the frequent bleeding that occurs when the patient develops portal hypertension. Also, technical difficulties will arise if subsequent LTx becomes necessary, and no reduction has been observed in the incidence of cholangitis.

A spur valve is created, which may prevent cholangitis. Two centimeters of half of the seromuscular layer facing the gastric limb are removed from the biliary limb proximal to the anastomosis (Fig. 41.8). The gastric and biliary limbs are then coapted over the denuded mucosa with sutures along the edges of the removed seromuscular layer (Fig. 41.9).

After the completion of valve creation, the hepatic limb is brought up retrocolically. The end of the intestine is anastomosed around the transected end of the fibrous remnants at the porta hepatis with full-thickness interrupted sutures using 5/0 monofilament absorbable sutures (Fig. 41.10). Sutures must not be placed on the transected surface of fibrous remnants, in which minute bile ducts are present. After all inter-

rupted sutures of the posterior row are placed in position, they are tied.

The anterior row is sutured in a manner similar to the posterior row of the anastomosis (Figs. 41.11). Again, it should be emphasized that sutures must not be placed on the transected surface of fibrous remnants but rather on the liver tissue around the transected area.

Several interrupted seromuscular sutures are added using 5/0 braided absorbable sutures between the jejunum and the quadrate lobe of the liver and/or the hepatoduodenal ligament, if reinforcement of the anastomosis is believed to be required (Fig. 41.12). Irrigation of the abdominal cavity with saline should be performed sufficiently. The intestine, particularly the jejunum including the biliary limb, is carefully placed in order within the abdominal cavity to prevent ileus. A Penrose drain is placed in the foramen of Winslow. After the viscera of the upper abdomen, including the small intestine and the liver, is covered by Seprafilm®, the abdominal cavity is closed in layers. A central venous catheter is placed for postoperative management, and surgery is concluded.

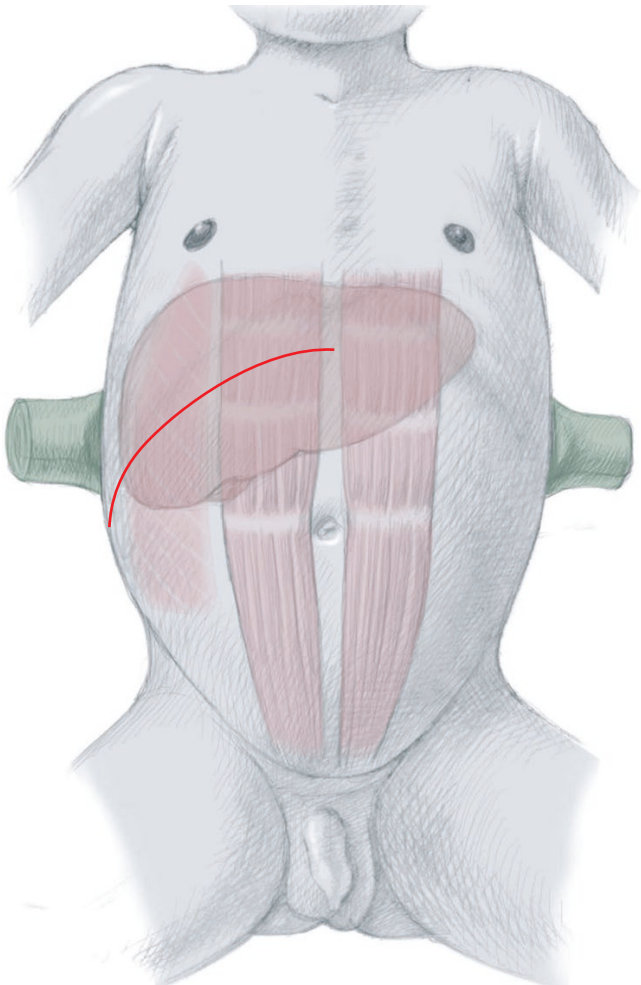

Fig. 41.1 Incision for Kasai portoenterostomy

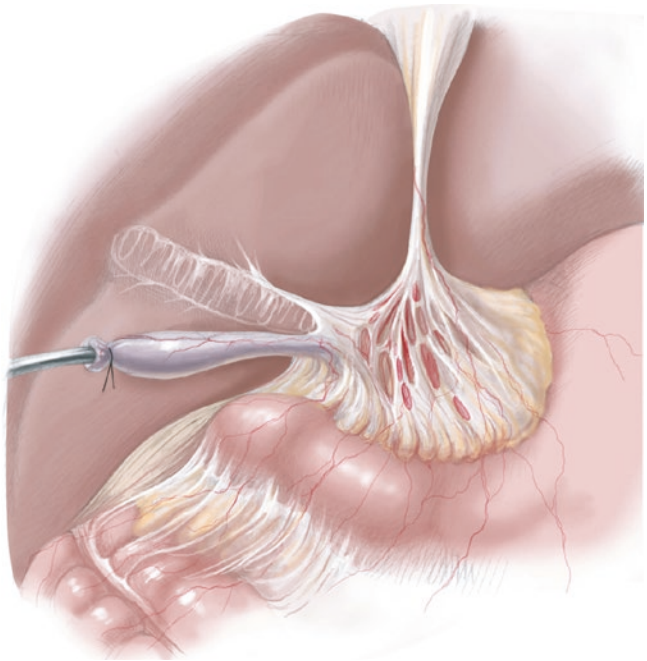

Fig. 41.2 Freeing the gallbladder from the liver

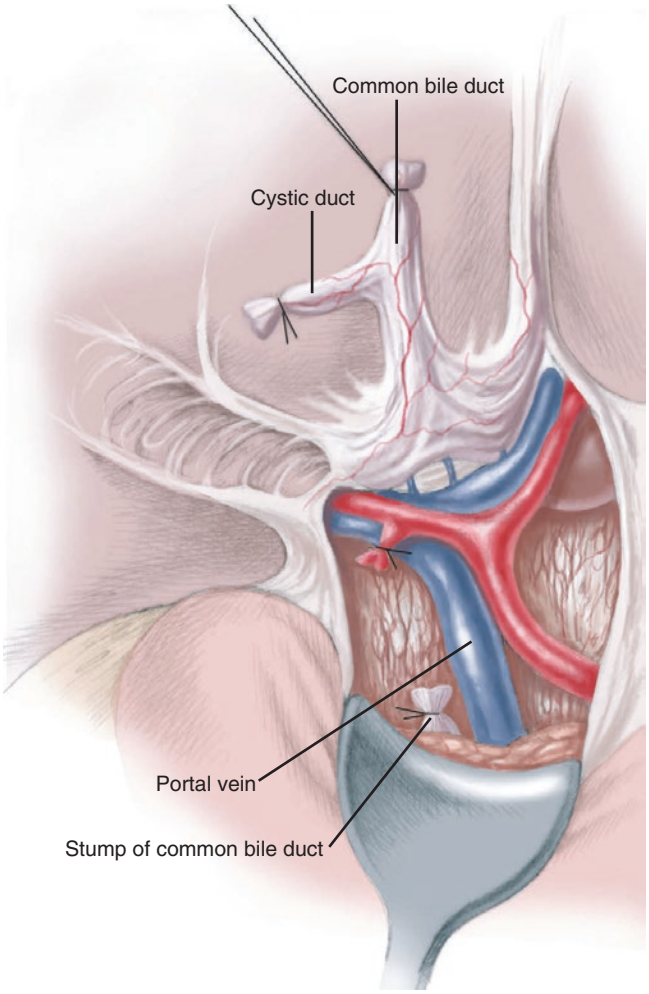

Fig. 41.3 Severed common bile duct

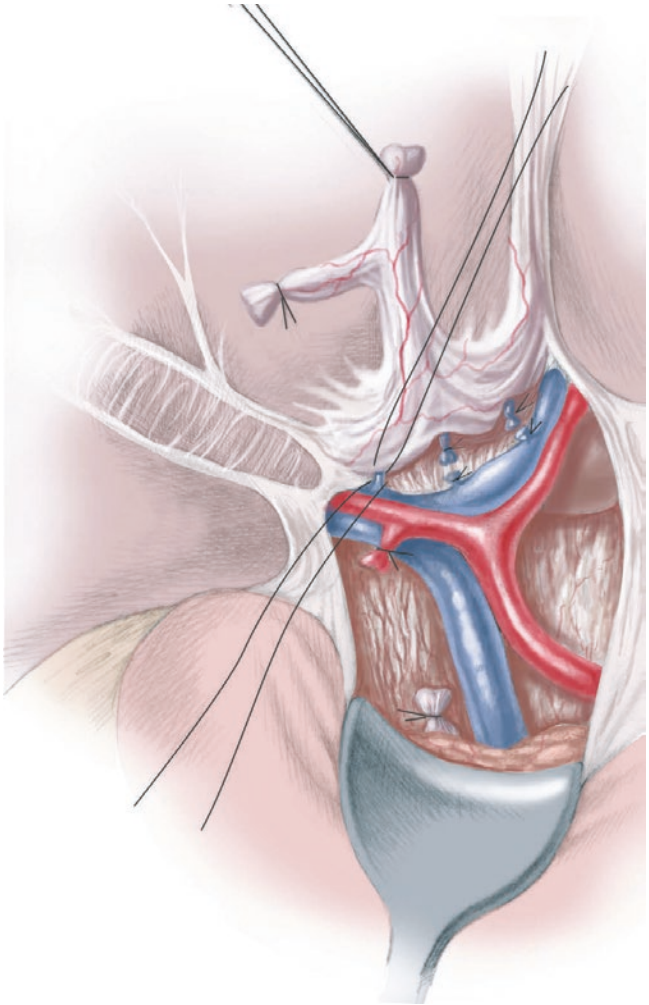

Fig. 41.4 Downward displacement of the portal vein to expose the porta hepatis

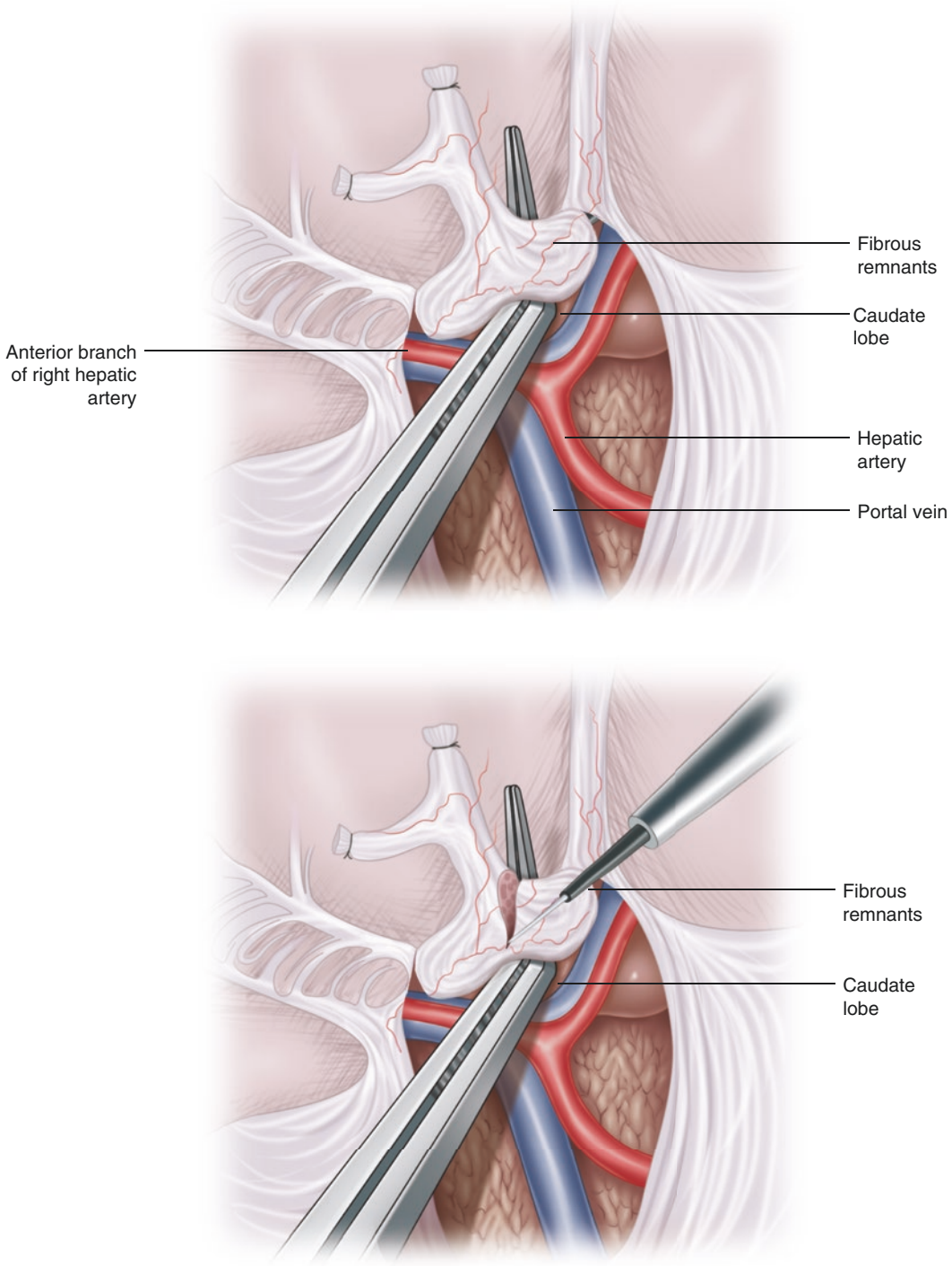

Fig. 41.5 Dissection of the fibrous remnant at the porta hepatis

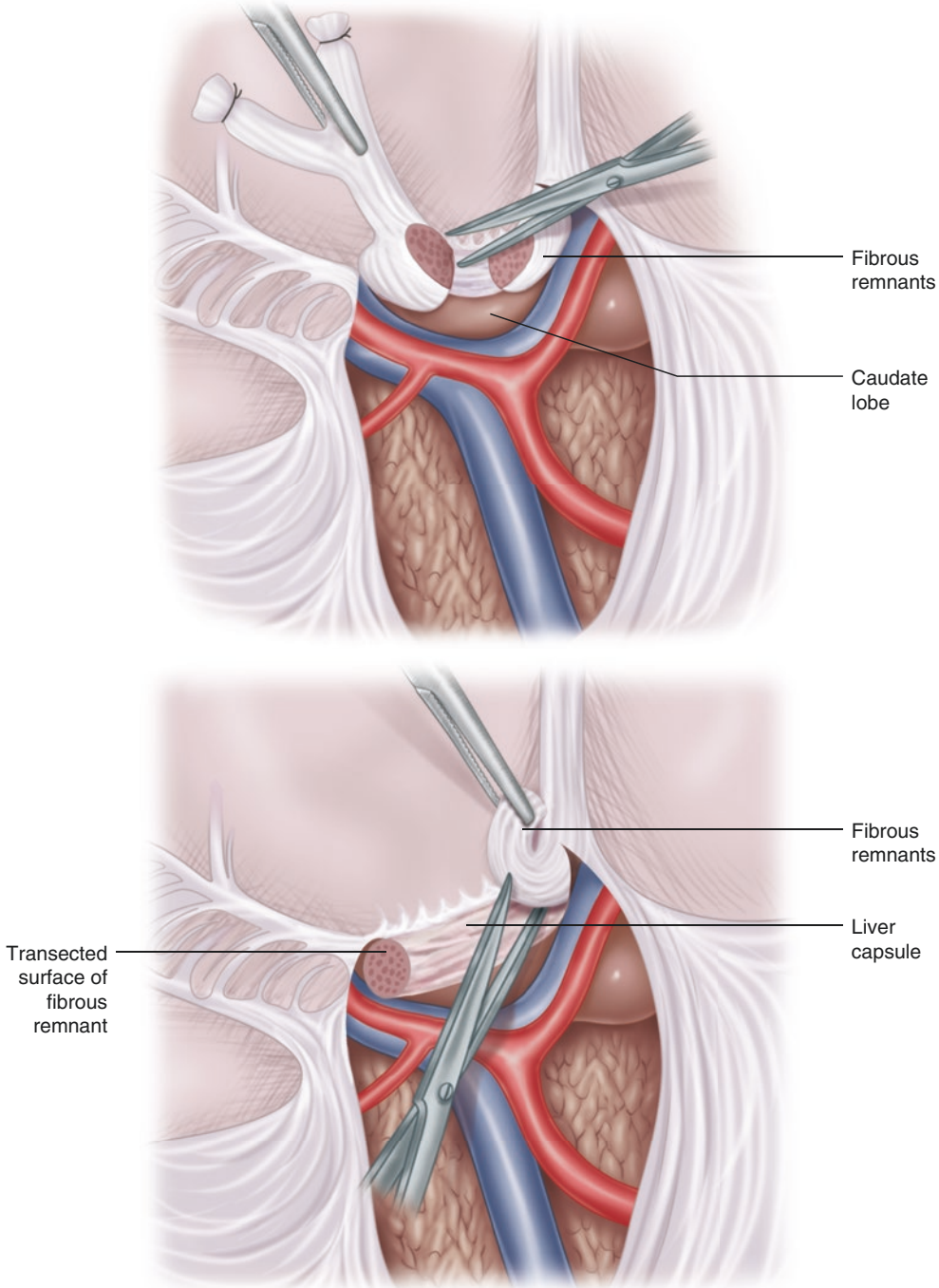

Fig. 41.6 Removal of the fibrous remnants

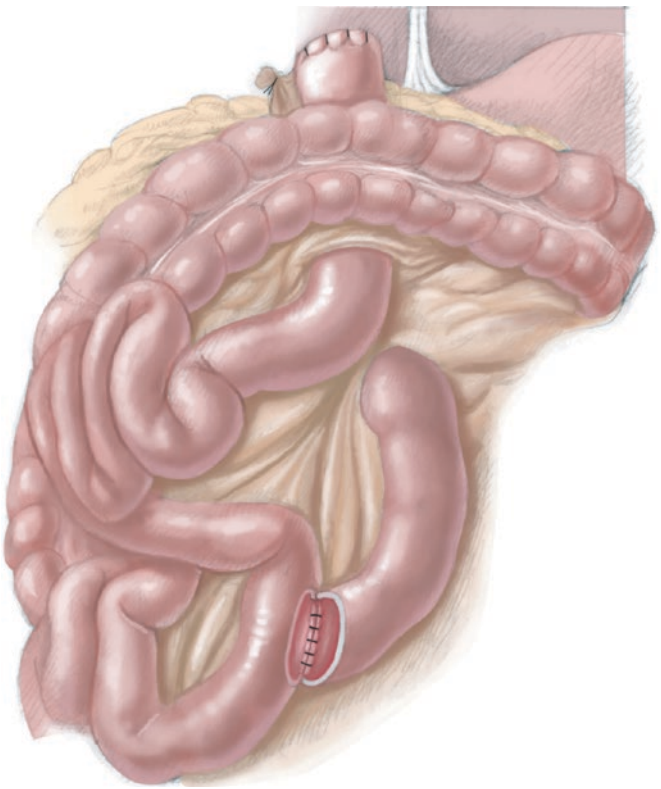

Fig. 41.7 Construction of a Roux-en-Y loop of the jejunum

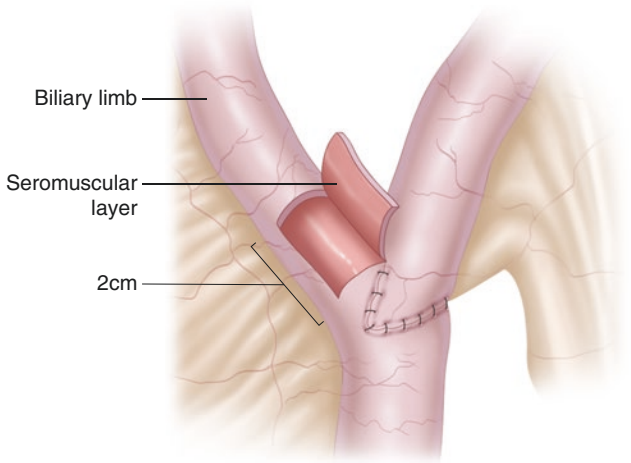

Fig. 41.8 Removal of a portion of seromuscular layer to create a spur valve

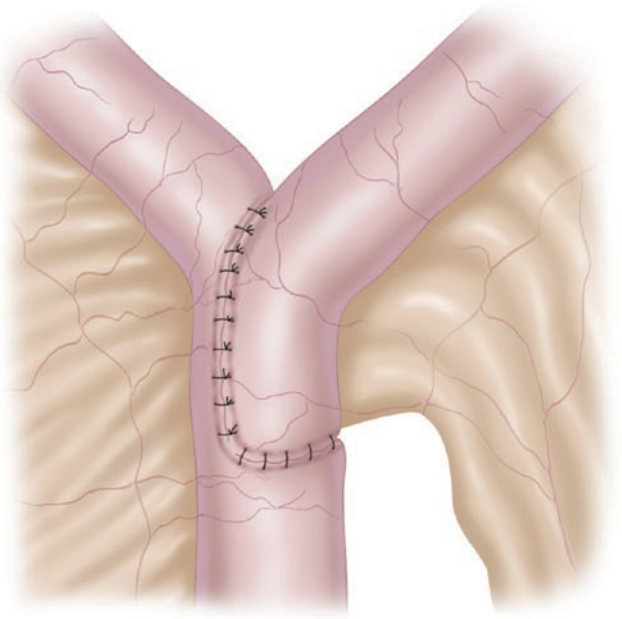

Fig. 41.9 Coaptation of the gastric and biliary limbs

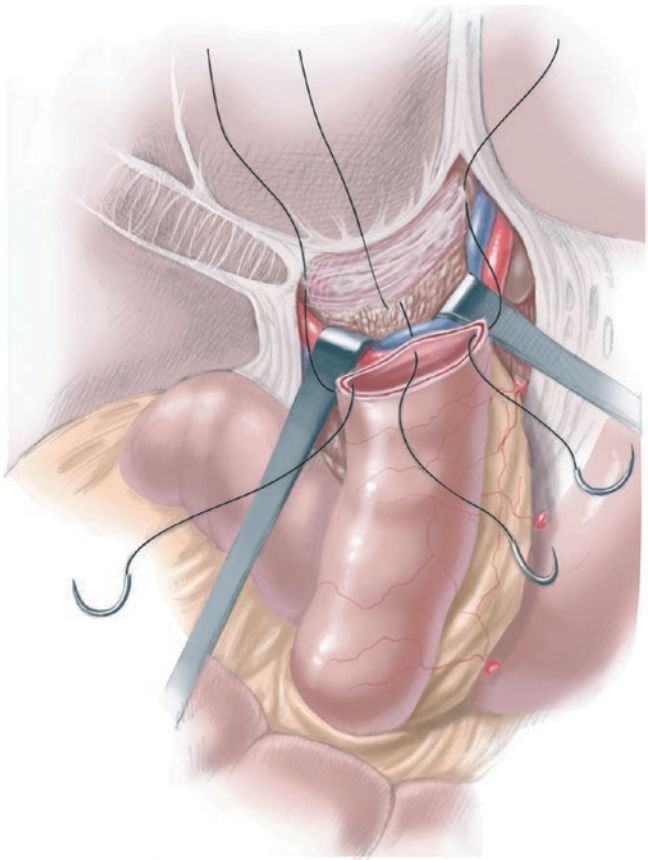

Fig. 41.10 Creation of the anastomosis: suturing of the posterior row

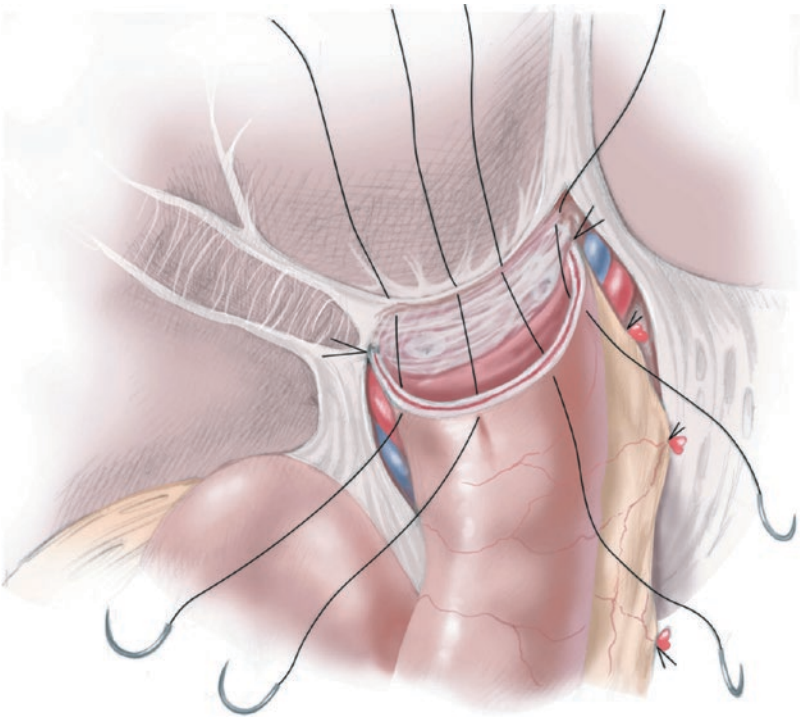

Fig. 41.11 Suturing of the anterior row

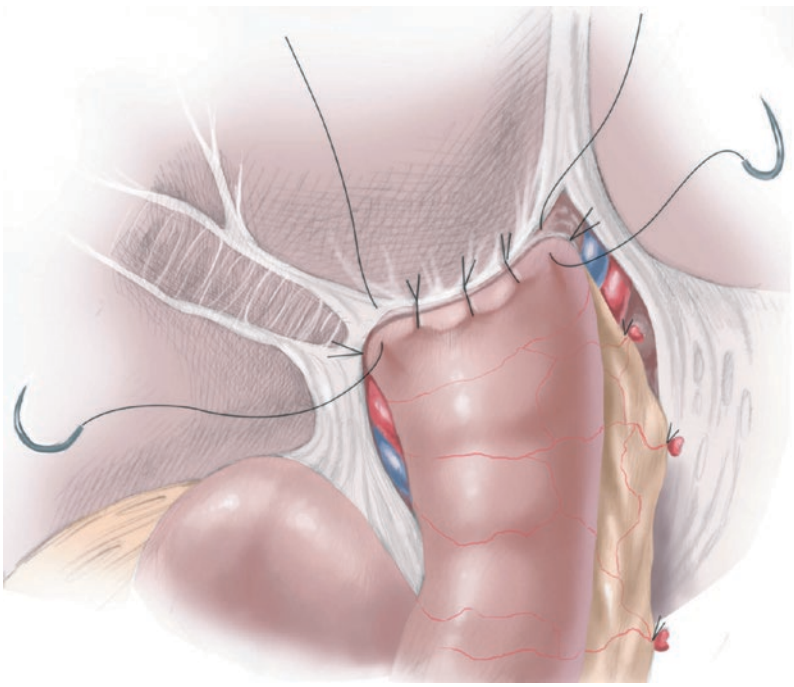

Fig. 41.12 Placement of sutures between the jejunum and the quadrate lobe of the liver

41.1.3 Postoperative Care

Patients are placed on oxygen via nasal cannula, and intravenous fluid is given. Decompression of the gastrointestinal tract should be performed by nasogastric aspiration and enema. Oral feeding is usually initiated on the fifth or sixth postoperative day, when bowel activity resumes. Prevention of postoperative cholangitis is the main goal of postoperative management. For this purpose, we routinely use antibiotics, choleretics, and steroids as postoperative medication. For antibiotics, amikacin sulfate at a dose of 8 mg/kg per day, q8h, is administered for 7 days postoperatively. Then cephalosporin at a dose of 50–80 mg/kg per day is given intravenously for a few months, until the serum bilirubin level is below 2 mg/dL. For choleretics, ursodeoxycholic acid at a dose of 20 mg/kg per day is given orally as soon as oral feeding is initiated. Prednisolone, 4 mg/kg per day, is initiated after the seventh postoperative day. Steroid therapy is continued intravenously for 4 days and is then switched to oral administration of prednisolone at 4 mg/kg bid every other day until the serum bilirubin level is below 2 mg/dL, or for a maximum of 3 months following hepatic portoenterostomy.

41.2 Complications

The most frequent and serious postoperative complication is cholangitis, which can result in fatal septicemia or reobliteration of the hepatic portoenterostomy. Although there are various modified reconstruction procedures to prevent cholangitis, the authors now perform hepatic portojejunostomy using a long Roux-en-Y limb with a length of 50 cm or a weight of 10 cm/kg equipped with a spur valve, as shown in this chapter. The incidence of cholangitis during the first year following Kasai portoenterostomy, however, remains at approximately 30%. Cirrhosis and portal hypertension and/or hypersplenism also have been documented as late complications. It has been shown that both late surgery and the complication of cholangitis aggravate hepatic fibrosis and induce portal hypertension.

41.3 Outcomes

Between 1953 and 2013, a total of 350 patients with biliary atresia underwent surgery in our hospital. Of these, 35 patients underwent hepatic portoenterostomy with a spur valve during the most recent 13 years. Bile drainage postoperatively was achieved in 100% of cases and 85.7% (30 patients) became jaundice-free. The incidence of postoperative cholangitis in the first year following hepatic portoenterostomy was 25.7%. Of the patients treated using this procedure, 26 (74.3%) are still alive with their native liver and without jaundice, eight underwent subsequent LTx, and one is waiting for LTx due to recurrent jaundice. No patient died.

Conclusions

The combination of hepatic portoenterostomy with subsequent LTx is the treatment of choice for patients with biliary atresia. It is important, however, to attempt preservation of the patient's native organ by continuing efforts to achieve the best possible results with hepatic portoenterostomy.

Our current strategy for surgical treatment of patients with this disease includes (1) early diagnosis, including prenatal diagnosis; (2) hepatic portoenterostomy without stoma formation; (3) careful postoperative care, particularly for the prevention of postoperative cholangitis; (4) revision of hepatic portoenterostomy only in selected patients who showed good bile drainage after the first procedure; (5) early LTx in patients with absolutely failed hepatic portoenterostomy; (6) avoidance of laparotomy for the treatment of esophageal varices and hypersplenism (endoscopic injection sclerotherapy and partial splenic artery embolization); and (7) consideration of primary LTx for patients with advanced liver damage at the time of referral.

Suggested Reading

Nio M, Ohi R. Biliary atresia. Semin Pediatr Surg. 2000;9(4):177–86.

Nio M, Sano N, Ishii T, Sasaki H, Hayashi Y, Ohi R. Cholangitis as a late complication in long-term survivors after surgery for biliary atresia. J Pediatr Surg. 2004;39(12):1797–9.

Nio M, Sasaki H, Tanaka H, Okamura A. Redo surgery for biliary atresia. Pediatr Surg Int. 2013;29(10):989–93.

Nio M, Wada M, Sasaki H, Kazama T, Tanaka H, Kudo H, et al. Technical standardization of Kasai portoenterostomy for biliary atresia. J Pediatr Surg. 2016;51:2105–8.

Nio M, Wada M, Sasaki H, Tanaka H. Effects of age at Kasai portoenterostomy on the surgical outcome: a review of the literature. Surg Today. 2015;45(7):813–8.