



Kasai Portoenterostomy (Open and MIS Approaches)

62

Stefan Scholz and Lea Wehrli

Indications and Benefits

- Persistent direct hyperbilirubinemia.
- Persistent infantile jaundice with negative medical work-up for alternative.
- Diagnose and treat biliary atresia.
- Benefits: Establish hepatic biliary drainage, improve gastrointestinal absorption for fat-soluble vitamins, and prevent progression of biliary liver cirrhosis.

Risks and Alternatives

- Standard risks (bleeding, infection, need for additional procedures, risks of anesthesia).
- Injury to hepatic arteries or portal vein, small or large bowel.
- Injury to liver at porta hepatis causing bleeding and granulation tissue obstructing bile drainage.

- Anastomotic leak of portoenterostomy (low flow, rare) or jejunio-jejunostomy.
- Postoperative obstruction of Roux limb through adhesions or retrocolonic window.
- Postoperative bowel adhesive obstruction.
- Ascites.
- Ascending cholangitis.
- Deficiency of fat-soluble vitamins.
- Progression of biliary cirrhosis.
- Alternatives: Direct liver transplant after definite diagnosis of biliary atresia by cholangiogram (or ERCP) in cases of advanced liver cirrhosis.

Essential Steps

Cholangiogram

1. Small right subcostal incision to enter the abdomen.
2. Evacuate possible ascites.
3. Retract the liver cranially with malleable retractor and identify gallbladder. Note the size and color of the gallbladder.
4. Place a 5-0 Prolene purse-string suture around the dome of the gallbladder (gallbladder dome may have to be mobilized).
5. Enter the gallbladder with a sharp knife in the center of the purse-string suture, and place a flushed Taut cholangiogram catheter into the

S. Scholz (✉)
University of Pittsburgh School of Medicine,
Children's Hospital of Pittsburgh, Department of
Pediatric Surgery, Pittsburgh, PA, USA
e-mail: stefan.scholz@chp.edu

L. Wehrli
Children's Hospital of Pittsburgh of UPMC,
Department of Pediatric Surgery,
Pittsburgh, PA, USA

- lumen. Note the amount and the color of bile (“white bile?”).
6. Tie the purse-string suture around the catheter to obtain a good seal (may add silk tie), and minimize leakage of contrast agent.
 7. Place the patient in a good Trendelenburg position.
 8. Inject contrast agent with a small syringe into catheter under fluoroscopy guidance (may use flat plate X-ray if quality of fluoroscopy is low).
 9. Contrast should flow with minimal resistance into bile system.
 10. If distal bile ducts and duodenal outflow is identified but no intrahepatic bile system, compress distal bile duct at the border of the duodenum with a retractor toward the spine to increase outflow pressure.
 11. Cholangiogram is complete and normal if contrast reaches the duodenum and the proximal biliary tree within the liver.
 12. In cases of biliary atresia, the gallbladder may be minuscule, and there may be only a small amount of “white bile” (mucous). In most cases of biliary atresia, the extrahepatic bile duct has no lumen at all. In about 20% of cases, there is a patent bile duct into the duodenum, but no proximal or intrahepatic bile system can be imaged.
 13. If there is a patent bile system, the injection pressure of the contrast, as felt injecting with a small syringe, is minimal. In a patient with Alagille syndrome, the bile system is patent but small in caliber and the pressure may be higher. In case of atretic bile ducts, the injection pressure becomes very high and the contrast may start leaking. High-pressure injections may penetrate into the lymphatic system of the porta hepatis or into the subperitoneal plane, which may be confusing on fluoroscopy.
2. If there is a good-sized gallbladder, the cholangiogram can be performed laparoscopically.
 3. Place an additional 3 mm instrument into the right abdomen for liver retraction.
 4. Under direct vision, place a Tuohy needle through the abdominal wall, through the liver into the gallbladder to minimize leakage of contrast. Alternatively, the needle (or other catheter) can be placed into the gallbladder dome using a laparoscopic clip.
 5. Follow all steps from 7 on for open cholangiogram above.

Kasai Portoenterostomy

1. After definite diagnosis of biliary atresia by means of cholangiogram, extend the right subcostal incision to both sides.
2. Divide the left and right triangular ligaments as well as the falciform ligament to mobilize the liver.
3. With care, rotate the liver anteriorly out of the abdomen using the corners of the incision as retractors behind the right and left hepatic lobes.
4. Mobilize the gallbladder completely, divide the cystic artery, and follow the cystic duct toward the portal vessels.
5. Dissect the peritoneum over the porta hepatis and identify the hepatic artery and its branches.
6. Identify the bile duct remnant and follow this toward the edge of the duodenum. Ligate the bile duct remnant at the border of the duodenum and divide.
7. Follow the bile duct remnant proximally toward the surface of the liver to reach its proximal fibrous cone (if present).
8. Dissect the peritoneum of the fibrous cone anteriorly and laterally to the branches of the hepatic artery. Typically, there are small branches of the hepatic artery entering into the fibrous cone from lateral at its anterolateral border, which can be divided using monopoly or bipolar cautery. Carry this dissection as far lateral as possible to maximize the liver surface covered by the portoenterostomy.

Laparoscopic Cholangiogram

1. Place a small trocar through the umbilicus, establish pneumoperitoneum and forward the laparoscope.

9. Dissect the peritoneum posteriorly behind the bifurcation of the portal vein. Take great care to identify two typical small portal vein branches (one from left and one from right portal vein) entering the posterior aspect of the fibrous cone. Ligate both branches with 6-0 Prolene vs. cautery.
10. Once the complete fibrous cone is dissected on its four borders, laterally to the hepatic arteries and posteriorly behind the portal vein, the biliary remnant is removed from the liver surface. Use small and very sharp scissors for meticulous removal of the scar tissue avoiding any injury to the surface of the liver.
11. Send the biliary remnant to pathology together with a fresh liver wedge biopsy.
12. Place Gelfoam sponge onto the bare hepatic plate and return the liver back into the abdomen.
13. Identify a loop of jejunum about 10 cm distal to the ligament of Treitz, make a window in the mesentery and divide the jejunum with an Endo GIA stapler.
14. Divide the mesentery further to its root and measure a 30–40 cm long Roux loop.
15. Perform a side-to-side jejuno-jejunostomy with a 4-0 PDS running suture directing the stream of enteric contents toward distal.
16. Close the mesenteric gap.
17. Create a window through the retrocolonic mesentery behind the right transverse colon and forward the Roux limb through the window toward the liver.
18. Extrovert the liver from the abdomen again and remove the Gelfoam sponge. Inspect the portal plate for bleeding.
19. Open an antimesenteric enterotomy just proximal to the end of the Roux loop in the appropriate length.
20. Perform an end-to-side portoenterostomy using interrupted 6-0 Maxon sutures maximizing the covered area on the liver surface. The sutures should be placed through the liver well behind the portal bifurcation and just next to the hepatic artery branches on both sides. First, place the back-row sutures. Typically, 8–10 sutures are necessary.
21. Place the liver back into its original position in the abdomen and position the Roux limb into the abdomen and through the transverse colonic mesentery to minimize the chance of kinking and obstruction.
22. Fixate the Roux limb with three sutures into the retrocolonic mesenteric window.
23. Check the orientation of the Roux limb and the jejuno-jejunostomy one last time.
24. Irrigate the abdomen with warm saline.
25. Close the abdominal incision and the skin in multiple layers.

Laparoscopic Kasai Portoenterostomy

1. After definite diagnosis of biliary atresia by means of laparoscopic cholangiogram, place additional three trocars into the right and left abdomen.
2. Retract the liver upwards using either a liver retractor and/or placing a transabdominal suture through the gallbladder remnant.
3. Leaving the gallbladder in place for retraction, divide the cystic artery with hook cautery, and follow the cystic duct toward the portal vessels.
4. Most tissue dissection is performed with hook cautery or careful blunt dissection.
5. Dissect the peritoneum over the porta hepatis and identify the hepatic artery and its branches.
6. Identify the bile duct remnant and follow this toward the edge of the duodenum. Ligate the bile duct remnant at the border of the duodenum and divide.
7. Follow the bile duct remnant proximally toward the surface of the liver to reach its proximal fibrous cone (if present).
8. Dissect the peritoneum of the fibrous cone anteriorly and laterally to the branches of the hepatic artery. Typically, there are small branches of the hepatic artery entering into the fibrous cone from lateral at its anterolateral border, which can be divided using monopoly or bipolar cautery. Carry this dissection as far lateral as possible to

- maximize the liver surface covered by the portoenterostomy.
9. Dissect the peritoneum posteriorly behind the bifurcation of the portal vein. Take great care to identify two typical small portal vein branches (one from left and one from right portal vein) entering the posterior aspect of the fibrous cone. Ligate both branches with cautery.
 10. Once the complete fibrous cone is dissected on its four borders, laterally to the hepatic arteries and posteriorly behind the portal vein, the biliary remnant is removed from the liver surface. Use very sharp scissors meticulous removal of the scar tissue avoiding any injury to the surface of the liver.
 11. Send the biliary remnant to pathology together with a fresh liver wedge biopsy.
 12. Place Gelfoam sponge onto the bare hepatic plate.
 13. Extend the umbilical incision up- and downwards and exteriorize the small bowel.
 14. Identify a loop of jejunum about 10 cm distal to the ligament of Treitz, make a window in the mesentery, and divide the jejunum with an Endo GIA stapler.
 15. Divide the mesentery further to its root and measure a 30–40 cm long Roux loop.
 16. Perform a side-to-side jejunum-jejunostomy with a 4-0 PDS running suture directing the stream of enteric contents toward distal.
 17. Close the mesenteric gap.
 18. Place all the bowel back into the abdomen, approximate the umbilical incision and replace the umbilical trocar.
 19. Laparoscopically, create a window through the retrocolonic mesentery behind the right transverse colon and forward the Roux limb through the window toward the liver.
 20. Remove the Gelfoam sponge. Inspect the portal plate for bleeding.
 21. Open an antimesenteric enterotomy just proximal to the end of the Roux loop in the appropriate length.
 22. Perform an end-to-side portoenterostomy using interrupted or running 6-0 Maxon sutures maximizing the covered area on the liver surface. The sutures should be placed through the liver well behind the portal bifurcation and just next to the hepatic artery branches on both sides. First, place the back-wall suture.
 23. Position the Roux limb into the abdomen and through the transverse colonic mesentery to minimize the chance of kinking and obstruction.
 24. Fixate the Roux limb with three sutures into the retrocolonic mesenteric window.
 25. Check the orientation of the Roux limb and the jejunum-jejunostomy one last time.
 26. Remove the gallbladder and send it to pathology.
 27. Irrigate the abdomen with warm saline.
 28. Remove all instruments and trocars from the abdomen, close the abdominal incision and the skin in multiple layers.

Note These Variations

- A 5-mm stapler may be utilized to divide the bowel and create the jejunum-jejunostomy and the Roux limb.
- An intraabdominal laparoscopic stapled jejunum-jejunostomy may be performed to create the Roux limb without exteriorization of the intestine.
- The 3 mm laparoscopic bipolar sealer or hook cautery can be used for laparoscopic dissection.

Template Operative Dictation

Preoperative Diagnosis Persistent direct hyperbilirubinemia, possible biliary atresia

Postoperative Diagnosis Biliary atresia

Findings Small gallbladder remnant containing “white bile,” atretic biliary system with proximal fibrous cone

Procedures Performed *Laparoscopically assisted/open* cholangiogram, liver wedge biopsy, *laparoscopic/open* cholecystectomy,

exploration of extra hepatic bile ducts to rule out biliary atresia, excision of atretic extrahepatic biliary remnant with proximal fibrous cone, jejunostomy with creation of 30 cm Roux-en-Y limb, and *laparoscopic/open* Kasai portoenterostomy

Anesthesia General anesthesia

Specimen Gallbladder, liver wedge biopsy, extrahepatic biliary remnant, and fibrous cone

Drains None

Implants None

Estimated Blood Loss Minimal

Indications This ___ -day/-week/-month-old boy/girl with persistent direct hyperbilirubinemia and jaundice was taken to the operating room for intraoperative cholangiogram to diagnose or rule out biliary atresia.

Procedure in Detail Following satisfactory induction of anesthesia, the patient was placed in supine position and appropriately padded. Timeouts were performed using both pre-induction and pre-incision safety checklists with participation of all present in the operative suite. These confirmed the correct patient, procedure, operative site, instrumentation, and additional critical information prior to the start of the procedure. The abdomen was then prepped and draped in the usual sterile fashion and a broad-spectrum antibiotic was given intravenously.

Cholangiogram

A small right subcostal incision was made to enter the abdomen. This was then carried down to the fascia using electrocautery. The peritoneum was identified and incised to enter the abdominal cavity. *There was a small amount of ascites, which was evacuated. The liver appeared soft and brown with a sharp edge/The liver appeared dark and indurated with a round edge (and a*

greenish color to it). The liver was retracted cranially and the gallbladder identified. The gallbladder was very small. A 5-0 Prolene purse-string suture was placed around the dome of the gallbladder after the gallbladder dome was mobilized. The gallbladder was opened with a knife in the center of the purse-string suture and a small amount of white mucous drained. A flushed *Taut* cholangiogram catheter was forwarded into the lumen of the gallbladder and the purse-string suture tied. The patient was placed in a Trendelenburg position.

If laparoscopic cholangiogram: An umbilical trocar was placed and pneumoperitoneum established. The peritoneal cavity was inspected for any injuries from abdominal access. The liver appeared soft and brown with a sharp edge/The liver appeared dark and indurated with a round edge (and a greenish color to it). The gallbladder could not be immediately visualized. We then placed a second laparoscopic instrument into the abdomen through a stab incision in the right abdomen. The right lobe of the liver was retracted and the gallbladder identified. (The gallbladder appeared very contracted, and the decision was made to convert to an open procedure.) Under direct vision, a Tuohy needle is introduced through the abdominal wall and directed (transhepatically) into the lumen of the gallbladder.

Under fluoroscopy guidance, a cholangiogram was performed.

The contrast only filled the small gallbladder lumen without progression into the cystic duct and the common bile duct/.The cholangiogram showed a small gallbladder with a patent distal bile duct and outflow into the duodenum. The distal bile duct was compressed at the border of the duodenum onto the spine but a patent proximal extrahepatic and intrahepatic bile system could not be demonstrated.

The cholangiogram was then completed and the parents notified that biliary atresia was confirmed. We then continued with the Kasai procedure.

If cholangiogram negative: The cholangiogram showed a patent extra- and intrahepatic bile system with evidence of a normal intrahepatic bile tree and outflow of contrast into the

duodenum. *The bile ducts appeared very small and Alagille syndrome should be excluded.* The subcostal incision was closed in a running manner in two layers. *All trocars were removed and the port sites closed.* Scarpa's fascia was approximated and the skin was closed with a subcuticular running stitch. Upon completion of the procedure, a debriefing checklist was completed to share information critical to the postoperative care of the patient. The patient tolerated the procedure well, was extubated in the operating room, and was transported to the postanesthesia care unit in stable condition.

Kasai Portoenterostomy

After confirmation of biliary atresia with the cholangiogram, the right subcostal incision was extended toward lateral and medial. The left and right triangular ligaments were divided to mobilize the liver as well as the falciform ligament. The liver was then rotated out of the abdomen using both corners of the incision as retractors behind the right and left hepatic lobes. The gallbladder was completely mobilized and the cystic artery divided and follow. We then explored the extra hepatic bile system to further confirm the diagnosis of biliary atresia. We carried our dissection along the cystic duct toward the portal vessels. The peritoneum was open at the porta hepatis and the hepatic artery and its branches identified. We then followed the atretic bile duct remnant to the edge of the duodenum. This was ligated at the border of the duodenum and divided. We then dissected the bile duct remnant toward proximal to the surface of the liver until the proximal fibrous cone became visible. We dissected the peritoneum around the fibrous cone anteriorly and laterally all the way to the branches of the hepatic artery. There were two small lateral branches of the hepatic arteries entering into the fibrous cone from lateral, which were divided using monopoly cautery. The dissection was carried as far lateral as possible to maximize the exposed liver surface covered by the

portoenterostomy. The peritoneum was then dissected posteriorly behind the bifurcation of the portal vein. Two portal vein branches, one from each portal vein, were seen entering the posterior aspect of the fibrous cone. Both branches were ligated with 6-0 Prolene and cautery. The peritoneal dissection was then completed behind the bifurcation of the portal vein, which was retracted with a vein retractor. The biliary remnant (fibrous cone) was then removed from the liver surface with sharp scissors. Injury to the surface of the liver was avoided and there was minimal bleeding. The biliary remnant was sent to pathology clearly marked for correct orientation. A fresh liver wedge biopsy was then performed at the edge of the liver. Two 4-0 PDS sutures were placed and the wedge was taken with a sharp knife between the two sutures. The liver surface was then cauterized at the cut surface for hemostasis. A small Gelfoam sponge was then placed onto the bare hepatic plate above the portal vein bifurcation and the liver returned back into the abdomen. We then identified a loop of jejunum about 10 cm distal to the ligament of Treitz and made a window in the mesentery. The jejunum was divided with an Endo GIA stapler and the mesentery further divided down to its root. A 30-cm-long jejunal Roux loop was measured. A side-to-side jejuno-jejunostomy was performed with a 4-0 PDS running suture after anti mesenteric enterotomies were opened. The anastomosis was oriented to direct the stream of enteric contents toward the distal small bowel. The mesenteric gap was then closed with interrupted Vicryl sutures. A window was opened through the retrocolonic mesentery to the right of the middle colonic vessels, and the Roux limb was brought through the window toward the liver. The liver was then again extroverted from the abdomen using the skin corners as retractors. The Gelfoam sponge was carefully removed and the portal plate inspected for bleeding. An antimesenteric enterotomy was opened just proximal to the end of the Roux loop. The end-to-side portoenterostomy was then performed using interrupted 6-0 Maxon

sutures. The portal plate area on the liver surface covered by the portoenterostomy was maximized behind the portal vein bifurcation and laterally just next to the hepatic artery branches on both sides. The liver back was then placed back into its original position in the abdomen. The Roux limb was then oriented at its optimal position through the transverse colonic mesentery, and the Roux limb was fixated with three sutures into the retrocolonic mesenteric window. The correct positioning and orientation of the Roux limb and the jejunio-jejunostomy were then reconfirmed one last time. The abdomen was irrigated with warm saline.

The subcostal abdominal incision was then closed in a running manner in two layers. Scarpa's fascia was approximated and the skin was closed with a subcuticular running stitch. Upon completion of the procedure, a debriefing checklist was completed to share information critical to the postoperative care of the patient. The patient tolerated the procedure well, was extubated in the operating room, and was transported to the post-anesthesia care unit in stable condition.

Laparoscopic Kasai Portoenterostomy

After definite diagnosis of biliary atresia by means of laparoscopic cholangiogram, three additional trocars are placed into the right and left abdomen. The liver was retracted upwards with a liver retractor and a transabdominal suture placed through the gallbladder remnant, which was left in place until the end of the procedure. The cystic artery was divided with hook cautery and followed toward the portal vessels. Careful blunt dissection and hook cautery was used for tissue dissection. The peritoneum over the porta hepatis was opened and the hepatic artery and its branches identified. The bile duct remnant was followed toward the edge of the duodenum and ligated there with a clip. We then followed the bile duct remnant toward the surface of the liver where the fibrous cone was identified. The peritoneum of the

fibrous cone was then dissected anteriorly and laterally to the branches of the hepatic artery. The peritoneum was then decided below the fibrous cone behind the bifurcation of the portal vein. The two portal vein branches entering into the fibrous cone were identified and cauterized. The biliary remnant was then removed from the liver surface with sharp scissors without injury to the surface of the liver. A percutaneous liver biopsy was then performed with a biopsy gun. Hemostasis was achieved at the entrance site with cautery. The biliary remnant was sent to pathology together with the fresh liver biopsy. A Gelfoam sponge was placed onto the bare hepatic plate. The umbilical incision was then extended up- and downwards and a neonatal wound retractor placed. The small bowel was exteriorized. A loop of jejunum about 10 cm distal to the ligament of Treitz was identified and a window made into the mesentery. The jejunum was divided with an Endo GIA stapler and the mesentery divided further to its root. A 30-cm-long Roux loop was measured. A side-to-side jejunio-jejunostomy with a 4-0 PDS running suture directing the stream of enteric contents toward distal. A 5 mm Endo-stapler was used to perform a side-to-side jejunio-jejunostomy, and the enterotomy was closed with interrupted Vicryl sutures. The mesenteric gap was closed with Vicryl sutures. The bowel back was placed back into the abdomen and the wound retractor removed. The umbilical incision was approximated with interrupted sutures and the umbilical trocar placed back into the abdomen. Laparoscopically, a window was made through the retrocolonic mesentery to the right of the middle colic vessels and the Roux limb pass through the window toward the liver. The Gelfoam sponge was gently removed from the liver surface and the portal plate inspected for any bleeding. An antimesenteric enterotomy just proximal to the end of the Roux loop was then opened with scissors. An end-to-side portoenterostomy was then completed using running 6-0 Maxon sutures for the back row and interrupted sutures for the front row maximizing the covered area

on the liver surface. The Roux limb into was then optimally positioned through the transverse colonic mesentery and fixated with three sutures. The orientation of the Roux limb and the jejuno-jejunostomy was reconfirmed one last time. The gallbladder was then removed with hook cautery and sent to pathology. The abdomen was irrigated with warm saline. All instruments and trocars were removed from the

abdomen. The fascia of all small abdominal incisions was closed as well as the skin in multiple layers.

A debriefing checklist was completed to share information critical to the postoperative care of the patient. The patient tolerated the procedure well, was extubated in the operating room, and was transported to the postanesthesia care unit in stable condition.