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CASE REPORT

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Laparoscopic Kasai portoenterostomy for biliary atresia

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Abstract Conventional surgery for extrahepatic bile-duct atresia (EHBDA) usually requires a large, painful, muscle-cutting laparotomy, dislodgment of the liver, and wide manipulations, followed by adhesions and possible complications that may disturb the postoperative course and hamper liver transplantation (LT). The main role of laparoscopy in EHBDA has been for diagnostic purposes. Besides all the advantages of minimally-invasive access, it allows excellent visibility and dissection of tiny hilar structures. The authors present the first two cases of successful Roux-en-Y laparoscopic portoenterostomy (LARP) for EHBDA, showing the importance of advanced technical skills and a new approach for extracorporeal enteroanastomosis. Laparoscopic hilar dissection and portoenterostomy was accomplished using four trocars. The umbilical site was used for extracorporeal Roux-en-Y enteroenterostomy, in the first case using a laparoscopic stapler and in the second a hand-sewn suture. Mean operative time was 190 min, and no operative complications were observed. Both girls became anicteric. The first is doing well 15 months after the operation with good hepatic function. The other was anicteric for 6 months, had one episode of cholangitis, developed an umbilical hernia, has shown slow and progressive hepatic failure, and is now being evaluated for possible LT. It is concluded that LARP for EHBDA can be done safely in infants using an extracorporeal transumbilical enteric anastomosis, with several advantages compared with open surgery. The role of LARP in facilitating LT is yet to be defined.

Keywords Biliary atresia · Laparoscopy · Hepatic portoenterostomy · Child · Roux-en-Y anastomosis

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Introduction

The surgical treatment of biliary atresia (BA) is still a great challenge for pediatric surgeons. Despite some controversies about the ideal age or age limit to indicate biliary drainage, standard operations include large, painful, muscle-cutting laparotomies to accomplish hilar dissection and one of several portoenterostomy (PEO) techniques, usually Kasai's operation [1]. Children with BA are operated upon with some degree of malnutrition and hepatic dysfunction, and show a high rate of perioperative complications due to surgical trauma, including pain, nerve damage, respiratory compromise, prolonged ileus, wound dehiscence, hernias, and peritoneal adhesions. Most of them will require liver transplantation (LT) due to intrahepatic progression of the disease, and substantial time may be spent to reopen the abdomen and release peritoneal and hilar adhesions while trying to avoid visceral damage and control blood loss[1, 14].

The incidence of all these complications could be minimized by minimally invasive access via laparoscopy. Pediatric laparoscopic instruments allow many delicate operations, so that laparoscopic Roux-en-Y portoenterostomy (LARP) could be of great use in infants with BA. However, this approach has been reported only for the diagnosis of BA to date.

We discovered that an umbilical approach could be used for safe extracorporeal Roux-en-Y enteric anastomosis in babies with laparoscopic assistance to localize the critical points. We could not find any report of this approach being used for this purpose. We report the first two infants with BA treated successfully by laparoscopy

and transumbilical extracorporeal Roux-en-Y reconstruction.

Case reports

Case 1 A 46-day-old girl who weighed 4.3 kg was admitted with a history of jaundice, choluria, and fecal acholia since birth. She was born by normal delivery at 39 weeks' gestation weighing 3.0 kg. The pregnancy had been uneventful and the mother's serologic studies were normal except for a slightly elevated IgG level for cytomegalovirus (166 ng/dl), which was also present in the baby (90 ng/dl). Laboratory data by the age of 20 days showed that total bilirubin (Tbil) was 13.8 mg/dl (normal 0.2–1.0); direct bilirubin (Dbil), 7.6 mg/dl; alanine aminotransferase (ALT) 94 IU/l (normal 5–32); aspartate aminotransferase (AST) 90IU/l (normal 9–32); alkaline phosphatase (AP) 340 IU/l (normal); and gamma-glutamyltransferase (GGT) 570 IU/l (normal 5–35). Other metabolic and hematologic test results were normal. At the age of 30 days she showed hepatomegaly, failure to thrive, signs of influenza, and intense cholestatic jaundice. An ultrasound scan (US) showed a small gallbladder, no intra- or extrahepatic bile ducts, and suggested the presence of a possible fibrous preportal triangle. Parenteral administration of vitamins A, D, E, and K was started.

A percutaneus liver biopsy at 38 days showed signs of BA and laparoscopic exploration was elected. Dbil was 13.6 mg/dl, albumin 2.1 g/dl (normal 3.5–5), and prothrombin time (PT) 92% (normal 70–100). Preoperative management included daily doses of vitamin K (5 mg) since the biopsy, fasting, a saline enema (80 ml) 24 h before operation, dimeticone orally every 4 h for 1 day, IV amikacin 7.5 mg/kg body weight every 12 h, and metronidazole 10 mg/kg every 8 h, the last doses 1 h prior to anesthesia.

The procedure was performed using general anesthesia with sevofurane, fentanyl and atracurium with the patient positioned at the foot of the table, the surgeon at the patient's feet, the assistant with the camera at the left, and the second assistant and instrument nurse at the right. A 4-mm incision was made in the inner infraumbilical ring and the peritoneum was opened under direct vision. A 3.5-mm trocar with CO_2 valve was introduced by an open technique into the abdomen, which was insufflated to 8–10 mmHg with CO_2. An additional 3.0 mm trocar was placed under direct laparoscopic vision in the left upper quadrant between the hemiclavicular and anterior axillary lines. A 3.5-mm 30° laparoscope was used.

The operation was divided into 5 stages:

1. *Confirmation of extrahepatic bile-duct atresia* by inspection or cholangiography; in this case, laparoscopy confirmed type III BA with a small, fibrotic gallbladder not amenable to cholangiography, a moderately cirrhotic appearance of the liver, mildly enlarged peritoneal collateral vessels, and no other anomalies. The circular umbilical incision was enlarged to 11 mm for a 10-mm reusable trocar, and two additional 5-mm trocars were placed under direct vision in the right mid- and lower abdominal wall. In order to rotate the liver upward and backward for better hilar exposure, the round and falciform ligaments were incised and two 3-0 chromic catgut sutures tied the hepatic borders to the abdominal wall at the level of the costal margin.

2. *Dissection of the biliary remnants and porta hepatis*: the gallbladder and all extrahepatic ducts were freed using a laparoscopic monopolar fine-needle cautery especially designed for delicate surgery (like the one used for Peña's anorectoplasty) followed by resection of enlarged hilar lymph nodes and blunt dissection close to the hepatic arteries and portal vein. Downward traction of the duodenum and frequent suction were performed by the assistant using the lower right port. Small arterial branches were ligated and very small vessels were cauterized, including the short portal tributaries under the cone-shaped porta hepatis (Fig. 1). This was widely exposed just under the portal bifurcation and entry point of the hepatic arteries, then transected just under the liver surface with a laparoscopic knife and scissors, leaving a crude surface with two visible tiny ducts draining bile, followed by *en bloc* resection of the biliary remnants and temporary covering of the crude area with moist gauze.

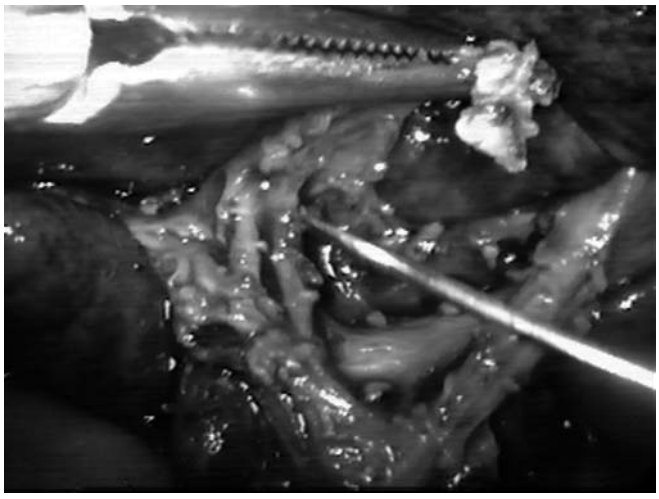

Fig. 1 Porta hepatis dissected with fine cautery needle

3. *Extracorporeal transumbilical jejunal Roux-en-Y anastomosis*: the jejunum was secured 15 cm from Treitz' ligament and drawn to the umbilical port under direct vision and withdrawn through the umbilicus. The pneumoperitoneum was interrupted to relieve tension on the mesentery as necessary; the jejunum was transected and the distal end closed temporarily with one suture. Another segment 30 cm beyond the distal end was selected by successive in-and-out movement of the distal bowel after the tied end had been replaced into the abdomen. Stay sutures identified the proximal and distal segments. The correct position of the intestine was checked by laparoscopy to avoid torsion; the bowel was again withdrawn for extracorporeal jejunal anastomosis. A 45-mm linear laparoscopic stapler (Endopath ET45B, Ethicon, Johnson and Johnson) was used for side-to-side approximation and a 2.0-cm transverse hand-sewn interrupted suture joined the proximal jejunal end and the stapler hole, closing the defect.

4. *PEO*: the tied proximal jejunum was passed through the transverse mesocolon, the knot cut, and the end anastomosed around the porta hepatis with delicate extra- and intracorporeal interrupted 5-0 polyglactin sutures (Fig. 2); the mesenteric defects were closed with 5-0 polyglactin.

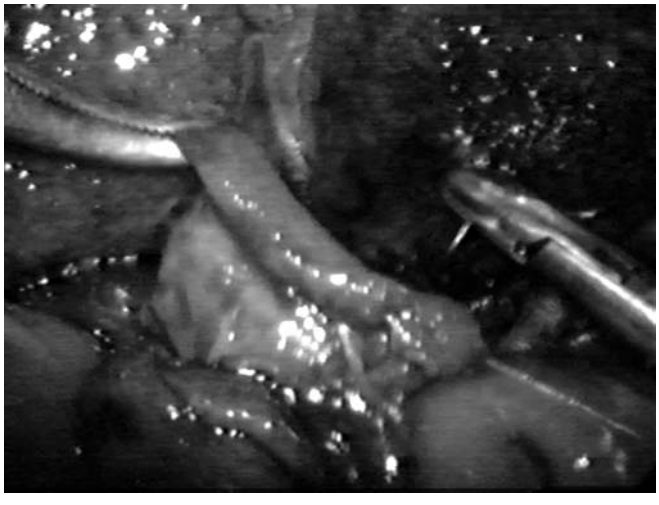

Fig. 2 Laparoscopic end-portoenterostomy

5. *Percutaneus liver biopsy* with a Tru-cut needle was performed. The suspending sutures of the liver were cut; the upper abdomen was drained by a Penrose drain exteriorized through the upper right trocar site. The other abdominal incisions were sutured at the fascia and the skin was closed with adhesive tape.

The operative time was 195 min. The infant needed no respiratory support postoperatively. She stayed in the intensive care unit (ICU) for 1 night, passed stools by the 2nd postoperative day, and began feedings of breast milk by the 3rd day. She received antibiotics (amikacin and metronidazole) for 48 h. Jaundice progressively declined until day 7, when she showed mild cholangitis (fever, acholia) treated by amikacin and metronidazole for 7 days. She had no complications due to the laparoscopy. After 15 months she was anicteric, weighing 9.5 kg, with stable hepatic and renal function on a low-fat diet.

Case 2 A 68-day-old girl who weighed 5.2 kg was admitted with a history of jaundice, choluria, and fecal acholia since the 4th day of life. She was born by cesarean delivery at 38 weeks' gestation weighing 3.1 kg. The pregnancy had been uneventful except for hyperemesis in the first trimester. The maternal and neonatal serologic tests were normal. Laboratory data by the age of 16 days showed Tbil 16.0 mg/dl; Dbil 8.8 mg/dl; ALT 145 IU/l; AST 170 IU/l AP 605 IU/l; GT 820 IU/l; and PT 25%. Other metabolic and hematologic test results were normal. Ten days later US showed a small gallbladder possibly containing fluid, no intra- or extrahepatic bile ducts, and a small, hyperechogenic porta hepatis. A percutaneus liver biopsy at another institution at 32 days suggested neonatal hepatitis; the child was kept under observation for almost a month, did not improve, and was refered to us for cholangiography at 65 days. Parenteral administration of vitamins A, D, E, and K was started. Since the PT had reached 90% 3 days later with other normal hematologic and biochemical parameters, laparoscopic exploration was planned. The preoperative management was the same as in case 1.

Laparoscopy revealed a moderately cirrhotic liver, a small, elliptical gallbladder, two large hilar lymph nodes, and type III atretic extrahepatic bile ducts. Percutaneous transhepatic cholecystography confirmed a closed, atretic gallbladder, and the operation was carried out as described above through step 2. An umbilical 10-mm trocar was used with a 3.5-mm laparoscope, and three other 5.3-mm trocars positioned as in case 1. The round ligament was not cut. At the porta hepatis no macroscopic ducts could be seen even after a deeper incision. During step 3, the transumbilical extracorporeal enteric Y-anastomosis was an end-to-side, one-layer, seromuscular, hand-sewn anastomosis with 4-0 polyglactin sutures (Fig. 3). The remaining procedures were the same as described above.

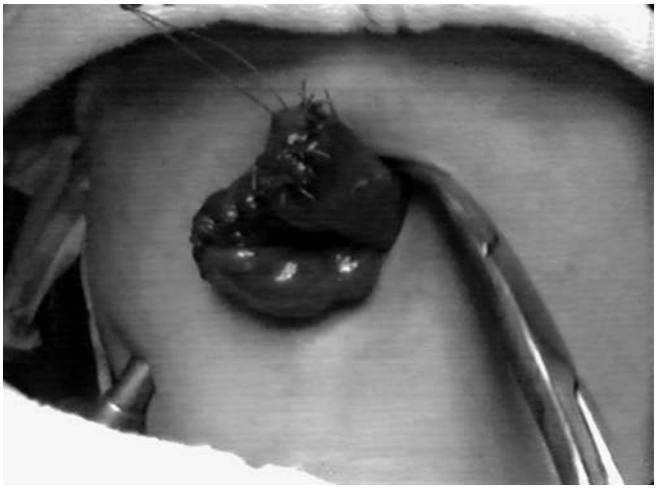

Fig. 3 Transumbilical Roux-en-Y jejunal anastomosis with hand-sewn suture

The duration of the procedure was 180 min. The baby stayed in the ICU for 2 days without respiratory support, passed stools by the 2nd postoperative day, and began feedings of breast milk by the 3rd day. She had no complications due to the laparoscopy. Antibiotics (amikacin and metronidazole) were given for 48 h and she was discharged home 6 days after the operation. The jaundice slowly decreased, but she had persistent Dbil around 1.5 mg/dl. After 2 months she developed diarrhea and cholangitis (fever, increased jaundice, acholia), and was treated in hospital with amikacin and metronidazole for 7 days. Discrete jaundice and recurrent fecal hypocholia have persisted. She developed an umbilical hernia and 8 months after surgery showed signs of moderate hepatic failure, and is being evaluated for LT. At 9 months during operation for umbilical hernia, laparoscopy showed only few adhesions at the hepatic hilum.

Discussion

Kasai's hepatic PEO or its variations are still the first and main procedures indicated for neonates and infants with BA, because almost 25% of affected children have a good life expectancy with stable hepatic function and bile drainage after these operations. LT is indicated at a later time if symptoms of end-stage liver disease occur [1, 9, 13]. A PEO provides a better chance to alleviate the progression of biliary cirrhosis until LT, and a previous Roux-en-Y anastomosis is already prepared for biliary connection.

Conventional surgical procedures to gain some bile drainage into the intestine in children have been achieved through large laparotomies in the upper abdomen, one of the largest incisions in pediatric abdominal surgery. Possible complications related to this approach include pain, breathing limitation leading to pulmonary complications, nerve damage, prolonged ileus, wound dehiscence, large or recurrent incisional hernias, rib damage, peritoneal adhesions, and the risk of retained surgical sponges. Dehiscence and hernias are usually favored by hypoproteinemia, ascites, and frequent crying, which are common features in infants with BA. Moreover, dense adhesions under the large scar along most of the abdomen can jeopardize reoperation for LT, when substantial time may be necessary to release or suture compromised intestinal segments, requiring larger blood transfusions [14].

The use of minimally-invasive access via laparoscopy could avoid or decrease these complications when adequately performed. Its advantages compared with laparotomies have been repeatedly demonstrated in both adults and children, showing significantly lower incidences of adhesions [12], related complications, pain, and postoperative breathing difficulty and resulting in very small scars [8]. Amplified, well-illuminated visibility of the tiny hilar structures is guaranteed by laparoscopy. Another advantage is that the whole operation can be recorded and subsequently reviewed by the transplantation team.

Laparoscopy has been widely applied to treat common biliary diseases in both adults and children, especially for cholecystitis and gallstones [3, 6]. During

the last few years, the use of laparoscopy in children with BA has been limited to diagnosis with or without cholangiography and liver biopsy [2, 7, 15]. After reviewing the literature, we concluded that this is the first report of LARP for BA. All standard steps of the operation, including hilar dissection to remove atretic extrahepatic bile ducts, exposure of the porta hepatis, PEO, and Roux-en-Y bilioenteric anastomosis could be accomplished in almost the same time as with open surgery, with no operative complications. We designed a special needle cautery like the one we had been using in open surgery for biliary diseases and anorectal anomalies, which helped in removing the atretic ducts and cauterizing tiny hilar vessels. A fine aspirator tip is important to constantly clean the operative field without removing much CO_2. Another useful aspect is lifting and suturing the fibrotic liver high on the anterior abdominal wall to expose the hilum without making extra ports. To do this and rotate the liver upward, it is not always necessary to cut the falciform and round ligaments.

Nowadays, special attention to anesthesia and careful monitoring of CO_2 in neonatal and infantile videosurgery allow safe procedures in conjunction with advanced surgical skills. LARP is not an easy operation: it should be done by a surgeon with adequate experience in laparoscopic hand-suturing and neonatal laparoscopy. Even in adults, laparoscopic biliodigestive anastomosis is rarely described [10]. The only reported pediatric laparoscopic bilioenteric Roux-en-Y anastomosis was used in Japan in a 6-year old girl with a choledocal cyst (CC) [16]. Two other cases of CC operated upon laparoscopically were reported in adults [5, 17]. No laparoscopic bilioenteric operation has been described in patients as young as ours with BA; however, we believe it is feasible with adequate instruments in experienced hands.

We have noticed that almost any small-bowel segment can be laparoscopically selected, secured, and exteriorized through an umbilical incision in children, allowing extracorporeal resection and anastomosis for Meckel's diverticulum, enteric duplications, intussusception, and enteric biopsies [4]. A semilunar umbilical incison coupled with an enlarged fascial defect becomes a sufficiently wide laparotomy incision for small-bowel exteriorization and anastomosis, not even requiring an omega-shaped amplification at the skin as used for hypertrophic pyloric stenosis [11]. The extracorporeal techniques are easier and faster than laparoscopic sutures, especially when using laparoscopic staplers extracorporeally, as we did in case 1, avoiding the use of an extra 12-mm port. The cost of the operation is lower if no stapler is used, as we did in case 2. To our knowledge, the technique of extracorporeal transumbilical jejunal anastomosis to assist laparoscopic Roux-en-Y reconstruction has not been described in the literature. This approach would also be useful for other reconstructions or small-bowel resections (for example, choledoeal cyst).

Although only two cases are described, we conclude that LARP for BA can be done safely in infants in association with extracorporeal transumbilical enteric anastomosis, with some advantages compared with open surgery. The role of LARP in facilitating LT is yet to be defined.

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