**Name of Journal:** *World Journal of Clinical Cases*

**Manuscript NO:** 65103

**Manuscript Type:** CASE REPORT

**Neonatal biliary atresia combined with preduodenal portal vein: A case report**

Xiang XL *et al*. Biliary atresia with PD-PV

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**Supported by** The Science Foundation of Suzhou Science and Technology Bureau, No. SYS201758 and No. SYS2020158; and Youth Science and technology project of revitalizing health by science and education in Suzhou in 2019, No. KJXW2019020.

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**Received:** February 28, 2021

**Revised:** May 28, 2021

**Accepted:** July 20, 2021

**Published online:** September 6, 2021

**Abstract**

BACKGROUND

Congenital biliary atresia is a type of obstruction of the bile ducts inside and outside the liver, which can lead to cholestatic liver cirrhosis and eventually liver failure. The preduodenal portal vein (PD-PV) is a rare developmental malformation of the PV. The PV courses in front of the duodenum. However, very few cases of neonatal biliary atresia combined with PD-PV have been reported in the scientific literature.

CASE SUMMARY

A 1-mo-and-4-d-old child was admitted to the hospital in January because of yellowish skin. After surgical consultation, surgical intervention was recommended. The child underwent Hilar-jejunal anastomosis, duodenal rhomboid anastomosis, and abdominal drainage under general anesthesia. During the operation, the PV was located at the anterior edge of the duodenum.

CONCLUSION

Diagnoses: (1) Congenital biliary atresia; (2) PD-PV; and (3) Congenital cardiovascular malformations. Outcomes: Recommendation for liver transplantation. Lessons: The choice of treatment options for neonatal biliary atresia combined with PD-PV.

**Key Words:** Neonatal; Biliary atresia; Preduodenal portal vein; Treatment; Case report

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**Citation:** Xiang XL, Cai P, Zhao JG, Zhao HW, Jiang YL, Zhu ML, Wang Q, Zhang RY, Zhu ZW, Chen JL, Gu ZC, Zhu J. Neonatal biliary atresia combined with preduodenal portal vein: A case report. *World J Clin Cases* 2021; 9(25): 7542-7550

URL: https://www.wjgnet.com/2307-8960/full/v9/i25/7542.htm

DOI: https://dx.doi.org/10.12998/wjcc.v9.i25.7542

**Core Tip:** Congenital biliary atresia is a type of obstruction of the bile ducts inside and outside the liver, which can lead to cholestatic liver cirrhosis and eventually liver failure. The preduodenal portal vein is a rare developmental malformation of the portal vein. A 1-mo-and-4-d-old child was admitted to the hospital in January because of yellowish skin. After surgical consultation, surgical intervention was recommended. The child underwent laparoscopic exploration under general anesthesia. During the operation, the portal vein was located at the anterior edge of the duodenum.

**INTRODUCTION**

Congenital biliary atresia accounts for half of the cases of long-term neonatal obstructive jaundice. Among the surviving infants, its incidence rate is 1:8000-1:14000. However, there is large variation in the incidence across races and geographic regions. Most cases have been reported in Asia, where the incidence rate in the Eastern population is 4-5 times higher than that in the Western population, and the ratio of affected males to females is 1:2. Congenital biliary atresia is a type of obstruction of the bile ducts inside and outside the liver, which can lead to cholestatic liver cirrhosis and eventually liver failure[1,2]. The preduodenal portal vein (PD-PV) is a rare developmental malformation of the PV. The PV courses in front of the duodenum, and the incidence of PD-PV is about 1:10000[3,4]. However, very few cases of neonatal biliary atresia combined with PD-PV have been reported in the published scientific literature[5].

**CASE PRESENTATION**

***Chief complaints***

A female infant, G1P1, had a gestational age of 37 wk and 4 d. On the fourth day after birth, the baby developed yellowing of the facial skin, which progressively aggravated. The skin over the trunk region also turned yellowish, and it progressed further.

***History of present illness***

On the fourth day after birth, the baby developed yellowing of the facial skin, which progressively aggravated. The skin over the trunk region also turned yellowish, and it progressed further. Admission diagnoses were: (1) Neonatal hepatitis syndrome; (2) Abnormal liver function; and (3) Congenital cardiovascular malformations. The patient’s body temperature was normal, all vital signs were stable, and the stool was still light yellow and slightly whitish. The electroconvulsive therapy report from the outside hospital showed that the liver function was disrupted with biliary obstruction. After surgical consultation, surgical intervention was recommended.

***History of past illness***

She was admitted to the Neonatology Department with the diagnosis of “newborn jaundice” in the outpatient clinic. The meconium passed by the child was resolved within 24 h after birth, and it turned yellow within 2-3 d. Bowel movements occurred 1-2 times a day, and the color of stool was pale yellow, without clay colored stool. Urine was normal.

***Personal and family history***

A female infant, G1P1, had a gestational age of 37 wk and 4 d.

***Physical examination***

Upon admission, clinical examination showed body temperature: 37 ºC, pulse: 140 beats/min, respiratory rate: 40 beats/min, weight: 3760 g, clearly conscious, good reaction, crying loudly, steady breathing, moderate yellowing of the skin on the face, trunk, and limbs, the sclera was yellowish, and the skull was not deformed. There were no special features in the face. The fontanelle measured about 2.0 cm × 2.0 cm, and it was flat. The nose did not move, the lips were not cyanosed, the neck was soft, the breath sounds of both lungs were thick, and no dry or wet rales were heard. The heart rhythm was uniform, the heart sound was medium, no murmur was heard, and the abdomen was soft. The liver was located 2 cm below the ribs, and it did not touch the spleen. Bowel sounds were normal, 3-4 sounds *per* minute. The umbilical cord had fallen off, the umbilicus was dry, and the umbilical chakra was not red. The muscle tension of the limbs was normal, and the foraging and sucking reflexes could be elicited.

***Laboratory examinations***

Outpatient examination of liver function revealed: γ-glutamyl transpeptidase: 114.5 U/L, total protein: 58.5 g/L, albumin: 58.5 g/L, prealbumin: 104 mg/L, globulin: 15.6 g/L, albumin-globulin ratio: 2.75, high-sensitivity C-reactive protein: 0.32 mg/L, glutamic-pyruvic transaminase: 91.3 U/L, glutamic oxaloacetic transaminase: 166.4 U/L, indirect bilirubin: 100.02 μmol/L, direct bilirubin: 129.88 μmol /L, total bilirubin: 229.9 μmol/L; and a normal TORCH test.

***Imaging examinations***

Color Doppler ultrasound showed no obvious abnormal echo in the liver, gallbladder, pancreas, and kidneys. Heart Doppler ultrasound revealed interruption of the inferior vena cava and continuation of the odd vein, persistence of the left superior vena cava, and a patent foramen ovale.

**MULTIDISCIPLINARY EXPERT CONSULTATION**

After surgical consultation, surgical intervention was recommended.

**FINAL DIAGNOSIS**

(1) Congenital biliary atresia; (2) PD-PV; and (3) Congenital cardiovascular malformations.

**TREATMENT**

The child underwent laparoscopic exploration under general anesthesia. During the operation, the PV was located at the anterior edge of the duodenum (Figure 1). Intraoperative diagnosis was PD-PV. Upon exploring the gallbladder, it was found that the gallbladder was poorly developed and had the shape of a cord. Intraoperative cholangiography showed that the intrahepatic bile duct was visualized by percutaneous puncture catheter-based injection of the contrast agent, but the biliary tract system was not clearly visualized, the duodenum was not visualized, and there was no contrast agent in the abdominal cavity. The intestinal loops were filled with gas and were dilated (Figure 2). Then the child was switched to open surgery, and the hilar tissues were carefully dissected. The fibrous mass in the hilar tissue was freed, and the fibrous mass and part of the liver parenchyma were removed. A light yellow bile secretion was noted. The gallbladder and choledochal cyst wall were removed from the trocar of the umbilical cord, and the mesangium was repaired. Hilar-jejunal anastomosis was performed.

**OUTCOME AND FOLLOW-UP**

The child’s symptoms were gradually relieved, and then she was discharged. During follow-up, the child’s condition gradually improved, but deterioration of the child’s condition could not be ruled out, which would require liver transplantation or other treatments. Finally, the child was lost to follow-up due to change in contact information of the child’s family.

**DISCUSSION**

The relationship between neonatal biliary atresia and PD-PV should be considered in depth. In this study, the baby developed yellowing of the facial skin, which progressively aggravated. The skin over the trunk region also turned yellowish, and it progressed further. The child was admitted to the neonatology department with the diagnosis of “newborn jaundice” in the outpatient clinic. Further investigation showed that the liver function was disrupted due to biliary obstruction. Intraoperative cholangiography showed that the intrahepatic bile duct was visualized by percutaneous puncture catheter-based injection of the contrast agent, but the biliary tract system was not clearly visualized, the duodenum was not visualized, and there was no contrast agent in the abdominal cavity. The intestinal loops were filled with gas and were dilated. On laparoscopic exploration, the PV was located at the anterior edge of the duodenum. Upon exploring the gallbladder, it was found that the gallbladder was poorly developed and had the shape of a cord. Based on the above findings, postoperative diagnoses of congenital biliary atresia and PD-PV were established. After surgical treatment combined with drug therapy and other comprehensive treatments, the child’s symptoms were gradually relieved, and then she was discharged.

Biliary atresia of the bile duct in the first part of the duodenum before the PV can occur individually, as found in such cases, or it can occur in combination. It is categorized as deformation of the congenital anatomic structure. There are a limited number of published studies describing the correlation of biliary atresia and PD-PV with the onset of deformity. Given the paucity of reported cases, the current study provides timely insights with regard to planned treatment and outcomes for this combination syndrome.

***Features of PD-PV***

Since 1921, Knight first described PD-PV[6], and so far, less than 100 cases have been reported in the literature. Each report often presents the case of only 1 child, and most of the reports have presented cases of only 5 children, which indicates that the deformity is very rare. Its true incidence cannot be accurately calculated because in some cases PD-PV does not produce any clinical symptoms before it is detected.

PD-PV originates from the persistent primordial yolk vein or is related to abnormal rotation of the midgut. For example, abnormal intestinal rotation and duodenum and stomach reversal may result in PD-PV[7]. Three quarters of children with PD-PV have some concomitant malformations, such as cardiovascular malformations, gastrointestinal malformations, and biliary malformations[8] or are considered to be part of other syndromes, such as polysplenia or heterotaxy. Isolated PD-PV, including symptomatic or asymptomatic PD-PV, accounts for only a quarter of all cases[9].

PD-PV is considered to be an external cause of congenital duodenal obstruction. Researchers have been studying how a low-pressure blood vessel can cause thick-walled duodenal obstruction[7]. In most cases, duodenal obstruction is caused by other related deformities, and PD-PV is just an accompanying deformity. In the study by Vilakazi *et al*[8], in only 10 cases duodenal obstruction in children was found to be caused by PD-PV alone. PD-PV may cause complete or partial duodenal obstruction. Characteristically, vomiting can occur within a few hours after birth, and feeding cannot be tolerated. Partial duodenal obstruction presents with repeated episodes of vomiting and growth retardation. Snavely andBreakell[10] reported that PD-PV caused portal hypertension, variceal bleeding in the fundal venous plexus, and death of the patient. Autopsy revealed that the PV became narrowed due to abnormal position, which affected the blood flow of the PV and caused portal hypertension.

***PD-PV diagnosis and treatment***

Preoperative diagnosis of PD-PV is very rare. This disease entity may not be discovered until childhood or adulthood, or it may be discovered accidentally on abdominal computed tomography. A total of 5%-10% of children with biliary atresia are accidentally diagnosed with PD-PV during surgery. PD-PV combined with the anterior common bile duct of the duodenum has also been reported; however, cholecystectomy poses a great risk for children with PD-PV[11], or it is accidentally discovered during cholecystectomy in adults[12].

The prenatal diagnosis of congenital duodenal obstruction is based on obvious polyhydramnios and the double bubble sign displayed by B-ultrasound. The PD-PV is a cause of prenatally diagnosed duodenal obstruction, established by B-ultrasound, which has not been found in the literature presented in domestic and foreign reports[13].

Abdominal color Doppler ultrasound and computed tomography can be used in cases with a clear diagnosis of duodenal obstruction before surgery. If the vascular structure is found in the front of the pancreas, it has an important diagnostic value. PD-PV is a rare cause of duodenal obstruction. It is not necessary to diagnose PD-PV before surgery because all children with duodenal obstruction require laparotomy or laparoscopic exploratory surgery. However, it is very important to identify PD-PV during the operation because PD-PV occasionally does not cause obstruction, but it may only be discovered accidentally during the operation, which may result in intraoperative complications, especially in children with intestinal rotation or abnormal internal organs. These unconventional anatomical positions put the children at risk of iatrogenic injuries, including iatrogenic bleeding from abnormal veins or damage to the bile duct and dilated duodenum.

Duodenal obstruction has the potential to progress to a surgical emergency. However, children should not undergo surgery immediately. Instead, they should receive gastrointestinal decompression, oxygen inhalation, electrocardiogram monitoring, and fluid rehydration. Surgery should be performed after the child achieves hemodynamic stability and electrolyte balance. Due to high incidence of malformations related to this disease entity, which is similar to that in splenic abnormalities[14], it is necessary to conduct a systematic review of children to detect malformations, including evaluation of heart malformations. Cases of children with biliary atresia are extremely rare. The only treatment for PD-PV-induced duodenal obstruction is surgical treatment, and the normal anatomical relationship should be restored as much as possible. The clear surgical method is duodenal rhomboid anastomosis in front of the PV or diversion surgery, such as gastroduodenal anastomosis in front of the PV. Thorough examination of the abdominal cavity should be performed to rule out other related malformations. Dissociation of the anterior wall of the duodenum needs to be carefully performed to avoid damage to the duodenum and PV. The duodenal papilla should be avoided when the proximal obstruction is cut open. For duodenal rhomboid anastomosis, the duodenal incision edge should not be very close to the PV to avoid stenosis of the PV. During the operation, the location of the obstruction must be accurately judged to determine the surgical approach. Due to complicated biliary atresia, the patient of the current study underwent “duodenal rhomboid anastomosis + duodenal jejunal Roux-en-Y anastomosis.”

The genetic origin of PD-PV is still unclear. Although it is very rarely found in clinical practice, it is a likely cause of fetal or infantile duodenal obstruction and may cause a potential risk to surgery; thus, it should receive the attention of clinicians.

***Surgical treatment of biliary atresia***

Kasai radical resection opened a new era of “uncorrectable” biliary atresia treatment. To date, Kasai radical resection is still the preferred surgical method for biliary atresia, and liver transplantation is a treatment method in case of failure of advanced Kasai radical resection[15,16]. Kasai radical surgery emphasizes early diagnosis and treatment; the age of surgery should be around 60 d, and the maximum age should not exceed 90 d[17].

The key to Kasai radical operation is to completely remove the hepatic hilar fibrous mass. The operation is best performed under a surgical magnifying glass, so that the side of the cut section reaches the liver parenchyma at the entry of the PV and the longitudinal level reaches the posterior wall of the PV. The depth of removal of the hilar fibrous mass is the key to this operation. Very superficial excision may not ensure reaching the appropriate small intrahepatic bile duct, and very deep excision may cause damage to the liver parenchyma and affect the healing of the surgical anastomosis. Generally, only a thin layer of the membrane is preserved on the liver surface when the hilar fibrous mass is removed; secondly, electrocoagulation should be performed cautiously to stop bleeding from the incision, especially when the left and right liver ducts enter the liver parenchyma. Compression may also be performed, and it has partial hemostasis effect.

**Various modified surgical approaches:** After the classic portojejunostomy described by Kasai, although many modified procedures have been proposed to reduce the possibility of complicated cholangitis, the results are not ideal. The most commonly used modifications include external drainage and intussusception type anti-reflux valve placement. However, neither the “ventilation” nor the “valve” method has much effect on reducing the incidence of retrograde cholangitis. In the early 1990s, some scholars proposed that intussusception anti-reflux valve can reduce the occurrence of reflux cholangitis after biliary atresia. However, more recent studies have shown that targeting the regurgitant valve may be effective for anti-reflux but less effective at preventing cholangitis. A possible explanation for this dichotomy is that cystic dilatation of the intrahepatic bile duct accompanied by cholestasis has become a potential target for bacterial colonization. Therefore, although the regurgitant valve works, infection cannot be avoided.

**Views on the application of laparoscopy:** With the widespread application of laparoscopy, there are related reports on laparoscopy for radical operation of biliary atresia, but its clinical efficacy remains to be explored. Because biliary atresia is a rare disease, individual physicians find it difficult to accumulate surgical experience. For hilar operations with abundant blood supply, electrocoagulation may also affect the bile flow of the remaining microbiliary ducts in the hilar region, which will also affect the postoperative efficacy. Therefore, the author believes that the radical operation of laparoscopic biliary atresia should be cautiously performed. Laparoscopy for cholangiography is indeed a minimally invasive method. If the ability to perform radical surgery for biliary atresia is limited, then it is not recommended to only perform cholangiography and then arbitrarily conclude that the extrahepatic biliary system in the hilar area has completely disappeared. This is due to more than 90% of children with biliary atresia developing hilar fibrous masses within 3 mo when the hilum is isolated.

**Drug treatment after biliary atresia surgery:** Effective drug treatment is extremely important for improving the prognosis after portoenterostomy. Although surgery can prolong the lifespan of children, it cannot reverse liver damage and progressive cirrhosis. Ultimately, 75%-80% of children need liver transplantation for long-term survival[18,19]. In recent years, it has been recognized that the immune-mediated damage of the bile duct and liver may be related to the onset of biliary atresia and the progressive deterioration of liver function after surgery. It is possible to change the course of the disease through drug adjuvant therapy.

**Postoperative hormone therapy:** Corticosteroids, the main component of adjuvant therapy, can significantly improve the quality of life after surgery and increase the survival. Due to the inflammatory nature of cholangitis itself and the abnormal immune mechanism, it may be related to the onset of biliary atresia. Theoretically, the application of drugs, such as steroids, after hepatoenteric anastomosis should be very effective in reducing immune-mediated liver damage, improving bile drainage, and reducing the incidence of reflux cholangitis. Since Gad *et al*[20] reported that short-term shock therapy with glucocorticoids can increase bile flow, many treatment institutions have adopted short-term shock therapy for 1 to 2 wk after surgery. Dillon *et al*[21] proposed that compared with the non-hormonal group, oral high-dose steroids [prednisone 4 ms/(kg·d), initial] in combination with ursodeoxycholic acid and antibiotic treatment at 6-22 wk after surgery can effectively enhance the bile clearance rate of children and improve the survival rate of autologous liver within 5 years. Meyers *et al*[22] introduced the application of 10, 8, 6, 5, 4, 3, and 2 ms/(kg·d) prednisone *via* the intravenous route for 7 d, followed by oral prednisone 2 ms/(kg·d). The method of continuous 8-12 wk application is also believed to significantly improve bile drainage and increase the survival time of children with autologous liver compared with the hormone-free group. Wang *et al*[23] summarized the application results of long-term use of high-dose steroids. Compared with short-term shock therapy, steroids can improve the short-term bile drainage in children with biliary atresia and reduce the incidence of cholangitis; however, the effect of prolonging the survival time of autologous liver has not been clearly demonstrated. Complications and safety during the use of hormones require further observation and evaluation. In any case, the application of hormones after biliary atresia is widely performed.

**Long-term application of choleretic drugs after surgery:** In addition to hormones, choleretic drugs also include dehydrocholic acid, glucagon, dinoprostone, and ursodeoxycholic acid. Among them, ursodeoxycholic acid has been studied in depth. It can significantly improve the deficiency of essential fatty acids and reduce the level of bilirubin. It is currently used as a routine drug and has provided good effects. No adverse reactions have been reported. It is clinically recommended to take ursodeoxycholic acid 10 mg/(kg/d) orally. Ursodeoxycholic acid is started after the operation and usually continued for 1 to 2 years. There are also reports of oral administration throughout life.

**The application of prophylactic antibiotics after surgery:** In the early 1980s, the second-generation cephalosporins (cephalosporin and cefuroxime) were combined with aminoglycosides (gentamicin and amikacin). After the 1990s, third-generation cephalosporins became dominant, and they were occasionally combined with aminoglycosides. The third-generation cephalosporins reach a sufficient level in the bile through the passive secretion pathway. Other advantages are that they can be administered at intervals of 12 to 24 h, which provides convenience for home treatment. Previous drug sensitivity tests have proved the effectiveness of cefoperazone and ceftriaxone. Unfortunately, in recent years, the sensitivity of cefoperazone in the treatment of cholangitis after Kasai operation has dropped from 88.9% to 75.0%[24,25], which increases the need to identify new first-line antibiotics. It has been reported that trimethoprim/sulfamethoxazole and neomycin can reduce the incidence of cholangitis. According to Bu *et al*[26], these drugs can reduce the recurrence rate of cholangitis to 9.1% and 7.5%, respectively, and the first episode of cholangitis was delayed from 3 mo after surgery to 6 and 7 mo after surgery, thereby improving survival.

***Liver transplantation and biliary atresia***

With the development of liver transplantation, the prognosis of biliary atresia has greatly improved. According to current reports on liver transplantation at home and abroad, biliary atresia is the most common indication. The average survival time of children with biliary atresia without surgery is 12 mo. After Kasai surgery, more than half of the children have repeated postoperative infections, and the survival rate is only 30% to 60%. Since Strong *et al*[27] reported the success of the first liver transplantation for extrahepatic biliary atresia, more than 90% of children with successful liver transplantation have developed biliary atresia. Some scholars have proposed whether to perform liver transplantation directly to reduce hilar adhesions after Kasai operation, which causes difficulties during liver transplantation. It is still unclear whether the treatment of biliary atresia should be to directly perform liver transplantation or to perform liver transplantation after Kasai surgery; however, the current view is that treatment should be considered based on the child’s condition.

Kasai surgery and liver transplantation complement each other; children whose age is less than 90 d should undergo Kasai surgery first. If there is no bile flow or only temporary bile drainage after the operation, and the histological examination of the hilar region of the liver shows that the biliary tract has a small caliber and a small number of ducts, these children do not need to undergo the Kasai operation because repeated operations increase the difficulty of future liver transplantation. If the child is older than 90 d and there is no obvious chronic liver disease, then the hepatic hilar region can be dissected first to determine whether there are residual liver ducts. If there are open residual liver ducts, then the Kasai operation can be performed; otherwise liver transplantation should be performed. If the child has any obvious liver disease, such as liver cirrhosis and portal hypertension, then liver transplantation should be performed. Even if the bile drainage is satisfactory after the Kasai operation and the jaundice has gradually reduced, close follow-up should be performed over a long time. If liver disease occurs, liver transplantation should be performed as soon as possible.

**CONCLUSION**

In short, Kasai surgery is the first choice for treatment of biliary atresia, which may allow the child to achieve healing or buy precious time for liver transplantation. Postoperative comprehensive drug treatment plays an important role in improving the efficacy, and the success of liver transplantation significantly improves prognosis. However, it is very important to deepen our understanding of the etiology of biliary atresia, strive to improve the level of early diagnosis, and continuously improve the technique of portoenterostomy and perioperative management.

**ACKNOWLEDGEMENTS**

The authors thank all the medical workers who helped us (Children’s Hospital of Soochow University).

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**Footnotes**

**Informed consent statement:** All study participants, or their legal guardian, provided informed written consent prior to study enrollment.

**Conflict-of-interest statement:** The authors have no conflicts of interest to disclose.

**CARE Checklist (2016) statement:** The authors have read the CARE Checklist (2016), and the manuscript was prepared and revised according to the CARE Checklist (2016).

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**Manuscript source:** Unsolicited manuscript

**Peer-review started:** February 28, 2021

**First decision:** April 14, 2021

**Article in press:** July 20, 2021

**Specialty type:** Medicine, research and experimental

**Country/Territory of origin:** China

**Peer-review report’s scientific quality classification**

Grade A (Excellent): 0

Grade B (Very good): B

Grade C (Good): C

Grade D (Fair): D

Grade E (Poor): 0

**P-Reviewer:** Kitamura K, Saad K **S-Editor:** Fan JR **L-Editor:** Filipodia **P-Editor:** Yuan YY

**Figure Legends**



**Figure 1 The child underwent laparoscopic exploration under general anesthesia.** During the operation, the portal vein was located at the anterior edge of the duodenum.



**Figure 2 Intraoperative cholangiography showed that the intrahepatic bile duct was visualized by percutaneous puncture catheter-based injection of the contrast agent, but the biliary tract system was not clearly visualized, the duodenum was not visualized, and there was no contrast agent in the abdominal cavity.**



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