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# Cystic Biliary Atresia Combined with Gallbladder Duplication: A Case Report

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# Abstract

Congenital biliary atresia (BA) is a disease involving obstruction of both intrahepatic and extrahepatic bile ducts, leading to cholestatic liver disease, progressive hepatic fibrosis and eventual liver failure. A specific subtype, cystic biliary atresia (CBA), is characterized by the presence of a cyst near the porta hepatis or in the proximal extrahepatic duct. However, as this cyst does not communicate with the intrahepatic biliary tree, it fails to alleviate progressive bile stasis and hepatic fibrosis. Additionally, gallbladder duplication is a rare biliary malformation where an accessory gallbladder is situated adjacent to or connected with the primary one. Notably, the cooccurrence of neonatal CBA and gallbladder duplication is exceedingly rare. To our knowledge, no similar literature has been reported.


# Introduction


Congenital biliary atresia (BA) is a disease involving obstruction of both intrahepatic and extrahepatic bile ducts, leading to cholestatic liver disease, progressive hepatic fibrosis and eventual liver failure. A specific subtype, cystic biliary atresia (CBA), is characterized by the presence of a cyst near the porta hepatis or in the proximal extrahepatic duct. However, as this cyst does not communicate with the intrahepatic biliary tree, it fails to alleviate progressive bile stasis and hepatic fibrosis. Additionally, gallbladder duplication is a rare biliary malformation where an accessory gallbladder is situated adjacent to or connected with the primary one. Notably, the cooccurrence of neonatal CBA and gallbladder duplication is exceedingly rare. To our knowledge, no similar literature has been reported.


# Case report

### Clinical history

A 12-day-old female infant was admitted with a prenatally diagnosed choledochal cyst. Physical examination was normal. Transabdominal ultrasound revealed a cystic structure at the porta hepatis and duplicated gallbladders with communication between them. The child underwent Hilar-jejunal anastomosis, duodenal rhomboid anastomosis, and abdominal drainage under general anesthesia. The gallbladder exhibited a bilobed duplication anomaly containing white transparent bile. Blood tests showed a white blood cell count of $7.50\times10^{9}$/L and a red blood cell count of $5.31\times10^{12}$/L.

### Laboratory findings

**Gamma-Glutamyl Transferase** (**GGT**): 538U/L, Alanine Aminotransferase(ALT): 20 U/L,total protein: 54 g/L,albumin: 34.5 g/L, prealbumin: 95mg/L, globulin: 19.5g/L, albumin-globulin ratio:1.8, high-sensitivity C-reactive protein:0.65 mg/L, Total bilirubin **( TBIL)**: 240.2 μmol/L;Direct (conjugated) bilirubin(**DBIL**): 75.14 μmol/L;Indirect bilirubin: 165.14 μmol/L.

### Imaging findings

Transabdominal ultrasound revealed two principal findings. First, a well-defined cystic lesion, measuring approximately 29× 16mm, was seen at the porta hepatis (Fig. 1a). Second, gallbladder duplication was noted, with the two separate gallbladders measuring approximately40 ×7 mm and35 × 7 mm, respectively (Fig. 1b, c). Both gallbladders demonstrated anechoic contents with good through-transmission, and communication between them was observed. Sonographic imaging showed two separate cystic ducts that converged into a single common cystic duct just before joining the common bile duct (Figs. 1d). Based on these findings, a final diagnosis of choledochal cyst (CCs) accompanied by gallbladder duplication was established.

**Treatment**

The infant underwent laparoscopic biliary exploration under general anesthesia at 15 days of age. However,Intraoperative findings revealed a red-colored liver with mild nodular hardening on the surface and visible fine vascular markings (Fig. 2a). The gallbladder exhibited a bilobed duplication anomaly containing white transparent bile(Fig. 2b). The common bile duct was cystically dilated, measuring approximately 3.0 cm in diameter. Aspiration of both the gallbladder and the common bile duct yielded white transparent bile. Based on these hepatic features, a diagnosis of biliary atresia (Type I, cystic variant) was established. Consequently, excision of the hilar cyst and Roux-en-Y hepaticojejunostomy were performed.The gallbladder and cyst were completely resected along the biliary bed. The two gallbladder cavities were observed to converge into a single cystic duct, which communicated with the cyst(Fig. 2b). The distal portion of the cyst was obstructed cephalad to the level of the pancreatic head, with no patent pathway identified. After removal of the cyst, the proximal common hepatic duct measured approximately 0.2 cm in diameter, with a small amount of light yellow bile drainage observed(Fig. 2c). A retrocolic end-to-side hepaticojejunostomy was subsequently performed, along with the construction of an anti-reflux valve. The procedure was completed successfully, and no intraoperative complications occurred. The pathological examination of the liver showed grade G2S3 cholestatic hepatitis with bridging fibrosis, marked by hepatocyte degeneration and necrosis, bile plug formation, portal fibroplasia, and inflammatory cell infiltration. Histological examination of the duplicated gallbladder revealed significant wall fibrosis accompanied by vascular congestion and inflammatory infiltrates. Focal epithelial denudation was also noted. Overall, the pathological features were diagnostic of cystic biliary atresia presenting within a double gallbladder anomaly.he patient was doing well at the 1-month postprocedure visit.

# Discussion

BA is a progressive, obstructive cholangiopathy of early infancy of unknown etiology. Clinically, differentiating CBA from CCs remains a formidable task for pediatric surgeons due to their striking similarities in initial presentation. Literature suggests that over half of CBA cases are preoperatively identified as CCs, underscoring the diagnostic challenges in neonates. Despite their distinct prognoses, the overlapping clinical and ultrasonographic patterns of CBA and CCs often make an accurate preoperative differentiation difficult..[1,2].Both of them are the common causes of neonatalcholestasis in

pediatric surgery and presented with a cyst at the hepatic hilum which could be detected prenatally by ultrasound .The two diseases share similar ultrasonographic patterns and clinical manifestations but with a different prognosis. The prognosis of BA is largely dependent on early surgical[3]. CBA, a subset of BA, is confined to presentation in early infancy. CCs is a well-established entity that may present soon after birth or later in infancy, in older children and in adults. The incidence of CBA and isolated CCs in the first 90 days of life is roughly similar. Both may be detected before birth by fetal ultrasound that may also detect signs of coexisting atresia. The cysts in CBA tend to be smaller and based on imaging studies are stated to be more spherical than fusiform;TBIL and total serum bile acids(TBA) are higher in BA than in isolated CCs. A small gallbladder and preoperative liver biopsy showing unequivocal evidence for LDO in an infant favors CBA over CCs[4]. According to the histopathologic criteria established by Lobeck et al., CBA is distinct from CC in its lack of a well-defined epithelial layer and the presence of a unique dense cicatricial inner layer characterized by myofibroblastic proliferation. Our case aligns with these findings, specifically showing a fibrotic cyst wall devoid of the organized smooth muscle structures typically retained in choledochal cysts[2]. Literature review indicates that the overall degree of liver function impairment in children with CCs is typically milder than in those with CBA, and the severity of cholestasis is also often lower. Consistent with our findings, multiple previous studies reported that levels of ALT, Aspartate Aminotransferase(AST), TBIL, and DBIL are typically significantly higher in CBA than in CCs. The markedly elevated liver function indicators observed in this patient align more closely with the characteristic profile of CBA, which may serve as a crucial reference for clinical differentiation. This dilemma is very clearly illustrated by the example of cystic BA: A study conducted in the UK reported that patients with CBA underwent surgical intervention approximately 10 days earlier than those with isolated BA (47 days vs. 58 days of life; $p = 0.02$)[5]. In contrast, a German study observed a slightly delayed surgical timing in children with CBA compared to those with the isolated form (63 days vs. 60 days of life; $p = 0.2$)[1]. While ultrasound is an excellent first-line tool, intraoperative cholangiography remains the definitive diagnostic procedure. It can conclusively map the biliary anatomy, confirm the absence of communication between the cyst and the intrahepatic bile ducts in CBA, and delineate the insertion pattern of the cystic ducts in a duplication.

Gallbladder duplication is an uncommon malformation of the biliary tract. with an estimated incidence of approximately 1 in 3,800–4,000 live births. To date, just over 210 cases have been documented in the literature [6,7]. The occurrence of a duplicated gallbladder is essentially a developmental anomaly arising during early embryogenesis. Specifically, during the 5th to 6th week of gestation, an atypical bifurcation or over-division of the hepatic diverticulum's caudal portion can lead to the formation of supplemental gallbladder primordia8. Anatomical variants of this condition are traditionally categorized using Boyden’s system. In our patient, the gallbladder presented a Y-shaped configuration—a 'true duplication' where two cystic ducts merge into a singular conduit before reaching the common bile duct(CBD), as opposed to the independent H-shaped entry pattern9. For the present case of a Y-shaped duplication associated with CBA, accurate preoperative identification was particularly crucial for surgical strategy. This required addressing both the biliary atresia (typically with a Kasai portoenterostomy) and the complete excision of the duplicated structure. Although transabdominal ultrasound serves as a valuable initial screening

tool, delineating the complex biliary anatomy in such combined anomalies often necessitates intraoperative cholangiography. This procedure provides definitive mapping, confirming the cystic duct configuration and the absence of communication between the cyst and intrahepatic ducts in CBA[9].Increasing awareness among sonographers of the specific features of duplication—such as two separate sacs and cystic ducts—is essential to raise preoperative suspicion. The differential diagnosis includes Phrygian cap shaped gallbladder, hepatic cyst adjacent to the gallbladder, Ladd's band which may twist the gallbladder, perivesicular effusion,and Todani II bile duct cyst (bile duct diverticulum) [7,10]. Beyond identifying two separate gallbladder sacs, sonographic clues to duplication include the presence of two distinct cystic ducts or the 'double-channel' sign. In the context of suspected BA, meticulous scanning is required to differentiate a true duplicated gallbladder from the cyst in CBA or other entities in the differential list. Although gallbladder duplication has previously been reported in association with duodenal atresia, to our knowledge, this case represents the first report of gallbladder duplication coexisting with biliary atresia.

In conclusion, The coexistence of CBA and gallbladder duplication is exceedingly rare, presenting a unique diagnostic challenge. CBA should be highly suspected if ultrasonography indicates the combination of a small subhepatic cyst, an atrophic gallbladder, the triangular cord sign, along with elevated laboratory values (ALT, AST, GGT,TBil, DBil, TBA). The unexpected finding of a potential duplicated gallbladder in this context should alert the clinician to this rare possibility. Cholangiography should be performed expediently for any suspected CBA to confirm the diagnosis and allow for timely surgical intervention.

## Declarations

This research received no specific grant from any funding agency in the public, commercial, or not-for-profit sectors.

# Consent for publication

Written informed consent was obtained from the parents/legal guardians of the patient for publication of this case report and any accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

# Author Contribution

A(LUAN Shengwei) and B(CHEN Shuai) share co-first authorshipC(GUO Fagang) provided surgical images.D(LIU Xiaofang )and E (LIU Qinghua )offered guidance on manuscript writing.F(YAN Yuxi )served as the corresponding author and provided guidance on manuscript writing.

## References

1. Madadi-Sanjani O, Rohrbacher V, Uecker M. Cystic Biliary Atresia—A Single-Center, Retrospective Analysis. Dtsch Arztebl Int. 2024;121(19):641–2. 10.3238/arztebl.m2024.0114.
2. Lobeck IN, Sheridan R, Lovell M, Dupree P, Tiao GM, Bove KE. Cystic Biliary Atresia and Choledochal Cysts Are Distinct Histopathologic Entities. Am J Surg Pathol. 2017;41(3):354–64. 10.1097/PAS.0000000000000805.
3. Yu P, Dong N, Pan YK, Li L. Comparison between cystic biliary atresia and choledochal cyst: a clinical controlled study. Pediatr Surg Int. 2022;38(1):109–14. 10.1007/s00383-021-05004-y.
4. Lobeck IN, Sheridan R, Lovell M, Dupree P, Tiao GM, Bove KE. Cystic Biliary Atresia and Choledochal Cysts Are Distinct Histopathologic Entities. Am J Surg Pathol. 2017;41(3):354–64. 10.1097/PAS.0000000000000805.
5. Davenport M, Caponcelli E, Livesey E, Hadzic N, Howard E. Surgical outcome in biliary atresia: etiology affects the influence of age at surgery. Ann Surg. 2008;247(4):694–8. 10.1097/SLA.0b013e3181638627.
6. Boyden EA. The accessory gall-bladder: an embryological and comparative study of aberrant biliary vesicles occurring in man and the domestic mammals. Am J Anat. 1926;38:177–231. doi.10.1002/aja.1000380202.
7. Darnis B, Mohkam K, Cauchy F, Cazauran JB, ,Bancel B, Rode A, et al. A systematic review of the anatomical findings of multiple gallbladders. HPB (Oxford). 2018;20(11):985–91. 10.1016/j.hpb.2018.04.002.
8. Vezakis A, Pantiora E, Giannoulopoulos D, et al. A Duplicated Gallbladder in a Patient Presenting with Acute Cholangitis. A Case Study and a Literature Review. Ann Hepatol. 2019;18(1):240–5. 10.5604/01.3001.0012.7932.
9. Huang XZ, Liu J. A case of double gallbladder with cholelithiasis diagnosed with transabdominal ultrasound. J Med Ultrason (2001). 2023;50(2):263–264. 10.1007/s10396-023-01287-x
10. Joshi HO, Poudel D, Khatiwoda P, Adhikari BB, Kunwar L. Gall bladder duplication: A rare biliary malformation. Radiol Case Rep. 2024;20(3):1492–5. 10.1016/j.radcr.2024.11.057. Published 2024 Dec 21.

# Figures

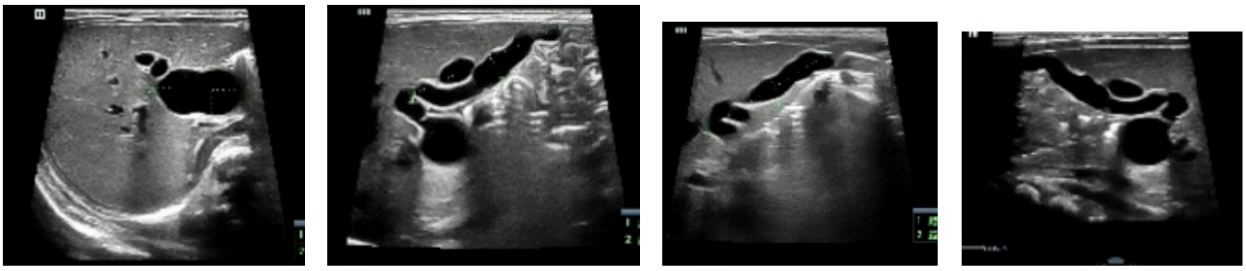


**Figure 1**

Transabdominal ultrasound in CBA patients.

(A) CBA: hepatic hilar cyst. (B) gallbladder duplication:one size 40 ×7 mm (C) gallbladder duplication :another size 35 × 7 mm(D) Y-shaped cystic duct configuration

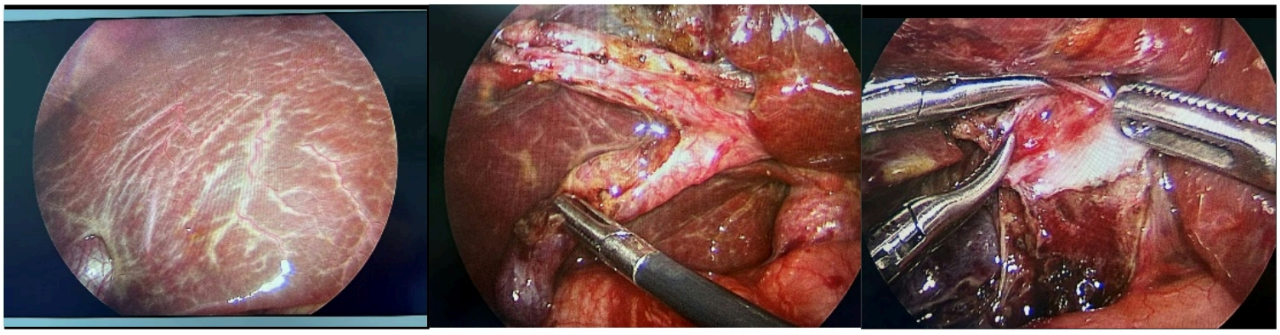


**Figure 2**

Laparoscopic in CBA patients.

(A) The liver surface exhibits mild nodular hardening with visible fine vascular markings.

(B)Intra-operative picture showing duplication of gallbladder.

(C) Intraoperative photograph showing the exposed orifice of the common hepatic duct after cyst excision (or resection).