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Case Report

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Coexistence of choledochal cyst and annular pancreas in a child: A rare case report and literature review

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Abstract

Choledochal cyst (CDC) and annular pancreas (AP) are rare congenital anomalies of the pancreatobiliary system, and their coexistence is exceedingly uncommon, with only a few pediatric cases reported. Recognition of this association is important, as it may influence clinical presentation, surgical planning, and long-term outcomes. Herein, we reported a two-year-old female who presented with recurrent abdominal pain and nonbilious vomiting. Laboratory investigations revealed elevated pancreatic enzyme levels, and magnetic resonance cholangiopancreatography demonstrated a fusiform dilatation of the common bile duct consistent with Todani type 1 CDC. The child underwent laparoscopic cyst excision, during which an AP encircling the duodenum was unexpectedly identified. To avoid the risk of future duodenal obstruction, hepaticoduodenostomy was not performed, and laparoscopic Roux-en-Y hepaticojejunostomy was completed. The postoperative course was uneventful, and at the three-year follow-up, the child remained asymptomatic. A review of the literature revealed only 10 previously reported pediatric cases of CDC associated with AP, most presenting in the neonatal period with duodenal obstruction. This case is unique in that the AP was clinically silent and discovered only intraoperatively, underscoring the need for surgical vigilance when managing such rare associations.

Keywords: Annular pancreas, choledochal cyst, laparoscopic surgery, pancreaticobiliary maljunction, roux-en-y hepaticojejunostomy.

Choledochal cysts (CDCs) are rare congenital malformations of the bile duct characterized by abnormal dilatations of the extrahepatic and/or intrahepatic biliary tree and are usually associated with pancreaticobiliary maljunction.[1] Its incidence varies globally, reaching as high as 1 in 1,000 in Southeast Asia, particularly Japan, compared to 1 in 100,000-150,000 in Western countries. It predominantly affects females, with a female-to-male ratio of approximately 3-4:1.[2] Annular pancreas (AP), a recognized cause of extrinsic duodenal obstruction, is another rare congenital anomaly in which pancreatic tissue encircles the descending duodenum, causing partial or complete obstruction.[3] Its incidence is estimated at 1 in 12,000-15,000 live births and 0.005 to 0.015% in adult autopsy series. It has been reported across all age groups, with no consistent sex predilection.[4]

Although CDC and AP are embryologically related entities,[5] their concomitant occurrence is extremely rare, with only a few cases reported in the literature. Herein, we reported a rare case of CDC associated with AP in a two-year-old child, along with a review of the relevant literature.

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CASE REPORT

A two-year-old female, the second child born at term via vaginal delivery following an uncomplicated pregnancy with no notable antenatal concerns,

was asymptomatic at birth and thereafter. Over the preceding six months, the child developed recurrent episodes of colicky, moderate to severe upper abdominal pain associated with nonbilious vomiting, resulting in multiple hospital admissions for treatment. There was no history of fever, weight loss, poor appetite, jaundice, urinary complaints, or blood transfusion. Bowel and bladder habits were normal, and there was no significant dietary or family history. Developmental milestones and immunization status were appropriate for age. On examination, the weight and height were 11 kg and 85 cm respectively, both at the 50th percentile, with no evidence of failure to thrive. Tenderness was present in the right upper abdomen, although no palpable lump was detected. A written informed consent was obtained from the parent of the patient.

The routine blood investigations, including complete blood count, liver and kidney function tests, were within normal limits, except for serum amylase and lipase levels, which were elevated to 254 U/L (normal range: 30-110 U/L) and 742 U/L (normal range: 20-120 U/L), respectively. The patient was diagnosed with acute pancreatitis and managed conservatively. On abdominal ultrasonography, the common biliary duct was dilated with no evidence of calculus. Magnetic resonance cholangiopancreatography (MRCP) showed fusiform dilatation of the common biliary duct with smooth distal tapering, with a common channel and a bulky pancreas (CDC type 1; Figure 1a, b). The patient was discharged and advised to undergo elective surgery.

After an interval of eight weeks, the child was taken up for elective laparoscopic excision of CDC with a planned hepaticoduodenostomy. A standard four-port technique was used, comprising a 10-mm umbilical camera port and three 5-mm working ports. A large fusiform cyst measuring approximately 5×4 cm, consistent with Todani type 1 CDC, was visualized. The cyst was carefully mobilized circumferentially and dissected distally up to the point of tapering. After application of ligaclips at the distal end, the cyst was transected proximally just distal to the confluence of the right and left hepatic ducts. During partial mobilization of the duodenum for hepaticoduodenostomy, annular pancreatic tissue encircling the second part of the duodenum was unexpectedly identified, which had not been detected on preoperative imaging and was mislabelled as a bulky pancreas (Figure 2a, b). In view of this intraoperative

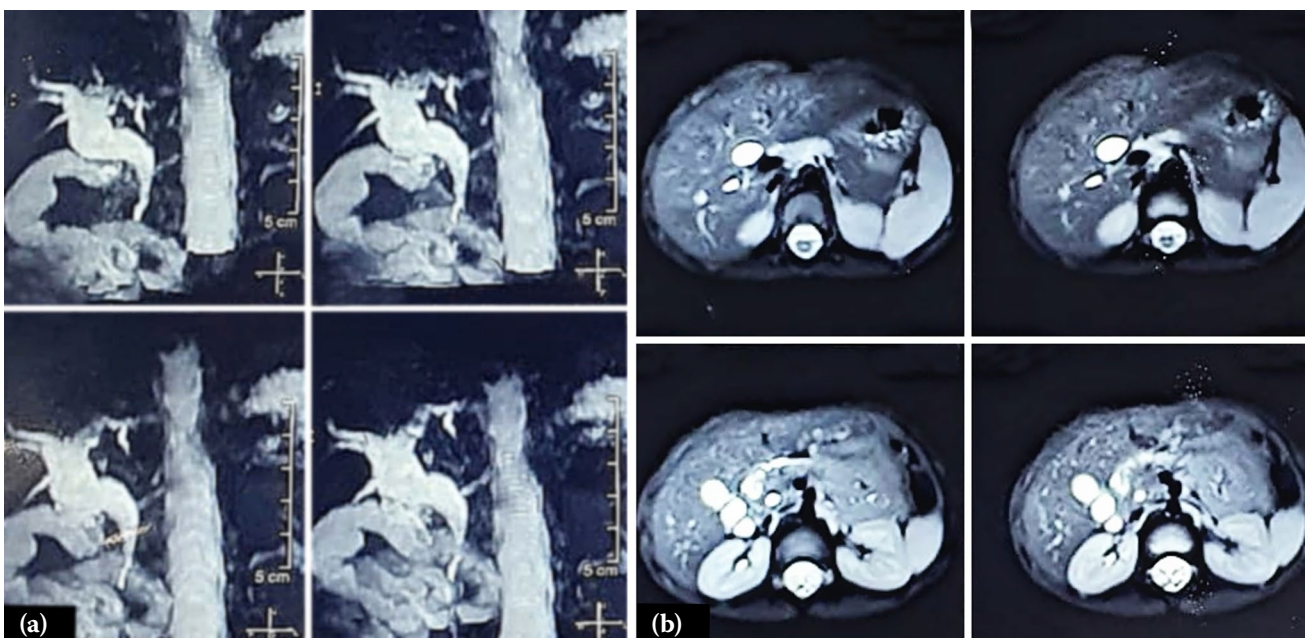


Figure 1. Preoperative MRCP images. **(a)** Coronal view showing a fusiform dilatation of the common bile duct consistent with Todani type 1 choledochal cyst. **(b)** Axial images demonstrating cystic dilatation of the extrahepatic bile duct. The presence of AP was not clearly appreciated on MRCP.

MRCP: Magnetic resonance cholangiopancreatography; AP: Annular pancreas.

finding, a decision was made to proceed with Roux-en-Y hepaticojejunostomy (RYHJ) instead of hepaticoduodenostomy. A Roux limb was fashioned approximately 15 cm distal to the duodenojejunal flexure. The distal limb was brought up through a window created in the transverse mesocolon, and an end-to-side hepaticojejunostomy was performed using 5-0 Vicryl (RB needle) in a single-layer interrupted fashion, entirely intracorporeally (Figure 2c). The proximal Roux limb was then anastomosed to the distal limb approximately 20 cm distal to the hepaticojejunostomy in an end-to-side jejunojejunostomy, which was performed extracorporeally after exteriorizing the bowel loops through the umbilical port site, using 4-0 Vicryl (RB needle) in a single-layer interrupted fashion. A drain was placed in the Morrison's pouch through one of the 5-mm port sites, and all port sites were closed in layers. The total operative time was approximately 260 min. There were no intraoperative complications, and the estimated blood loss was minimal (approximately 10 mL).

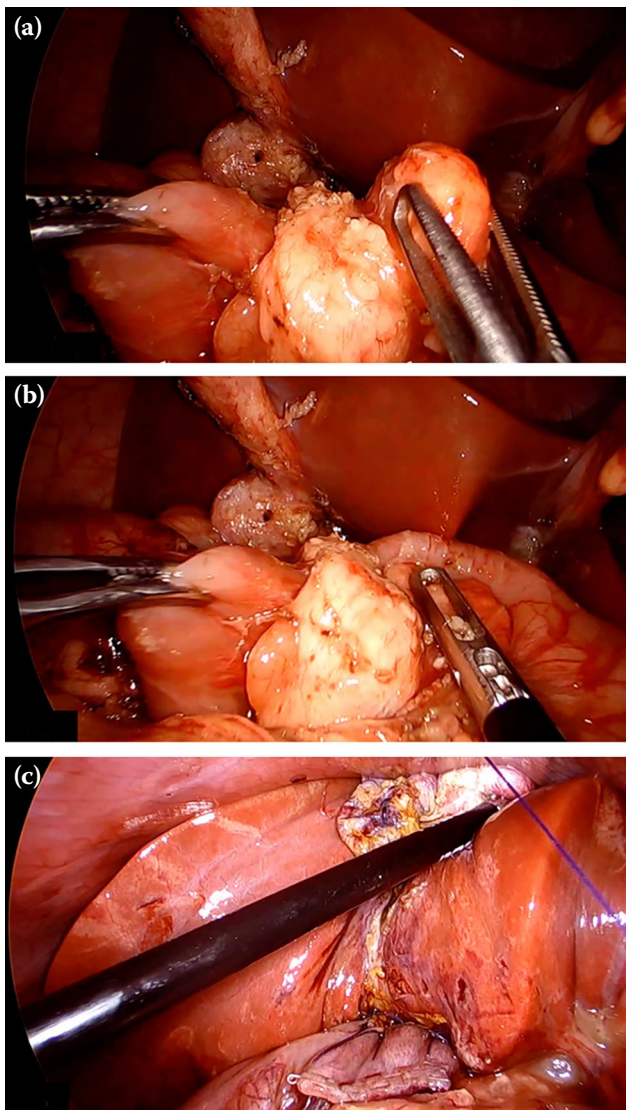


Figure 2. Intraoperative laparoscopic findings **(a, b)**. Dissection of the choledochal cyst with identification of annular pancreatic tissue encircling the duodenum **(c)**. Completed RYHJ following cyst excision.
RYHJ: Roux-en-Y hepaticojejunostomy.

Intraoperatively, there was no evidence of proximal duodenal or gastric dilatation, and the duodenal lumen appeared patent without luminal narrowing. Clinically, the child had no prior symptoms such as bilious vomiting, failure to thrive, weight loss, or poor appetite suggestive of chronic duodenal obstruction. These findings supported the diagnosis of a nonobstructive AP. Accordingly, the child underwent RYHJ alone, without any additional procedure for the AP, as it was found to be nonobstructive both clinically and intraoperatively.

The postoperative course was uneventful, and the patient was discharged on the sixth day. On histopathological examination, the gallbladder showed features of chronic inflammation. The wall of the CDC was composed of dense fibrous tissue lined by columnar or flattened epithelium, with areas of focal ulceration and denudation. Round cell infiltration, along with bile duct wall thickening and fibrosis, was also noted. There was no evidence of dysplastic or malignant changes. At the three-year follow-up (now five years old), the child remained well and asymptomatic, with no associated complaints.

DISCUSSION

Although CDC can occur at any age, about 80% of the cases are diagnosed in childhood.[2] The exact etiology is uncertain, but pancreaticobiliary maljunction with reflux of pancreatic secretions into the bile duct is considered a key factor, leading to chronic inflammation, wall damage, and cystic dilatation. Pancreaticobiliary maljunction-associated CDC is frequently complicated by hepatitis, cholangitis, pancreatitis, or histological inflammation.[1] Clinical presentation is variable,

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TABLE 1

Reported case of CDC associated with AP in the pediatric population

No	Author & year of the publication	Sex of the patient	Age of AP diagnosis & surgery	Surgery performed for AP	Associated duodenal lesion	CDC diagnosis-age & type	CDC surgery-age & type	Other anomalies	Outcome
1	Okada et al.[9] (1993)	Male	Antenatal polyhydramnios present, 2nd day	Duodeno-duodenostomy	DA	12 year, type 4	12 year, Roux-Y-choledochojejunostomy	–	Survived
2	Okada et al.[9] (1993)	Female	Antenatal polyhydramnios present, 7th day	Duodeno-duodenostomy	DA	3 year, type 4	3 year, Roux-Y-choledochojejunostomy	Jejunal aberrant pancreas	Survived
3	Komuro et al.[10] (2000)	Male	Both 11 months	Duodeno-duodenostomy	DS	Neonatal age, NA	11 month, Roux-Y-choledochojejunostomy	Intrapacreatic cyst	Survived
4	Sugimoto et al.[11] (2002)	Female	Antenatally diagnosed DA, 1st day	Duodeno-duodenostomy	DA	32 month, type 4	32 month, HD (open)	–	Survived
5	Oowari et al.[12] (2003)	Female	Both day 1	Duodeno-duodenostomy	DA	8 year, type 1	8 year, NA	–	Survived
6	Martin-Hirsel et al.[13] (2004)	Female	Antenatally diagnosed AP with polyhydramnios, 1st day	Duodeno-duodenostomy	–	Antentally diagnosed CDC, NA	Not done during 1st surgery	–	Survived
7	Shih et al.[14] (2005)	Female	Antenatal polyhydramnios present with DA, 3rd day	Duodeno-duodenostomy	DA	7 year, type 4	7 year, HD (open)	–	Survived
8	Iwai et al.[15] (2009)	Female	Antenatal polyhydramnios present, 2nd day	Duodeno-duodenostomy	DA	4 year, type 4	4 year, RYHJ (open)	–	Survived
9	Raman et al.[16] (2015)	Male	Antenatal polyhydramnios with single gastric bubble present, 3rd day	Duodeno-duodenostomy with Ladds procedure	–	Intraoperatively (3rd day), type 1	Intraoperatively (3rd day), RYHJ (open)	Malrotation	Survived
10	Downing et al.[17] (2020)	Female	Antenatal polyhydramnios present, 4th day	Duodeno-duodenostomy with Ladds procedure	DA	After birth, type 4	2 year, HD (open)	Malrotation	Survived
11	Index case (2025)	Female	Intraoperatively during CDC surgery	Not operated	–	2 year, type 1	2 year, RYHJ (laparoscopic)	–	Survived

CDC: Choledochal cyst; AP: Annular pancreas; DA, duodenal atresia; DS, duodenal stenosis; HD, hepaticoduodenostomy; NA, not available; RYHJ, Roux-en-Y-hepaticojejunostomy.

with nonspecific abdominal pain being common, while the classic triad of jaundice, abdominal pain, and right upper quadrant mass is rare. Other manifestations include jaundice, cholangitis, pancreatitis, portal hypertension, liver dysfunction, and coagulopathy.[1] Choledochal cyst has also been linked to other congenital anomalies, particularly cardiac malformations in infants and duodenal atresia, colonic atresia, gastroschisis, annular pancreas, and pancreatic cysts.[2]

Annular pancreas can present in neonates, infants, children, or adults, with 25% forming a complete and 75% a partial pancreatic ring. It results from abnormal embryonic fusion of the dorsal and ventral pancreatic buds during duodenal rotation, and such developmental errors may also involve the pancreatobiliary system, leading to associated malformations.[3,4] The clinical spectrum varies with the degree of duodenal obstruction and coexisting anomalies, ranging from asymptomatic cases to bilious vomiting in neonates/infants. In older children and adults, recurrent abdominal pain, pancreatitis, failure to thrive, and weight loss may predominate. Annular pancreas is frequently associated with congenital anomalies such as Down syndrome, duodenal atresia or stenosis, malrotation, imperforate anus, esophageal atresia, and congenital heart disease.[6]

Embryologically, CDC and AP share a close developmental relationship as they both arise from the foregut. The dorsal pancreatic bud forms the body and tail, while the ventral bud gives rise to the head and typically rotates to fuse with the dorsal bud. The ventral pancreas develops near the hepatic diverticulum and is carried dorsally with the bile duct during duodenal rotation, later fusing with the dorsal pancreas. Both structures are closely related and arise around the fourth week of embryogenesis, when the hepatic diverticulum is in the solid stage, with canalization beginning in the fifth week. Abnormal ventral bud migration or persistence may encircle the duodenum, resulting in AP, while defective canalization of the bile duct can lead to CDC.[7] Babbitt's hypothesis further links the CDC to the anomalous pancreaticobiliary junction, which promotes biliary stasis and ductal dilatation.[8] Several theories, including Lecco's (adhesion of the ventral bud to the duodenum) and Baldwin's (persistence of a left ventral bud), attempt to explain AP formation.[3] Since these developmental events occur simultaneously and in proximity, disruptions during this critical period may account for the rare coexistence of CDC and AP.

Reviewing the literature, we found only 10 reported pediatric cases of CDC associated with AP, making our index case the 11th.[9-17] Table 1 summarizes these reports. In 1993, Okada et al.[9] first described two cases from Japan, where both patients presented with duodenal obstruction due to AP in the neonatal period and underwent duodeno-duodenostomy, followed years later by surgery for CDC. Since then, most reports have described a similar pattern: AP diagnosed antenatally or at birth due to duodenal obstruction and treated surgically in the neonatal period, with CDC identified and managed at a later stage. A consistent observation from the literature is that AP is usually symptomatic early in life, most often with duodenal atresia or stenosis and antenatal polyhydramnios. Choledochal cyst is then diagnosed later, commonly as Todani type 4 or 1, and managed with hepaticojejunostomy or hepaticoduodenostomy. Associated anomalies, such as malrotation, jejunal heterotopic pancreas, intrapancreatic cyst, and congenital heart disease, have also been described.[9,10,16,17] Importantly, all reported patients, including ours, survived after surgical management.

Our case is distinct in that the AP remained asymptomatic, likely due to partial encirclement of the duodenum, and therefore was not suspected preoperatively. Preoperative MRCP did not clearly demonstrate AP, and the anomaly was detected only intraoperatively during laparoscopic excision of the CDC and duodenal mobilization. This highlights the diagnostic challenge of identifying AP in the absence of neonatal duodenal obstruction and underscores the importance of intraoperative vigilance.

The preoperative MRCP images were retrospectively reviewed and did not clearly demonstrate an AP. No direct radiological evidence of annular pancreatic tissue encircling the duodenum was identified, apart from a doubtful bulky pancreatic head. This limitation may be attributed to the absence of dedicated multiplanar reconstructions and to the fact that partial or nonobstructive AP often consists of a thin band of pancreatic tissue incorporated into the duodenal

wall, which may not be readily visualized on routine MRCP.

On further analysis, indirect clues on cross-sectional imaging that may suggest AP include asymmetric or bulky pancreatic head tissue extending in a posterolateral or anterolateral direction around the descending duodenum, or pancreatic tissue located both anterior and posterior to the duodenum, producing a characteristic "crocodile jaw" configuration. Additional subtle features include mild duodenal indentation, angulation, or focal narrowing without significant proximal dilatation, particularly in patients presenting with symptoms of gastric outlet obstruction. Other suggestive findings may include a thin rim or incomplete cuff of pancreatic tissue partially encircling the second part of the duodenum, best appreciated on multiplanar reconstructions. An aberrant or accessory pancreatic duct may be seen coursing semicircumferentially around the duodenum before joining the main pancreatic duct; however, this duct may be nondilated and occasionally not visualized on standard MRCP.[18] In selected cases, additional preoperative imaging modalities such as high-resolution contrast-enhanced computed tomography or endoscopic ultrasonography may complement MRCP. Secretin-enhanced MRCP, where available, may be particularly useful as a noninvasive technique for detailed evaluation of pancreatic ductal anatomy.[18]

Hepaticoduodenostomy and RYHJ are the two most commonly employed techniques of biliary reconstruction in the surgical management of CDC. Hepaticoduodenostomy offers several advantages, including a single anastomosis, shorter operative time, reduced bowel handling, and preservation of physiological bile drainage into the duodenum, thereby allowing easier endoscopic access to the biliary tree if required in the future. It is technically simpler and may be associated with faster postoperative recovery. However, hepaticoduodenostomy has potential disadvantages, such as an increased risk of duodenogastric bile reflux, alkaline gastritis, and ascending cholangitis due to reflux of duodenal contents. In addition, prolonged exposure of the biliary epithelium to pancreatic enzymes has raised concerns regarding long-term malignant potential, although this remains controversial. On the other hand, RYHJ provides effective diversion of bile away from the duodenum, thereby minimizing biliary reflux and reducing the risk of ascending cholangitis. It has demonstrated durable long-term outcomes with low rates of anastomotic complications. Nevertheless, RYHJ is technically more demanding, requires multiple anastomoses, involves greater bowel manipulation, and is associated with longer operative time. Endoscopic access to the biliary anastomosis is more challenging, and complications such as Roux limb stasis, adhesive bowel obstruction, or anastomotic stricture may occur.[19]

In the present case, laparoscopic CDC excision with RYHJ was performed instead of the planned laparoscopic hepaticoduodenostomy due to the unexpected intraoperative finding of AP, which significantly influenced the choice of biliary reconstruction. The decision was based on the anticipated difficulty in achieving adequate duodenal mobilization for a tension-free hepaticoduodenostomy, the potential risk of future duodenal narrowing or functional obstruction even in the absence of preoperative symptoms, and the increased likelihood of biliary stasis, duodenobiliary reflux, and ascending cholangitis if hepaticoduodenostomy were performed in the presence of AP. Additionally, preserving the duodenum was considered important to facilitate a future duodeno-duodenostomy should AP-related obstructive symptoms develop later. From a surgical standpoint, all previous cases underwent open procedures, while our patient was successfully treated laparoscopically.

In conclusion, the coexistence of CDC and AP in children is exceedingly rare, with only 11 cases reported to date, including the present case. Unlike previously reported cases in which AP was symptomatic in the neonatal period, this case demonstrates that AP may remain clinically silent and be detected only intraoperatively. Awareness of this rare association is essential, as it may significantly influence surgical planning and long-term management.

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Data Sharing Statement: The data that support the findings of this study are available from the corresponding author upon reasonable request.

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