

## Bile Duct Perforation in Children: Is it Truly Spontaneous?

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### Abstract

**Introduction:** Bile duct perforation (BDP) with resultant biliary ascites in children is a rare clinical condition. The aetiopathogenesis is still an enigma, with increasing evidence suggesting anomalous union of pancreaticobiliary ductal (AUPBD) system as the prime causative factor. **Clinical Picture:** We report 2 cases of spontaneous perforation of the bile duct confirmed on histopathological examination as choledochal cyst, in a 6-month-old female child and a 4-year-old boy who presented with subtle clinical symptoms. **Treatment and Outcome:** Both patients were successfully managed by excision of the gall bladder and common bile duct and Roux-en-Y hepaticojejunostomy. This procedure was performed following initial cholecystostomy drainage in the second case. **Conclusions:** From the available literature and experience with our patients, BDP is not merely spontaneous but may be related to AUPBD and choledochal cyst.

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**Key words:** Aetiology, Biliary ascites, Choledochal cyst, Treatment

### Introduction

Bile duct perforation (BDP) and biliary ascites in children is a rare clinical entity. The aetiopathogenesis is still elusive. Growing evidence suggests that BDP and choledochal cyst (CC) are interrelated entities with a common pathogenetic factor, the anomalous union of the pancreaticobiliary ductal system (AUPBD).<sup>1-4</sup> Virtually all fully investigated cases of BDP in oriental patients have demonstrated AUPBD, which is also an aetiopathogenetic factor for genesis of choledochal cyst.<sup>1-4</sup> The differentiation between BDP and rupture of the choledochal cyst is debatable and this has bearing on the final management plan. Simple drainage with or without suture closure, T-tube or cholecystostomy has been proposed as the treatment of choice. Definitive surgery in the form of excision of the CC, cholecystectomy and Roux-en-Y hepaticoportoenterostomy seems to be the ideal option. We present 2 cases of bile duct perforation in children and discuss the dilemmas in the aetiopathogenesis and management in the light of available literature.

### Case Report

#### Case 1

A 6-month-old girl with no previous medical problems

was admitted to the paediatric ward with complaints of poor feeding and lethargy of 2 days' duration and 1 episode of non-bilious vomiting. Over the next 2 days, the child developed fever and seemed to draw up her legs when abdominal examination was performed. The child could tolerate oral feeds but had clay-coloured stools.

The preliminary investigations were unremarkable except for raised total leukocyte count ( $23 \times 10^9/L$ ). The septic screen was also unremarkable. The blood investigations showed total bilirubin  $55 \mu\text{mol/L}$  ( $\leq 14$ ), direct bilirubin  $31 \mu\text{mol/L}$  (0-7), total protein  $47 \text{ g/L}$  (53-78), albumin  $28 \text{ g/L}$  (38-53), alkaline phosphatase  $110 \text{ U/L}$  (157-363 U/L), alanine transaminase  $8 \text{ U/L}$  (9-48 U/L), aspartate transaminase  $29 \text{ U/L}$  (16-54 U/L) and gamma glutamyl transferase  $269 \text{ U/L}$  (5-26 U/L) (Figures in brackets indicate normal range). Serial abdominal radiographs showed stagnant bowel loops. Ultrasonography revealed fairly large amount of ascites with low level internal echoes, septations and loculations on the right side of the abdomen, likely inflammatory in nature. The gall bladder contained sludge and had a thickened wall (26 mm). The intra-hepatic biliary radicles were normal and the common bile duct was obscured by bowel gas. A few bowel loops in the right lower quadrant were thickened and oedematous. The

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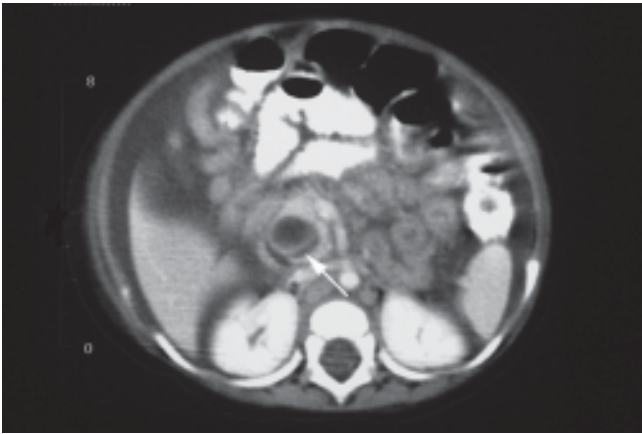


Fig. 1. CT scan of the abdomen showing dilated CBD (arrow) at the pancreatic head with periductal fluid collection.

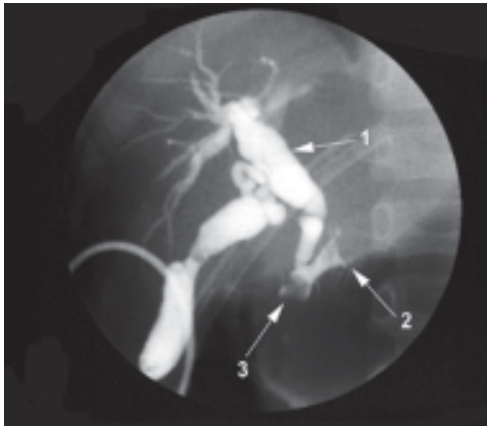


Fig. 3. Cholangiogram showing dilated CBD suggestive of a fusiform choledochal cyst (1) with reflux into the pancreatic duct (2) and a long bilio-pancreatic channel (3).

findings were suggestive of intraabdominal sepsis although the primary source could not be ascertained. Computed tomography (CT) scan of the abdomen confirmed the same findings and also revealed a dilated common bile duct (CBD) with its largest diameter (1 cm) at the level of the pancreatic head (Fig. 1).

The child was subjected to diagnostic laparoscopy with provisional diagnosis of infective peritonitis. The findings revealed bile in the peritoneum, which was found later to be sterile with total bilirubin of 383 mmol/L. Conversion to laparotomy was done and extensive bile staining of abdominal viscerae was noted with a perforation on the posterior wall of the CBD, which appeared to be mildly dilated. The liver was unremarkable and the gall bladder was distended. As the condition of the baby was stable, excision of the common bile duct and gall bladder with Roux-en-Y hepaticojejunostomy was done. The diagnosis of perforated Type 1 CC with bile duct measuring 7 mm in diameter was confirmed on histopathology. The post-operative recovery was uneventful and the child is well at 2 years after surgery.

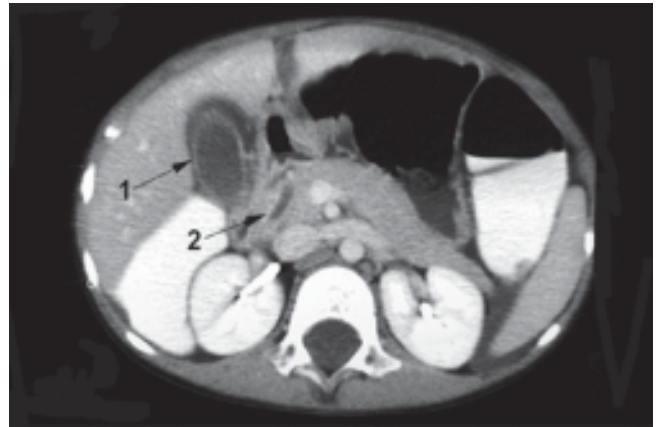


Fig. 2. CT scan of the abdomen showing thickened gall bladder with pericholecystic collection (1) and dilated CBD (2).

### Case 2

A 4-year-old male child with no previous medical history, presented with complaints of abdominal pain and non-bilious vomiting of a day's duration. The right upper quadrant of the abdomen was tender on palpation. The haemogram showed leukocytosis ( $19.6 \times 10^9/L$ ) and neutrophilia (89%). The liver function test revealed transaminitis and elevated gamma glutamyl transferase (total bilirubin  $17 \mu\text{mol/L}$  ( $\leq 14$ ) direct bilirubin  $5 \mu\text{mol/L}$  (0-7), alanine transaminase 227 U/L (8-29), aspartate transaminase 132 U/L (16-54), gamma glutamyl transferase 75 U/L (9-41) alkaline phosphatase 256 U/L (110-345) and amylase 123 U/L (39-123).

Ultrasonography of the abdomen revealed a grossly distended gall bladder with pericholecystic fluid collection and a dilated CBD extending to the intrapancreatic portion. Intravenous cefazolin was administered with a provisional diagnosis of acute acalculus cholecystitis. CT scan of the abdomen was done as the symptoms worsened. This revealed significant ascites as well as dilated hepatic and CBD (8 mm) and thickening of the gall bladder wall with pericholecystic fluid collection (Fig. 2).

Laparoscopy was performed with a provisional diagnosis of biliary ascites due to spontaneous BDP or rupture of the CC. At surgery, significant biliary ascites was noted, but definite site of perforation could not be seen. The antrum of the stomach was adherent to the undersurface of the liver. Cholecystostomy was performed and drains were placed in the subhepatic space and pelvis after peritoneal lavage.

Cholangiography performed on the eighth postoperative day through the cholecystostomy tube revealed a fusiform Type 1 CC with a long common bilio-pancreatic channel (Fig. 3). The patient was discharged and readmitted 3 weeks postoperatively for the excision of the choledochal cyst. At laparotomy, a Type 1 fusiform CC was noticed with a long common bilio-pancreatic channel. The CC was

excised and Roux-en-Y hepaticojejunostomy was performed. The postoperative recovery was uneventful and the child is well 1 year after the surgery.

## Discussion

Biliary ascites in children due to BDP is a rare clinical condition with fewer than 100 cases reported in the literature worldwide.<sup>5-7</sup> The peak age of occurrence is around 6 months with age ranging from 25 weeks gestation to 7 years.<sup>8</sup> The aetiopathogenesis is yet to be elucidated as various theories have been proposed such as congenital weakness of the bile duct, pancreaticobiliary anomalies including AUPBD, pancreatitis, distal obstruction of the bile duct due to inspissated bile, stones or stenoses, presence of a diverticulum or abnormal gland of the bile duct wall, viral infection of the bile duct, tuberculosis, necrotising enterocolitis and birth trauma.<sup>2,6,9</sup> Obstruction of the bile duct by protein plugs, inspissated bile or stone is considered as a consequence of BDP, slow bile transit and biliary stasis rather than the cause of perforation.<sup>8</sup> Filling defects in the common channel seen on cholangiograms may be misleading, as stenoses, calculi, protein plugs and biliary sludge have resolved spontaneously with adequate drainage.<sup>6,7</sup>

Although BDP and CC are considered as different entities there is growing evidence that both may be interrelated problems with a common pathogenesis. AUPBD has been demonstrated in fully investigated cases of BDP<sup>1-4,6</sup> but many former reports have not taken into consideration the AUPBD.<sup>1,2</sup> The difficulty in ascribing the cause of perforation secondary to CC is due to the first time presentation, small size of the cyst and collapse of the cyst wall following perforation. This was true with our first case. Yamaguchi et al<sup>9</sup> reported 26 cases of perforation (1.8%) in 1433 patients with CC. The incidence of perforation was 2.1% as reported by the American Academy of Pediatrics in a review of 188 patients with CC.<sup>10</sup>

BDP is frequently diagnosed only at laparotomy as the condition is rare and the clinical presentation is usually insidious with subtle symptoms.<sup>7,8,11</sup> The symptomatology includes mild jaundice, progressive abdominal distension, ascites, vomiting and acholic stools. Acute onset of symptoms in the form of abdominal distension, fever, vomiting and signs of fulminant bile peritonitis is exceptional.<sup>7,8</sup>

The investigations usually reveal cholestatic jaundice with near normal liver function tests, which differentiate BDP from biliary atresia and neonatal hepatitis.<sup>7</sup> Ultrasonography shows ascites, localised fluid collection around the porta and non-dilated bile duct.<sup>8</sup> CT scan of the abdomen reveals the same findings as ultrasound and it is difficult to diagnose the perforation preoperatively. On

retrospective review of the CT scan in our first case, the CBD had a crinkled appearance and there appeared to be a defect in its posteromedial aspect. The crinkled appearance was postulated to be due to the release of tension when the cyst ruptured. Abdominal paracentesis can be diagnostic with ascitic bilirubin levels ranging from 7.7 mg% to 420 mg%, but carries the risk of infecting the sterile bile.<sup>8</sup> The presence of bile in the peritoneal cavity associated with obstructive jaundice in the absence of liver derangement is considered pathognomonic for BDP.<sup>12</sup> Hepatobiliary scintigraphy with Tc 99m imidodiacetic acid derivatives may demonstrate the leak of radionuclide tracer and aid in the definitive diagnosis of BDP.<sup>13</sup> Magnetic resonance cholangiopancreatography (MRCP) provides a non-invasive method of evaluating the biliary tree using heavily T2-weighted sequences.<sup>14</sup> The signals from either static or slowly moving liquids are augmented, and liquid-filled compartments like bile ducts or pancreatic ducts and the contrast between ducts and base are made evident without using a paramagnetic contrast media. Owing to its excellent anatomical and contrast resolutions, MRCP can delineate BDP. MRCP also evaluates the pancreaticobiliary junction anomalies such as AUPBD, which would aid in discerning the aetiopathology and management plans for BDP. Recently, the effectiveness of dynamic MRCP with secretin stimulation has been reported in the evaluation of pancreaticobiliary maljunction anomalies.<sup>15</sup> However, subtle clinical signs, cost and availability limit the use of MRCP in evaluation of children presenting with features of acute abdomen. Endoscopic retrograde cholangiopancreatography aids in the delineation of BDP and AUPBD, but the invasiveness and risk of complications such as pancreatitis, cholangitis, sepsis, adverse reactions to contrast media and bile duct or duodenal injury limits its utilisation in evaluation of suspected BDP.<sup>16</sup> Laparoscopy is useful in ascertaining the diagnosis and also to drain the perforation.<sup>8</sup> Due to worsening abdominal distension, our patients were subjected to diagnostic laparoscopy with a provisional diagnosis of appendicular perforation or primary peritonitis in the first case and BDP in the second case.

The perforation is usually seen as a punched-out hole in the anterior aspect of BD at the junction with the cystic duct as it has been suggested that this may be prone to mural malformation during embryogenesis.<sup>6</sup> On the contrary, there are reports stating that the usual location of perforation is on the posterior wall of the CC as was seen with our patient. Operative cholangiogram confirms the diagnosis, sites the perforation, evaluates the status of the distal BD including AUPBD and provides a road map for surgery.<sup>7,8</sup> It was not done in our first case as the perforation was evident but retrospectively, we believe that it would aid as described.

Management options vary from simple drainage to definitive correction in the form of excision and bilioenteric drainage.<sup>7,8</sup> Operative modalities suggested include simple drainage with or without suture closure,<sup>5,10</sup> T tube drainage and/or cholecystostomy.<sup>8</sup> Other options like cholecystectomy, hepaticocholedochostomy, hepaticoduodenostomy, hepaticojejunostomy, choledochocholedochostomy, choledochojejunostomy or resection of biliary pseudocyst are considered to be unnecessary and carry a high morbidity and mortality risk.<sup>8</sup> Simple drainage with or without suture closure entails less morbidity and may cure the condition or stabilise the patient for a second look definitive surgery. Presence of T tube/cholecystostomy facilitates biliary drainage and postoperative assessment of the BD anatomy.<sup>7</sup>

AUPBD has been claimed to be responsible for the development of CC and BDP in infancy and may be the cause of cancer of BD in later life.<sup>8</sup> The risk of malignant degeneration of CC is well documented in the literature. This risk appears to be age related with less than 1% risk in patients less than 10 years of age and about a 15% risk in patients over 20 years of age. Since there is potential risk of subsequent malignant degeneration, we believe that these patients with BDP should undergo excision of the BD/CC and bilioenteric drainage. There are few case reports of successful treatment of the condition by definitive surgery, as most believe that extensive procedures bear high risk of morbidity and mortality.<sup>11</sup> We were prompted towards cholecystostomy drainage with peritoneal lavage for the second case, based on the reports in the literature.<sup>5,7,8,10</sup> The postoperative cholangiogram delineated the choledochal cyst with a long common bilio-pancreatic channel.<sup>17</sup>

Although around 100 cases of BDP have been described and the incident is spontaneous, the etiopathogenesis is still an enigma. In the light of available literature evidence and experience with our patients, there are reasons to deem that BDP is not merely spontaneous and may be related to AUPBD and choledochal cyst.<sup>1-4,6</sup> Hence, we believe that if the condition of the baby were stable, definitive surgery as described would be an ideal option, reserving simple drainage procedure with or without T tube/cholecystostomy for patients with poor general condition.<sup>3,6,11</sup> Proper meta analysis of all the cases reported with long-term follow-up may provide an answer.

## REFERENCES

- Ohkawa H, Takahashi H, Maie M. A malformation of the pancreatico-biliary system as a cause of perforation of the biliary tract in childhood. *J Pediatr Surg* 1977;12:541-6.
- Yano H, Matsumoto H. Choledochal cyst following operation for idiopathic perforation of the biliary tract in childhood. *Jpn J Surg* 1983;13:441-5.
- Hasegawa T, Udatsu Y, Kamiyama M, Kimura T, Sasaki T, Okada A, et al. Does pancreatico-biliary maljunction play a role in spontaneous perforation of the bile duct in children? *Pediatr Surg Int* 2000;16:550-3.
- Ng WT, Cheung CH, Chan S. Is spontaneous perforation of the bile duct in children due solely to pancreatico-biliary maljunction? *Pediatr Surg Int* 2002;18:565-6.
- Kasat LS, Borwankar SS, Jain M, Naregal A. Spontaneous perforation of the extrahepatic bile duct in an infant. *Pediatr Surg Int* 2001;17:463-4.
- Ando H, Ito T, Watanabe Y, Seo T, Kaneko K, Nagaya M. Spontaneous perforation of choledochal cyst. *J Am Coll Surg* 1995;181:125-8.
- Spigland N, Greco R, Rosenfeld D. Spontaneous biliary perforation: does external drainage constitute adequate therapy? *J Pediatr Surg* 1996;31:782-4.
- Banani SA, Bahador A, Nezakatgoo N. Idiopathic perforation of the extrahepatic bile duct in infancy: pathogenesis, diagnosis, and management. *J Pediatr Surg* 1993;28:950-2.
- Yamaguchi M. Congenital choledochal cyst. Analysis of 1,433 patients in the Japanese literature. *Am J Surg* 1980;140:653-7.
- Kim SH. Choledochal cyst: survey by the surgical section of the American Academy of Pediatrics. *J Pediatr Surg* 1981;16:402-7.
- Moss RL, Musemeche CA. Successful management of ruptured choledochal cyst by primary cyst excision and biliary reconstruction. *J Pediatr Surg* 1997;32:1490-1.
- Ford WD, Sen S, Morris L, LeQuesne G. Spontaneous perforation of the common bile-duct in the neonate: imaging and treatment. *Aust Paediatr J* 1988;24:306-8.
- Fawcett HD, Hayden CK, Swischuk LE, Lobe TE. Spontaneous extrahepatic biliary duct perforation in infancy. *Can Assoc Radiol J* 1986;37:206-7.
- Sugiyama M, Baba M, Atomi Y, Hanaoka H, Mizutani Y, Hachiya J. Diagnosis of anomalous pancreaticobiliary junction: value of magnetic resonance cholangiopancreatography. *Surgery* 1998;123:391-7.
- Fukukura Y, Fujiyoshi F, Sasaki M, Nakajo M. Pancreatic duct: morphologic evaluation with MR cholangiopancreatography after secretin stimulation. *Radiology* 2002;222:674-80.
- Farrell RJ, Noonan N, Mahmud N, Morrin MM, Kelleher D, Keeling PW. Potential impact of magnetic resonance cholangiopancreatography on endoscopic retrograde cholangiopancreatography workload and complication rate in patients referred because of abdominal pain. *Endoscopy* 2001;33:668-75.
- Waghlikar GD, Chetri K, Yachha SK, Sikora SS. Spontaneous perforation – a rare complication of choledochal cyst. *Indian J Gastroenterol* 2004;23:111-2.