

Usefulness of Thin-Slice Contrast-Enhanced Computed Tomography in Detecting Perforation Site in Congenital Biliary Dilatation: A Case Report

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The site of perforation is difficult to identify preoperatively in many cases with spontaneous perforation of congenital biliary dilatation (CBD). We report a case of spontaneous perforation of CBD in which the perforation site was identified preoperatively using thin-slice contrast-enhanced computed tomography (CT). The patient was a girl aged 1 year and 4 months. She was admitted to our hospital because of vomiting and diarrhea that had continued for 3 days prior to admission. Abdominal contrast CT on admission showed dilated common bile duct, thickening of the gall bladder wall, and marked ascites. In addition, an area of low density with a diameter of 1 cm was detected near the neck of the gallbladder. We evaluated the area via thin-slice contrast-enhanced CT and detected a defect in the wall of the bile duct. Cholangiography revealed abnormal confluence of the pancreaticobiliary duct and a protein plug in the common duct. A diagnosis of CBD with perforation of the bile duct was made, and surgery was performed. The intraoperative findings matched that seen on the enhanced CT. There are some reports of pseudocysts and fluid retention around the perforation site; however, no reports are found in which the perforation site was confirmed by preoperative CT. If localized fluid retention is observed in cases with biliary perforation, confirmation with thin-slice contrast-enhanced CT might be useful for identifying the perforation site. (J Nippon Med Sch 2022; 89: 623–628)

Key words: congenital biliary dilatation, fluid accumulation, perforation site, spontaneous perforation, thin-slice contrast-enhanced computed tomography

Introduction

Spontaneous bile duct perforation in cases with congenital biliary dilatation (CBD) is a relatively rare condition that occurs in 1.8%-18% of all cases of CBD¹. Many cases involve girls aged ≤ 4 years, and many patients experience non-specific symptoms such as fever and abdominal distension². If treatment is delayed, it might cause peritonitis. Therefore, surgery must be performed without delay in patients with CBD in whom there is a suspicion of spontaneous bile duct perforation. Since the symptoms of bile duct perforation in young children are non-specific in many cases, imaging studies such as abdominal

sonography, contrast-enhanced computed tomography (CT), magnetic resonance cholangiopancreatography (MRCP), and hepatobiliary scintigraphy are required for the diagnosis³. Only in rare cases, the location of the perforation identified on a preoperative imaging study by MRCP³, however, no reports are found in which the perforation site was confirmed by preoperative CT.; hence, the site of the perforation is usually identified intraoperatively. We report here a case of bile duct perforation in a patient with CBD in which the perforation site was identified preoperatively using thin-slice contrast-enhanced CT. We obtained written consent from participants re-

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Fig. 1 Abdominal X-ray showed displacement of intestines medially, indicating ascites.

garding the publication of this report.

Case Presentation

The patient was a girl aged 1 year and 4 months. She presented with a chief complaint of vomiting and diarrhea, starting 3 and 2 days before hospital admission, respectively. On the day prior to hospital admission, the vomiting increased in frequency. The patient was examined by a primary physician and admitted to the hospital with a diagnosis of acute gastroenteritis. The primary physician noted negligible improvement even after hospital admission and that the patient showed increasing somnolence. The patient was then transferred to our hospital for further examinations and treatments. Her medical history and family history was unremarkable.

Her vital signs at the time of our hospital admission were as follows: body temperature 37.0°C, respiratory rate 30/min, SpO₂ 98% (indoor air), heart rate 153 bpm, and blood pressure 111/67 mmHg. Clear respiratory sounds with no heart murmur were heard. Soft but distended abdomen and diminished abdominal sounds indicated decreased bowel motility. No obvious masses and no hepatosplenomegaly were found by palpation. There was no rash and no yellowing of the skin.

Findings of blood examinations at admission were as follows: white blood cell count, 12,100 / μ L; C-reactive protein (CRP) level, 0.39 mg/dL; total bilirubin level, 1.01 mg/dL; aspartate aminotransferase level, 76 IU/L;

alanine aminotransferase level, 53 IU/L; lactate dehydrogenase level, 291 IU/L; amylase level, 273 IU/L; urea nitrogen level, 7.1 mg/dL; creatinine level, 0.23 mg/dL; sodium level, 145 mmol/L; potassium level, 4.4 mmol/L; chloride level, 110 mmol/L; prothrombin time-international normalized ratio, 1.10; activated partial thromboplastin time, 22.5 seconds; fibrinogen level, 90 mg/dL; fibrin degradation product level, 10.3 mg/mL.

Abdominal X-ray at admission found marked mild bowel dilatation and displacement of intestines medially, indicating ascites (**Fig. 1**). Abdominal sonography at admission observed marked ascites and thickening of the gallbladder wall. Because it was difficult to visualize the bile duct and pancreas due to the effects of gas in the intestinal tract by abdominal sonography, contrast-enhanced CT was performed without sedation. The contrast-enhanced CT protocol was as follows: scans using a 320-detector row CT machine (Aquilion one, CANON MEDICAL SYSTEMS CORPORATION, Tochigi, Japan); tube voltage, 100 kV; tube current, 150 mAs; rotation time, 0.35 s; slice thickness, 1.0 mm (volume computed tomography dose index 22.20 mGy, dose length product 675.60 mGy \cdot cm). The enhanced CT scans was started at 60 s after injection manually.

Findings of contrast-enhanced CT at admission were as follows: marked ascites, dilatation of the common bile duct, and thickening of the gallbladder wall (**Fig. 2A, B and C**). A region of low density was found, indicating accumulation of bile; this region measured approximately 1 cm in the vicinity of the neck of the gallbladder (**Fig. 2D**). The bile duct was confirmed to be adjacent to this region of low density on thin-slice contrast-enhanced CT. We also identified loss of the bile duct wall measuring approximately 2 \times 3 mm in the vicinity of the junction of the common hepatic duct, the cystic duct, and the common bile duct (**Fig. 2**). The anomalous pancreaticobiliary junction was not able to be identified.

Post-admission Course

Analysis of the contrast-enhanced CT performed at admission led to the bile duct perforation associated with CBD; hence, surgery was performed on 3 days of the hospital admission. The ascitic fluid resembled bile. Findings of the fluid showed elevated levels of total bilirubin (18.19 mg/dL), direct bilirubin (12.23 mg/dL), and amylase (4,103 IU/L). Intraoperative cholangiography revealed dilation of the common bile duct and leakage of contrast medium in the vicinity of the junction of the common hepatic duct, the cystic duct, and the common bile duct. Areas of non-visualization of contrast were

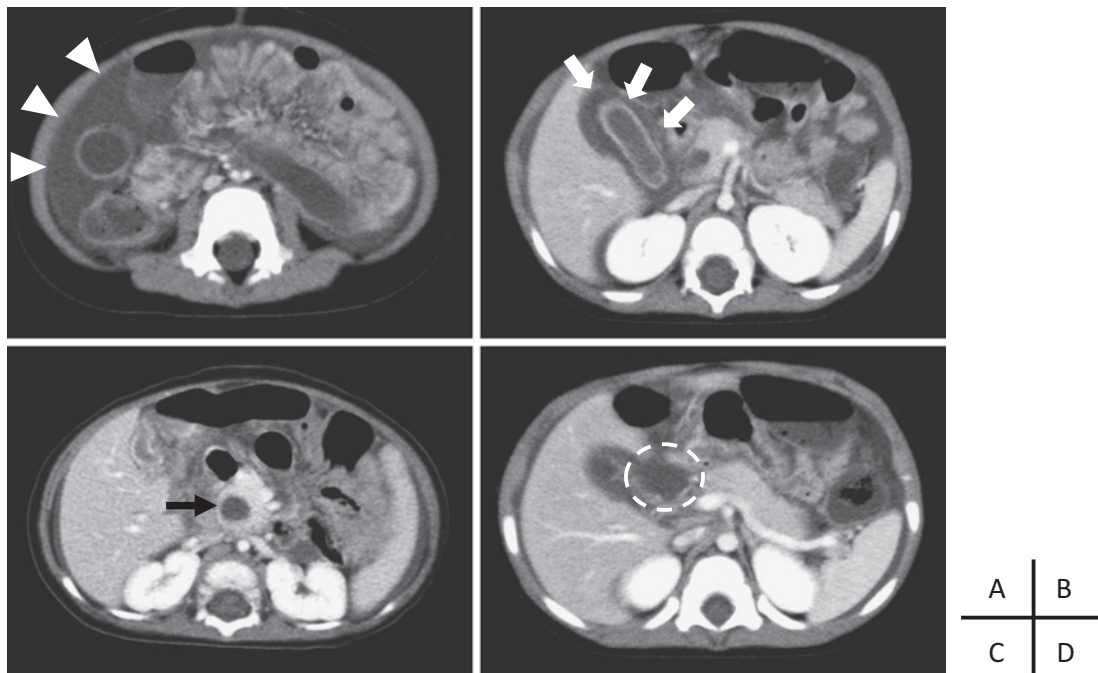


Fig. 2 Abdominal contrast-enhanced computed tomography (volume computed tomography dose index 22.20 mGy, dose length product 675.60 mGy · cm, slice thickness 3.00 mm)
 Ascites (A, arrow heads), thickening of the gallbladder wall (B, white arrows), and common bile duct dilatation (C, black arrow) are seen. A region of low density indicating exclusion of the gallbladder wall in the vicinity of the junction of the common hepatic duct, the cystic duct, and the common bile duct are observed (D, dotted circle).

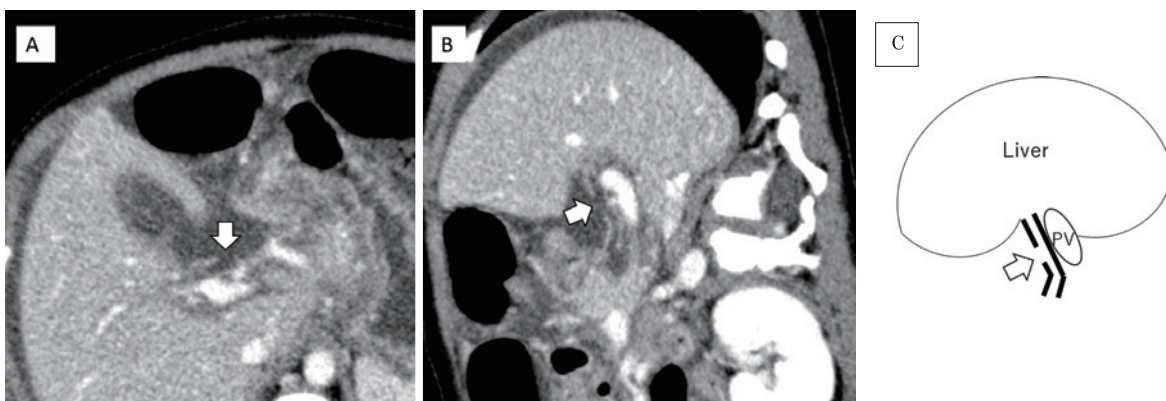


Fig. 3 Thin-slice contrast-enhanced computed tomography (slice thickness of 1.00 mm)
 A) Horizontal slice, B) oblique sagittal slice parallel to the common bile duct, loss of the bile duct wall measuring approximately 2×3 mm in the vicinity of the junction of the common hepatic duct, which suggests bile duct perforation (white arrow). C) Illustration of B), PV: portal vein.

found, indicating an abnormality at the junction of the pancreatic duct and the common bile duct and protein thrombus. The patient was diagnosed with bile duct perforation associated with CBD (Fig. 3). Intraoperative findings indicated bile leakage in the vicinity of the junction of the common hepatic duct, the cystic duct, and the common bile duct and bile accumulation localized between this region and the nearby serosa (Fig. 4). There were strong adhesions in the region surrounding the area

of localized bile accumulation. Although we were unable to identify the site of perforation during surgery, the site of bile accumulation and the site of the leakage of cholangiography contrast medium were consistent with the findings on contrast-enhanced CT at admission (Fig. 5).

Surgery included trans-gallbladder and common bile duct drainage, and peritoneal lavage. On 81 days of hospital stay, secondary extrahepatic bile duct resection and hepaticojejunostomy were performed. The patient was

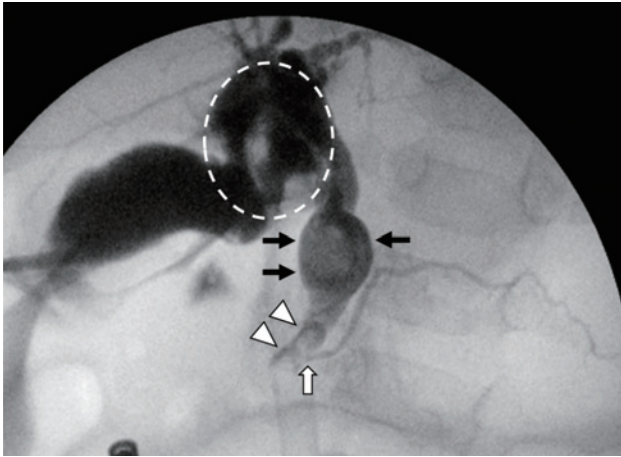


Fig. 4 Intraoperative cholangiography

Contrast medium leakage (dotted circle) is observed in the vicinity of the junction of the common hepatic duct, the cystic duct, and the common bile duct. Bile duct dilatation (black arrows), filling defects suggestive of protein thrombus (arrow heads), and abnormal pancreatic duct confluence (white arrow) are observed. Based on these findings, the diagnosis was bile duct perforation in association with congenital biliary dilatation.

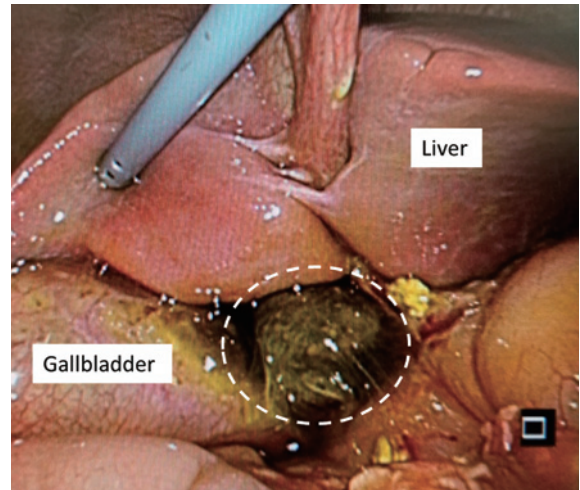


Fig. 5 Intraoperative findings

Accumulation of bile is observed in the vicinity of the junction of the common hepatic duct, the cystic duct, and the common bile duct, which was consistent with the findings on contrast-enhanced computed tomography.

discharged on 100 days of hospital stay.

Discussion

In this case, findings from physical examination led to suspicion of acute gastroenteritis; however, a contrast-enhanced CT led to the diagnosis of bile duct perforation associated with CBD. Detailed analyses using thin-slice contrast-enhanced CT allowed preoperative identification of the perforation site.

Spontaneous perforation has been considered to be a rare complication of CBD¹. Although the specific cause of spontaneous perforation of CBD remains obscure, some authors believed that irritation from refluxed pancreatic juice coupled with immature CBD wall was responsible for the perforation⁴. The immaturity of the ductal wall in children younger than 4 years could be an important contributory factor of perforation². Therefore, a majority of the perforation cases were distributed in children younger than 4 years⁵. In this case as well, she was 1 year and 4 months old. The site of spontaneous perforation of CBD has been reported almost always at the union of the common bile duct and the cystic ducts, and is believed to occur due to congenital bile duct weakness, possibly due to a mural malformation during early embryogenesis⁴. Also in this case, the site of perforation was the vicinity of the junction of the common hepatic duct, the cystic duct, and the common bile duct.

The symptoms of bile duct perforation associated with CBD are often non-specific and include abdominal distension, fever, and abdominal pain. Thus, not only many cases are misdiagnosed, but also the site of perforation can only be identified during surgery. Chen et al. reported that bile duct perforation is diagnosed preoperatively in approximately 13% of the cases, and the common misdiagnoses are intestinal obstruction (35%) and appendicitis (26%); blood γ -glutamyl transpeptidase and amylase levels are elevated in 60% and 4% of the cases, respectively; however, leukocytosis and elevated CRP level are not observed⁶. In the present case, the physical examination revealed only non-specific findings such as abdominal distension and fever, and the blood examination results did not show leukocytosis or elevated CRP level, while amylase was only slightly elevated. It was difficult to diagnose bile duct perforation based on the physical examination and results of the blood examinations. Hence, it is extremely important to conduct imaging studies to diagnose bile duct perforation associated with CBD.

The imaging studies used for preoperative diagnosis include ultrasound, contrast-enhanced CT, MRCP, and hepatobiliary scintigraphy. It has been generally accepted that abdominal ultrasonography is the most useful diagnostic tool for detection of CBD and is the first modality for the perforation with CBD. In their analysis of the ultrasound findings, Chen et al. reported that among 23 subjects, ascites was found in all subjects (100%), thicken-

ing of the gallbladder wall was found in 19 subjects (83%), and biliary dilatation was found in 16 subjects (70%)⁶. Hence, when such findings are observed on imaging, bile duct perforation in association with CBD must be suspected. Lee et al. reported that in some cases, it was difficult to visualize the bile duct on abdominal sonography due to the effects of gas in the intestinal tract³. Tani et al. reported that in an emergency setting, CT, in addition to US, is a useful diagnostic tool, especially for abnormalities in the retroperitoneal space. In this case, it was difficult to visualize the bile duct on abdominal sonography, therefore in emergency setting, CT was performed. MRCP is also a useful of noninvasively depict an abnormal connection between the pancreatic duct and the common bile duct. MRCP allows detailed anatomical abnormalities to be identified from sites, such as the junction of the pancreatic duct. This also allows local accumulation of fluid in the vicinity of the perforation site to be confirmed, which is useful in suspecting the perforation site. Hepatobiliary scintigraphy also allows the site of biliary perforation to be suspected as it reveals leakage of bile into the peritoneum. Fluid accumulation was detected in the region surrounding the site of perforation in the present case, and this clue allowed us to identify the perforation site. However, it is difficult to perform a detailed evaluation of the bile duct wall on MRCP or hepatobiliary scintigraphy; hence, it is impossible to identify the site of perforation accurately using these modalities.

Contrast-enhanced CT allows visualization of the structure of the bile duct. Usually, the bile duct wall cannot be visualized using contrast-enhanced CT in many cases; however, since the enhancement of the bile duct wall is more pronounced in cases with pathological changes such as cholangitis, it can be seen on contrast-enhanced CT⁷. It is known that in cases of CBD, pancreatic enzymes backflow into the bile duct and irritate the bile duct mucosa⁸. Based on the postoperative pathological findings in cases of CBD, Kim et al. reported hypertrophy of the bile duct wall⁹. The pathological findings in the present case also showed fibrous hypertrophy of the wall of the common bile duct and gallbladder, as well as neutrophil infiltration (data not shown). Thus, since enhancement of the bile duct walls was seen on contrast-enhanced CT in this case of CBD, similar to that seen in cholangitis, this modality can be used to evaluate the bile duct walls.

The bile duct perforation might be extremely small in some cases of CBD. Chen et al. reported that the mean \pm

SD diameter of perforations is $4.6 \text{ mm} \pm 1.4 \text{ mm}$ ⁶. Thus, it might be impossible to identify the perforation site on contrast-enhanced CT slices between 3 and 5 mm in thickness. In the present case, the size of the perforation was approximately $2 \times 3 \text{ mm}$, and identification using contrast-enhanced CT slices of 3 mm thickness was difficult. However, on contrast-enhanced CT slices of 1 mm thickness, the perforation site was clearly visible. Hence, we believe that it is important to use thin-slice contrast-enhanced CT imaging to evaluate bile duct perforations.

Conclusion

Presence of ascites, thickening of the gallbladder wall, and biliary dilatation are useful in diagnosing the perforation of CBD. Fluid accumulation localized to the area surrounding the perforation site might also be seen at times. Detailed observations of the bile duct wall in the vicinity of the region of fluid accumulation on thin-slice contrast-enhanced CT might help in identification of the perforation site.

Conflict of Interest: The authors declare that they have no conflicts of interest. This case study received no external funding.

References

1. Chiang L, Chui CH, Low Y, Jacobsen AS. Perforation: a rare complication of choledochal cysts in children. *Pediatr Surg Int*. 2011 Aug;27(8):823-7.
2. Tani C, Nosaka S, Masaki H, Kuroda T, Honna T. Spontaneous perforation of choledochal cyst: a case with unusual distribution of fluid in the retroperitoneal space. *Pediatr Radiol*. 2009 Jun;39(6):629-31.
3. Lee MJ, Kim MJ, Yoon CS. MR cholangiopancreatography findings in children with spontaneous bile duct perforation. *Pediatr Radiol*. 2010 May;40(5):687-92.
4. Ando K, Miyano T, Kohno S, Takamizawa S, Lane G. Spontaneous perforation of choledochal cyst: a study of 13 cases. *Eur J Pediatr Surg*. 1998 Feb;8(1):23-5.
5. Fukuzawa H, Urushihara N, Miyakoshi C, et al. Clinical features and risk factors of bile duct perforation associated with pediatric congenital biliary dilatation. *Pediatr Surg Int*. 2018 Oct;34(10):1079-86.
6. Chen J, Tang Y, Wang Z, Wang Q, Wang D. Clinical value of ultrasound in diagnosing pediatric choledochal cyst perforation. *AJR Am J Roentgenol*. 2015 Mar;204(3):630-5.
7. Schulte SJ, Baron RL, Teefey SA, et al. CT of the extrahepatic bile ducts: wall thickness and contrast enhancement in normal and abnormal ducts. *AJR Am J Roentgenol*. 1990 Jan;154(1):79-85.
8. Fumino S, Iwai N, Deguchi E, et al. Spontaneous rupture of choledochal cyst with pseudocyst formation-report on 2 cases and literature review. *J Pediatr Surg*. 2006 Jun;41(6):e19-21.
9. Kim JH, Choi TY, Han JH, et al. Risk factors of postoperative anastomotic stricture after excision of choledochal cysts with hepaticojejunostomy. *J Gastrointest Surg*. 2008

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