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ORIGINAL ARTICLE

MDCT of biliary cysts in children with biliary atresia: clinical associations and pathologic correlations

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PURPOSE
To evaluate the association of biliary cyst formation with cholangitis, portoenterostomy, biochemical abnormalities, using multidetector computed tomography (MDCT) and pathologic findings of end-stage liver disease.

MATERIALS AND METHODS
We retrospectively reviewed the 42 MCDT studies, clinical history and laboratory findings of 36 children with biliary atresia.

RESULTS
Biliary cysts were detected in 58% of the patients on MDCT images. The cysts were not associated with cholangitis, portoenterostomy surgery, or biochemical abnormality. Hepatic artery anomaly was also common in our series (25%) and more common in patients with biliary cysts which was statistically significant ($P < 0.05$). Eighteen livers were available for pathologic examination. The only statistically significant finding between the patients with and without biliary cysts were biliary epithelial damage and inflammatory reaction around the cysts which were common in the patients with biliary cysts ($P < 0.05$).

CONCLUSION
The damage to the bile duct epithelium and inflammatory reaction around the biliary epithelium support the theory of obstruction and bile leaks in the etiogenesis of biliary cysts. This is the first report of the association between hepatic artery variations and the biliary cysts; this may be important in pretransplant evaluation.

Key words: • *biliary system, abnormalities* • *diagnostic imaging* • *cysts*

Biliary atresia is a liver disease of newborns in which there is obliteration of intrahepatic and extrahepatic bile ducts leading to progressive liver injury (1). It is the third most common cause of neonatal cholestatic jaundice and is the most common indication for liver transplantation in children (2). The etiology is unknown, but perinatal viral infections, genetic factors, defects in immune response, autoimmune disorders, and defects in morphogenesis of biliary tree have been postulated. The disorder is commonly classified as classic (or perinatal) type and the embryonic (or fetal) type. In the perinatal type, bile ducts are patent at birth. A progressive inflammatory and sclerotic reaction results in the eventual obliteration of the biliary tree. The consequence is the development of obstructive jaundice, indicated by direct hyperbilirubinemia and acholic stools. The embryonic or fetal type often is associated with other structural anomalies such as the polysplenia sequence, preduodenal portal vein, and situs inversus. In this setting, the extrahepatic biliary tree may not have formed (3). The prognosis of patients with biliary atresia has improved since the introduction of Kasai and Suzuki's hepatic portoenterostomy in 1959 (4). Long-term follow-up has shown that intrahepatic cystic lesions develop after hepatic portoenterostomy in some patients. These cysts may be a source of recurrent infection and affect the morbidity and mortality of patients with biliary atresia (5–7). Although the clinical manifestations are clear, the cause of cyst formation in the biliary system is unclear. The aim of this study is to present multidetector computed tomography (MDCT) findings of biliary cysts and to evaluate the association of biliary cyst formation with cholangitis, portoenterostomy, biochemical abnormalities, and pathologic findings of end-stage liver disease.

Materials and methods

The study group consisted of children with known biliary atresia examined at our hospital from January 1996 to June 2008 for whom MDCT images were available for review. All patients except one were referred for MDCT as part of an evaluation protocol for possible liver transplantation to detect any vascular anomalies or complications of biliary disease in pretransplant period; 42 studies were available in 36 patients in the pretransplant period. The diagnosis of biliary atresia was confirmed by histologic examination in all patients. Abdominal MDCT examinations were performed with 4-row CT scanners (Volume Zoom and Somatom Plus 4; Siemens, Erlangen, Germany), and a 16-row CT scanner (Sensation 16, Siemens, Erlangen, Germany). Hepatic angiography was performed in all patients except one. The CT acquisition, which was designed to cover the entire craniocaudal extent of the liver and vascular structures, was performed during the precontrast phase, the arterial phase, the portal venous phase, and the late phase.

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Each patient was administered 3 mL/kg nonionic contrast material intravenously at a rate of 0.5 to 4 mL/s, depending on the age of the patient. Sedation was given to children younger than 6 years old and to patients unable to cooperate with the procedure. The CT parameters were 120 kV, 0.75 mm slice thickness, 12 mm feed/rotation, and 0.7 mm reconstruction thickness. We used 80 to 100 mA depending on the weight of children. MDCT images were evaluated for the presence, number, and location of biliary cysts. Findings that may be related to cirrhosis or portal hypertension were also assessed on CT images, including liver size (small or large), parenchymal heterogeneity, liver configuration (enlarged caudate lobe or left lobe), contour irregularity or lobularity, decrease in hepatic vein caliber, portal vein stenoses and cavernous transformation, splenomegaly, ascites, and portosystemic collateral vessels. We used age-dependent standards to determine the presence of hepatomegaly and splenomegaly (8).

We noted whether a portoenterostomy was performed. The patients were clinically diagnosed with cholangitis if they had fever associated with increased serum bilirubin and liver enzymes. We evaluated liver and biliary enzymes routinely obtained at our hospital in patients with biliary atresia. We included biochemical data if the data had been obtained within 1 month of CT images.

Pathologic examination of liver was performed in 27 cases. Only hepatectomy specimens in 18 patients who underwent liver transplantation were included in the study. Biliary cirrhosis was classified according to Scheuer's classification (9). Pathological specimens were examined for cholestases (ductular or hepatocanalicular), cystic ductus, foci of hepatocellular necrosis, cholangitis, bile plug-pseudorosette formation, bile duct damage, and inflammatory reaction and the loss of bile ducts.

We divided the patients into two groups on the basis of the presence or absence of biliary cysts on MDCT images. Differences in imaging, clinical, and biochemical data between two groups were tested for significance by using Mann Whitney U test for continuous variables or chi-square test for categorical variables. Probability values of less than 0.05 were considered significant.

Results

Clinical evaluation

The patients (19 females, 17 males) ranged from 2 days to 11 years of age (mean age, 14 months). Eighteen patients (50%) had liver transplantation for progressive liver injury; the other eighteen patients did not have liver transplantation because suitable donors could not be found. Table 1 shows the clinical and laboratory findings of patients with biliary atresia. A history of cholangitis was not significantly different between the two groups; instead it was common in patients without biliary cysts (19% versus 46.7%). Twenty-seven of 36 patients had Kasai operations previous to this evaluation. Although early portoenterostomy has been recommended for children with biliary atresia, 5 of 21 patients with biliary cysts and 4 of 15 patients without biliary cysts did not have surgery. Biochemical data were available for all patients. Except for one patient all patients had abnormal liver enzymes and serum bilirubin levels. Both groups had similar distributions of values for total and direct serum bilirubin, alanine aminotransferase, and aspartate aminotransferase levels.

The median follow-up time was 20 months in patients with liver transplantation and 11 months in patients without liver transplantation. Only one patient with liver transplantation died from hepatic failure. Of 18 patients without liver transplantation, 7 died from hepatic failure. Three of them also had intraabdominal hemorrhage, and one had pulmonary hemorrhage. In addition, one patient died from upper gastrointestinal hemorrhage. Four patients had no clinical follow-up.

Table 1. CT findings in children with biliary atresia with and without biliary cysts

	Patients			
CT findings	Total (n, %) n = 36	With cysts (n, %) n = 21	Without cysts (n, %) n = 15	χ^2, *P*
Small liver	1 (2.8)	1 (4.8)	0	$\chi^2 = 0.391$, $P > 0.05$
Large liver	15 (41.7)	11 (52.4)	4 (26.7)	$\chi^2 = 0.123$, $P > 0.05$
Enlargement of the left or caudate lobe	10 (27.8)	4 (19)	6 (40)	$\chi^2 = 0.166$, $P > 0.05$
Heterogenous parenchyma	11 (30.6)	9 (42.9)	2 (13.3)	$\chi^2 = 0.058$, $P < 0.05$
Contour irregularity or lobularity	18 (50)	12 (57.1)	6 (40)	$\chi^2 = 0.310$, $P > 0.05$
Splenomegaly	27 (75)	16 (76.2)	11 (73.3)	$\chi^2 = 0.845$, $P > 0.05$
Ascites	15 (41.7)	7 (33.3)	8 (53.3)	$\chi^2 = 0.230$, $P > 0.05$
Collaterals	17 (47.2)	12 (57.1)	5 (33.3)	$\chi^2 = 0.158$, $P > 0.05$
Decrease in hepatic vein caliber	3 (8.3)	1 (4.8)	2 (13.3)	$\chi^2 = 0.359$, $P > 0.05$
Portal vein stenosis and cavernous transformation	5 (13.9)	2 (9.5)	3 (20)	$\chi^2 = 0.370$, $P > 0.05$
Hepatic artery anomaly	9 (25)	9 (42.9)	0	$\chi^2 = 0{,}003$, $P < 0.05$

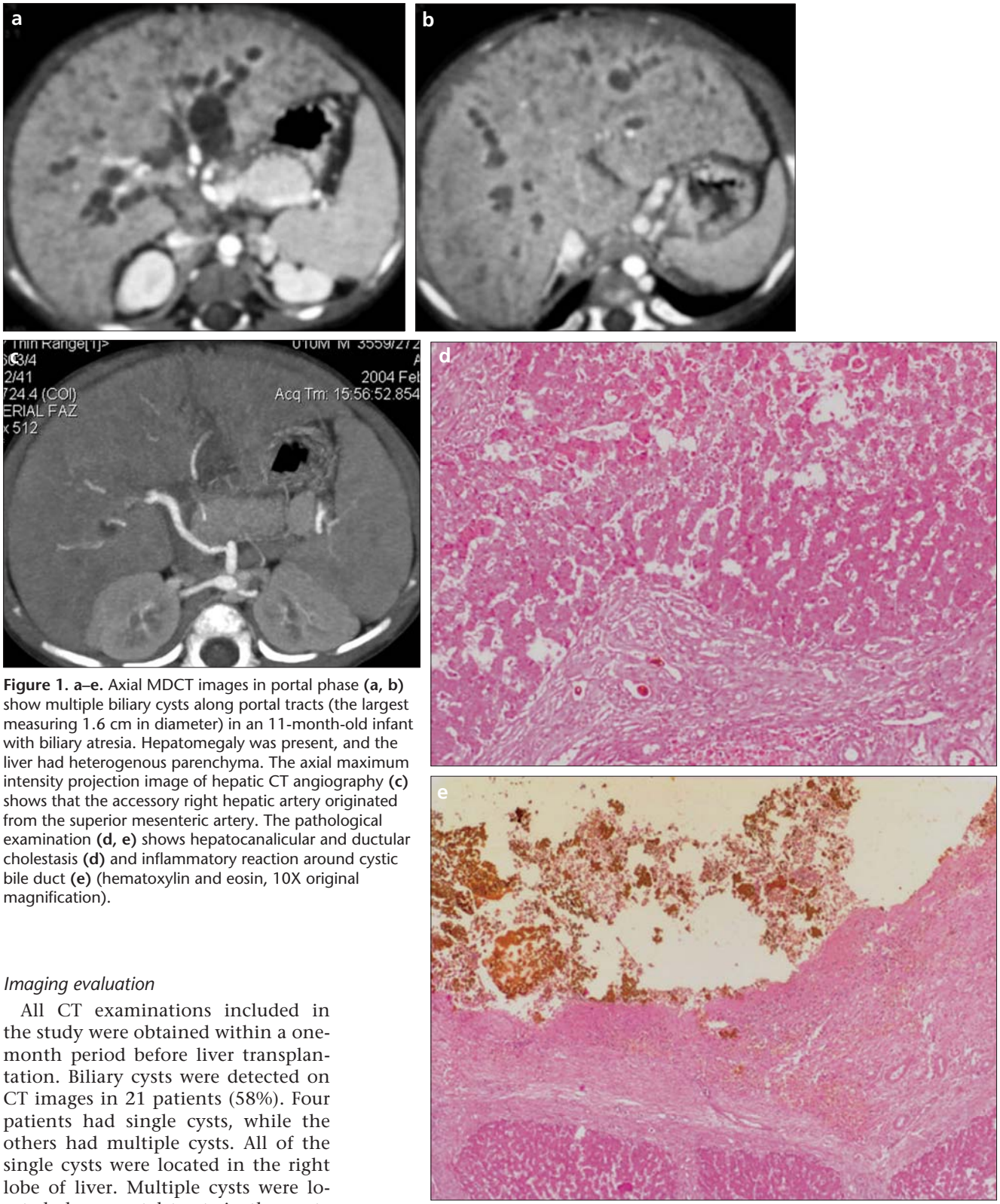

Figure 1. a–e. Axial MDCT images in portal phase **(a, b)** show multiple biliary cysts along portal tracts (the largest measuring 1.6 cm in diameter) in an 11-month-old infant with biliary atresia. Hepatomegaly was present, and the liver had heterogenous parenchyma. The axial maximum intensity projection image of hepatic CT angiography **(c)** shows that the accessory right hepatic artery originated from the superior mesenteric artery. The pathological examination **(d, e)** shows hepatocanalicular and ductular cholestasis **(d)** and inflammatory reaction around cystic bile duct **(e)** (hematoxylin and eosin, 10X original magnification).

Imaging evaluation

All CT examinations included in the study were obtained within a one-month period before liver transplantation. Biliary cysts were detected on CT images in 21 patients (58%). Four patients had single cysts, while the others had multiple cysts. All of the single cysts were located in the right lobe of liver. Multiple cysts were located along portal tracts in the porta hepatis (Fig. 1). Eleven patients had cysts <1 cm in size, and 9 had cysts >1 cm in size (1–2.5 cm in diameter). In one patient with multiple cysts, cysts covered large parts of both lobes of the liver; the largest was 10 × 9 cm in diameter (Fig. 2). There were no changes in size and number of cysts in patients with multiple examinations.

Table 2 lists the CT findings in patients with and without biliary cysts. The most common abnormality observed on CT images in patients with biliary atresia was contour irregularity or lobularity (50%). Hepatomegaly (41.7%), heterogenous parenchyma (30.6%), and enlargement of the left

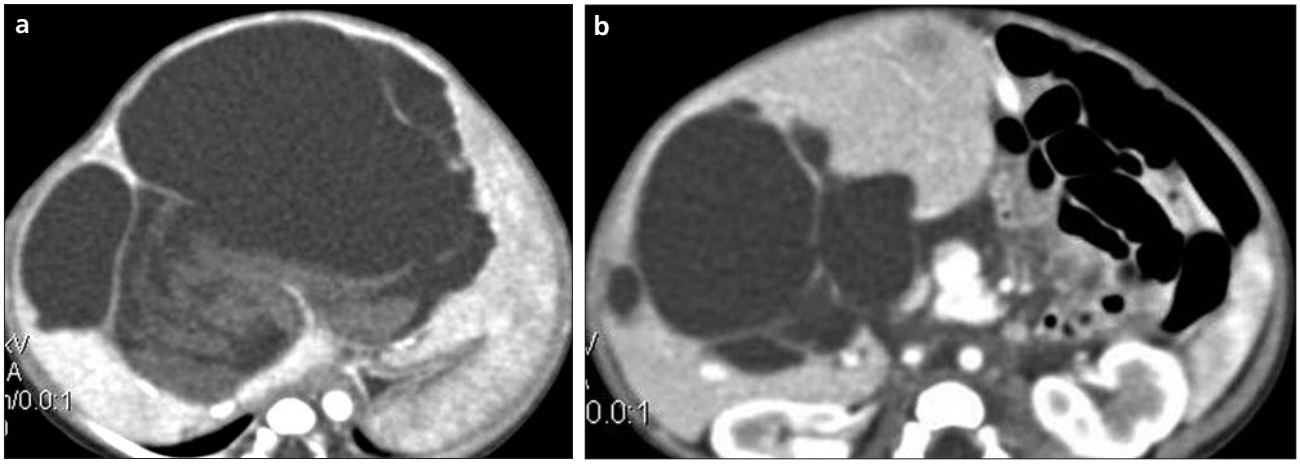

Figure 2. a, b. Axial MDCT images in arterial phase **(a, b)** show multiple multiloculated giant cysts in both lobes of the liver with hyperdense debris in posterior part of the biggest lesion in the left lobe in a 1-month-old patient. The hyperdense material was 32 HU in precontrast images (not shown). The patient died due to intraabdominal hemorrhage after cystojejunostomy procedure.

or caudate lobe (27.8%) were also common liver findings in patients with biliary atresia. The frequency of liver findings did not differ significantly between two groups, with the exception of heterogenous parenchyma, which had limited significance ($P = 0.058$). Signs of portal hypertension (splenomegaly [75%], collateral formation [47.2%], ascites [41.7%]) were also commonly detected on CT images and did not differ significantly between the two groups. Hepatic artery variations were significantly more common in patients with biliary cysts ($P = 0.003$). Hepatic arterial anomalies detected with CT imaging were accessory left hepatic artery arising from the left gastric artery (4), trifurcation (1), accessory right hepatic artery arising from the SMA (3), and common hepatic artery arising from the aorta (1). There were two patients with polysplenia syndrome without biliary cysts in the liver. All hepatic artery anomalies detected in patients who underwent liver transplantation were confirmed during surgery.

Pathologic correlation

Pathologic correlation was obtained in 18 cases in patients with or without biliary cysts (Table 3). The time between CT and pathologic examination of surgical specimens was 1 to 30 days. Thirteen of 18 patients with biliary atresia had biliary cysts in hepatectomy specimens. All of the cysts detected with pathologic examination were multiple cysts. All cysts were covered with biliary epithelial cells. The number of patients who had detection of biliary cysts by MDCT imaging and pathological evaluation did not differ between the groups, as assessed by McNemar test. All patients with biliary atresia had hepatocanalicular cholestasis and foci of hepatocellular necrosis in hepatectomy specimens. All patients with biliary atresia, except one patient with precirrhotic stage (stage 3), had stage 4 biliary cirrhosis of liver. The only statistically significant different findings between patients with and without biliary cysts were bile duct damage and inflammatory reaction, which were more common in the patients with biliary cysts ($\chi^2 = 0.017$, $P > 0.05$) (Fig. 1).

Table 2. Clinical and laboratory findings in children with biliary atresia with and without biliary cysts

	Patients			
Clinical and laboratory findings	Total (n, %) n = 36	With cysts (n, %) n = 21	Without cysts (n, %) n = 15	χ^2, P
Those with portoenterostomy	27 (75)	16 (76)	11 (73)	$\chi^2 = 0.845$, $P > 0.05$
Those with clinical history of cholangitis	11 (30.6)	4 (19)	7 (46.7)	$\chi^2 = 0.076$, $P > 0.05$
Age (month) (mean ± SD)	14.17 ± 21.0	13.15 ± 19.3	15.53 ± 23.7	$P^a = 0.893$
AST (mean ± SD)	342.94 ± 432.97	366.14 ± 539.96	310.47 ± 224.09	$P^a = 0.395$
ALT (mean ± SD)	169.47 ± 232.16	195.76 ± 296.46	132.67 ± 81.87	$P^a = 0.748$
Total bilirubin (mean ± SD)	19.52 ± 12.19	17.74 ± 11.06	22.01 ± 13.62	$P^a = 0.432$
Direct bilirubin (mean ± SD)	12.60 ± 7.99	11.91 ± 7.68	13.57 ± 8.58	$P^a = 0.724$

[a]Mann-Whitney U test
AST, aspartate transaminase; ALT, alanine transaminase; SD, standard deviation.

Discussion

Biliary atresia is a progressive disorder characterized by an inflammatory, sclerotic process of the extrahepatic and intrahepatic bile ducts with resultant ductular luminal obliteration and the development of biliary cirrhosis. Numerous ultrasonographic features have been described as useful for the diagnosis of biliary atresia.

Abnormalities in the shape and the wall of the gallbladder have yielded sensitivities and specificities >90% in the diagnosis of biliary atresia (10). The triangular cord sign, a focal area of increased echogenicity anterior to the bifurcation of the portal vein representing the fibrotic remnant of the extrahepatic biliary tree in biliary atresia, has been considered an important diagnostic feature. Kendrick et al. reported that gallbladder "ghost triad" is a very accurate sign of biliary atresia in ultrasound examination. This consists of an atretic gallbladder length <1.9 cm; thinned or absent smooth, complete echogenic mucosal lining with indistinct walls; and a knobbly, irregular, or lobular contour (11). The most serious complications of the disease are cholangitis (40–60%), and portal hypertension (35–75%) (3).

Development of intrahepatic biliary cysts or bile lakes has been reported in some patients with this disease; these findings may be caused by obstruction of biliary radicles by surgical portoenterostomy, cirrhotic changes of the liver, ongoing inflammation in the portal area, intra- or extrahepatic fibro-obliterative process, cholangitis, and ductal plate formation (12–15). The reported incidence of intrahepatic biliary cysts or bile lakes in biliary atresia ranged from 18% to 25% in reported *in vivo* studies and 24% to 36% in autopsy series (12–17). The prevalence of cysts in our series (58%) is the highest reported in the literature. In addition, of 21 cases with intrahepatic cysts, 17 (90%) had multiple cysts. These findings may be attributed to the advanced stage of disease, as most patients referred to our hospital for pretransplantation evaluation have cirrhosis. In addition, genetic factors may have a role; intrahepatic cyst formation is more commonly reported in Asian countries. Nine patients who did not undergo the Kasai procedure developed biliary cysts. Biliary cysts may occur, therefore, in patients with poor biliary drainage and cirrhosis regardless of the Kasai procedure. The only statistically significant finding detected with CT imaging is hepatic artery variations, which were more common in patients with intrahepatic cysts. Hepatic artery variations were detected in 9 of 36 patients with biliary atresia (25%); all patients with hepatic artery anomalies had intrahepatic cysts. Although anomalous origin of the hepatic artery is known to be associated with biliary atresia (18, 19), the cause of the high incidence in our series is unknown. Biliary and vascular anomalies have also been reported in patients with choledochal cysts (20). They have clinical importance since they must be diagnosed preoperatively to prevent intraoperative complications, particularly hemorrhage. MDCT is an effective imaging modality for detecting anatomic variations with the capability of multiplanar reconstructions. Other anomalies reported in patients with biliary atresia are polysplenia, bilateral bilobed lungs, preduodenal portal vein, azygos continuation of inferior vena cava, intestinal malrotation, and situs inversus (21, 22). There were two patients with polysplenia syndrome in our study.

In the majority of patients in this study the development of multiple cysts was not associated with concurrent clinical infections or with the pathologic examination, in contrast to other studies in literature. Bu et al. reported that fifteen patients with intrahepatic cysts (93.8%) had a history of cholangitis. After antibiotic treatment, cysts decreased in size in seven patients and disappeared in one patient in their study (23). Watanabe et al. also reported eight of twelve patients with biliary cysts had cholangitis, and six with multiple cysts were refractory to antibiotic treatment. They suggested that intrahepatic biliary cysts without cholangitis are not a source of infection and require no treatment, but they also suggested that patients with multiple complicated intrahepatic cysts have poor prognosis and require liver transplantation to control cholangitis (24). Takahashi et al. reported five

Table 3. Pathological findings in children with biliary atresia with and without biliary cysts

	Patients			
Pathological findings	Total (n, %) n = 18	With cysts (n, %) n = 13	Without cysts (n, %) n = 5	χ^2, *P*
Precirrhotic stage	1 (5.6)	1 (7.7)	0	$\chi^2 = 0.523$, $P > 0.05$
Cirrhosis	17 (94.4)	12 (92.3)	5 (100)	$\chi^2 = 0.523$, $P > 0.05$
Ductular cholestasis	14 (77.8)	11 (84.6)	3 (60.0)	$\chi^2 = 0.261$, $P > 0.05$
Hepatocanalicular cholestasis	18 (100)	13 (100)	5 (100)	
Foci of hepatocellular necrosis	18 (100)	13 (100)	5 (100)	
Cholangitis	2 (11.1)	2 (15.4)	0 (0)	$\chi^2 = 0.352$, $P > 0.05$
Bile plug-pseudorosette formation	16 (88.9)	12 (92.3)	4 (80)	$\chi^2 = 0.457$, $P > 0.05$
Bile duct damage and inflammatory reaction	14 (77.8)	12 (92.3)	2 (40)	$\chi^2 = 0.017$, $P < 0.05$
Presence of bile duct	2 (11.1)	1 (7.7)	1 (20)	$\chi^2 = 0.457$, $P > 0.05$
Biliary cysts	18 (100)	13 (100)	5 (100)	

of seven patients had intrahepatic cysts discovered during an episode of cholangitis (25). In our study, 11 of 36 patients had at least one clinical episode of cholangitis (31%); only four of them (36%) had cysts. The lower incidence of cholangitis may be attributed to short follow-up period of patients. But in 9 of 11 patients with cholangitis, cholangitis was refractory to antibiotic treatment, and the patients underwent liver transplantation. These findings show that cholangitis is not always associated with cyst formation, for which factors other than cholangitis may have a role.

No imaging findings or biochemical results had predictive value for the presence of cysts. The cysts were not associated with CT findings that might indicate more advanced biliary atresia, such as abnormal liver size, contour irregularity, parenchymal liver changes, splenomegaly, ascites, and portosystemic collateral vessels. The liver enzymes and serum bilirubin levels were abnormal in both groups except one patient with single intrahepatic cysts. The difference in liver enzyme levels was not statistically significant between the two groups, although the mean values of aspartate transaminase (AST) and alanine transaminase (ALT) were higher in patients with biliary cysts. Interestingly, the mean serum bilirubin levels were higher in the patients without biliary cysts. These results show that the cysts were not associated with aggravation of cholestasis.

The pathogenesis of cyst formation is unknown; proposed thories include intrahepatic duct fibro-obliterative process leading to erosion and ulceration of biliary epithelium, resulting in bile leakage (12); exaggeration of the irregular configuration of intrahepatic bile ducts during the course of cirrhotic changes leading to multicystic dilatation (13); and fetal ductal plate malformation (14). In addition, immune-mediated mechanisms of damage to bile duct epithelium may play a role in the pathogenesis of disease, with ongoing inflammation leading to cyst formation (15). Fonkalsrud and Arima referred to intrahepatic cystic lesions found at autopsy as bile lakes and described them histologically as pseudocysts surrounded by a rim of fibrous connective tissue (17). Fain and Lewin, however, reported that intrahepatic biliary cysts were lined by cuboidal and flattened epithelial cells (12). Betz et al. also reported the histology of biliary cysts lined with epithelium (6). Tainaka et al. reported that distinction of two separate entities, bile lakes and dilated bile ducts, are important in the treatment strategy for intrahepatic cystic lesions; the former indicates a poor prognosis, and the latter can be improved surgically without transplantation (26, 27). In our study, all of the cysts detected in pathological examination were multiple; we did not evaluate the single cysts in pathological examination detected by MDCT in four patients. Thus we preferred the name "bile cysts" instead of bile lakes. Histologically, the only statistically significant difference between the patients with and without biliary cysts was damage to the bile duct epithelium and inflammatory reaction around the cysts. Cholangitis was only found in two patients at the time of examination. Development of these cysts did not correlate with hepatic function, portoenterostomy surgery, the extent of morphologic change in the liver, or signs of portal hypertension in our study. These findings support the notion that cysts form in response to a fibro-obliterative process. A bile leak then occurs from the cyst wall, followed by inflammation in the same area. In addition, in our series all patients except one had abnormal liver enzymes and serum bilirubin levels and were in the cirrhotic stage, although portoenterostomy procedure had been done in 27 of 36. This implicates ongoing liver inflammation and fibrosis in the etiology of biliary atresia. Liver transplantation is thus indicated in the majority of patients, and the prognosis is good.

In conclusion, multiple intrahepatic cyst formation is a common complication in patients with biliary atresia. There was no association between the clinical features of patients and MDCT findings of biliary cysts in our patients, who had advanced disease. But damage to the bile duct epithelium and inflammatory reaction around the biliary epithelium was significantly higher in patients with biliary cysts. This supports the theory of obstruction and bile leaks in the etiogenesis of biliary cysts. This is the first report of the association between hepatic artery variations and the biliary cysts; this may be an important factor in pretransplant evaluation.

References

1. Pall H, Jonas MM. Pediatric hepatobiliary disease. Curr Opin Gastroenterol 2005; 21:344–347.
2. Balistreri WF. Neonatal cholestasis: medical progress. J Pediatr 1985; 106:171–184.
3. Narkewicz MR. Biliary atresia: an update on our understanding of the disorder. Curr Opin Pediatr 2001; 13:435–440.
4. Kasai M, Suzuki S. A new operation for 'non correctable' biliary atresia: portoenterostomy. Shijitsu 1959; 13:457–481.
5. Takahashi A, Tsuchida Y, Suzuki N, et al. Incidence of intrahepatic biliary cysts in biliary atresia after hepatic portoenterostomy and associated histopathologic findings in the liver and porta hepatis at diagnosis. J Pediatr Surg 1999; 34:1364–1368.
6. Betz BW, Bisset GS III, Johnson ND, Daugherty CC, Balistreri WF. MR imaging of biliary cysts in children with biliary atresia: clinical associations and pathologic correlation. AJR Am J Roentgenol 1994; 162:167–171.
7. Nakama T, Kitamura T, Matsui A, Makino S, Senyuz OF, Kanazawa K. Ultrasonographic findings and management of intrahepatic biliary tract abnormalities after portoenterostomy. J Pediatr Surg 1991; 26:32–36.
8. Konus OL, Ozdemir A, Akkaya A, Erbas G, Celik H, Isık S. Normal liver, spleen and kidney dimensions in neonates, infants and children: evaluation with sonography. AJR Am J Roentgenol 1998; 171:1693–1698.
9. Scheuer PJ. Primary biliary cirrhosis: chronic non-suppurative destructive cholangitis. Am J Pathol 1965; 46:387.
10. Humphrey TM, Stringer MD. Biliary atresia: US diagnosis. Radiology 2007; 244:845–851.
11. Tan Kendrick AP, Phua KB, Ooi BC, Tan CE. Biliary atresia: making the diagnosis by the gallbladder ghost triad. Pediatr Radiol 2003; 33:311–315.
12. Fain JS, Lewin KJ. Intrahepatic biliary cysts in congenital biliary atresia. Arch Pathol Lab Med 1989; 113:1383–1386.
13. Tsuchida Y, Honna T, Kawarasaki H. Cystic dilatation of the intrahepatic biliary system in biliary atresia after hepatic portoenterostomy. J Pediatr Surg 1994; 29:630–634.
14. Takahashi A, Tsuchida Y, Hatakeyama S, et al. A peculiar form of multiple cystic dilatation of the intrahepatic biliary system found in a patient with biliary atresia. J Pediatr Surg 1997; 32:1776–1779.
15. Dillon P, Belchis D, Tracy T, Cilley R, Hafer L, Krummel T. Increased expression of intercellular adhesion molecules in biliary atresia. Am J Pathol 1994; 145:263–267.
16. Danks DM, Campbell PE. Extrahepatic biliary atresia: comments on the frequency of potentially operable cases. J Pediatr 1966; 64:21–29.
17. Fonkalsrud EW, Arima E. Bile lakes in congenital biliary atresia. Surgery 1975; 77:384–390.
18. Day DL, Mulcahy PF, Dehner LP, Letourneau JG. Post-operative abdominal CT scanning in extrahepatic biliary atresia. Pediatr Radiol 1989; 19:379–382.

19. Casaccia G, Catalano OA, Marcellini M, Bagolan P. Biliary atresia associated with multiple unrelated anomalies: what about it? Pediatr Surg Int 2007; 23:277–280.
20. Todani T, Watanabe Y, Toki A, Ogura K, Wang ZQ. Co-existing biliary anomalies and anatomical variants in choledochal cyst. Brit J Surg 1998; 85:760–763.
21. Davenport M, Savage M, Mowat AP, Howard ER. Biliary atresia splenic malformation syndrome: an etiologic and prognostic subgroup. Surgery 1993; 113:662–668.
22. Carmi R, Magee CA, Neill CA, Karrer FM. Extrahepatic biliary atresia and associated anomalies: etiologic heterogeneity suggested by distinctive patterns of associations. Am J Med Genet 1993; 45:683–693.
23. Bu LN, Chen HL, Ni YH, et al. Multiple intrahepatic biliary cysts in children with biliary atresia. J Pediatr Surg 2002; 37:1183–1187.
24. Watanabe M, Hori T, Kaneko M, et al. Intrahepatic biliary cysts in children with biliary atresia who have had a Kasai operation. J Pediatr Surg 2007; 42:1185–1189.
25. Takahashi A, Tsuchida Y, Suzuki N, et al. Intrahepatic biliary cysts in biliary atresia in the era of liver transplantation. J Pediatr Gastroenterol Nutrition 2003; 36:608–612.
26. Tainaka T, Kaneko K, Seo T, Ono Y, Sumida W, Ando H. Intrahepatic cytsic lesions after hepatic portoenterostomy for biliary atresia with bile lake and dilated bile ducts. J Pediatr Gastroenterol Nutrition 2007; 44:104–107.
27. Tainaka T, Kaneko K, Nakamura S, Ono Y, Sumida W, Ando H. Histological assessment of bile lake formation after hepatic portoenterostomy for biliary atresia. Pediatr Surg Int 2008; 24:265–269.