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Original article

Clinical characteristics of cystic biliary atresia

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Abstract

Background: Cystic biliary atresia (BA) is a unique form of BA that can be mistaken for a choledochal cyst. This study reviews the clinical characteristics of cystic BA to better understand its nature.
Objective: To characterize cystic biliary atresia in terms of clinical presentation, imaging findings, and surgical outcomes, and to highlight key features that aid in differentiating it from choledochal cysts.
Methods: We retrospectively reviewed the charts of all infants diagnosed with cystic BA from January 2005 to December 2019. The diagnosis of BA was confirmed in all cases by intraoperative cholangiography. Moreover, clinical data, pre-operative laboratory and imaging, intraoperative findings, and postoperative outcome were recorded, and data were expressed as mean ± standard deviation.
Results: Out of the 227 patients with BA, there were 14 patients with cystic BA (6.2%) during the studied period. The mean pre-operative ultrasound size at the maximal part of the cyst was 2.1 ± 1.2 cm, and 11 of 14 patients underwent a Kasai operation. The mean age at the time of Kasai operation was 95.3 ± 37.6 days, and only 1 in 11 patients (9.0%) underwent a Kasai operation before 60 days. Moreover, 6 patients (54.5%) underwent a Kasai operation at 61–90 days, whereas 4 patients (36.3%) underwent a Kasai operation at 113–154 days. Intraoperative findings revealed BA type III proximal to the cysts in all cases. Based on the surgeons' decision, 3 patients did not receive a Kasai operation, and only a liver biopsy was performed. After the Kasai operation, 8 out of 11 patients (72.7%) experienced no jaundice within 3 months. In addition, gross cirrhosis was recorded by the surgeons in 9 patients, and of these patients, 7 underwent a Kasai operation. Interestingly, 5 of the 7 patients with gross cirrhosis undergoing surgery (71.4%) were jaundice-free within 3 months. Briefly, an overall jaundice-free state was achieved in 72.7% of patients with cystic BA who underwent a Kasai operation. In addition, 71.4% of patients with cystic BA and gross cirrhosis undergoing surgery were jaundice-free.
Conclusion: Cystic BA is a rare pathology that needs to be distinguished from choledochal cysts in neonates. Awareness of the existence of cystic BA may facilitate the diagnosis and dissection around the porta hepatis area during the Kasai operation. This may increase the chance of better results for the patients. Even in the presence of gross cirrhosis, the Kasai procedure may still be considered to provide an opportunity for bile drainage.

Keywords: Cystic biliary atresia, jaundice, Kasai operation.

Cystic biliary atresia (BA) is a unique form of BA that can be mistaken for a choledochal cyst.[1, 2] The clinical manifestation of obstructive jaundice together with direct hyperbilirubinemia during the neonatal period is a common presentation in cystic BA as well as in choledochal cysts. Although the triangular cord sign in the ultrasonographic findings has high diagnostic performance for BA[3, 4], this feature does not appear at the early stage of the disease, which results in reduced sensitivity.[5, 6] Although there are some differences in the pathological characteristics between cystic BA and choledochal cysts, there may also be some overlap.[7] Intraoperative cholangiography is usually required to confirm the differences between the two conditions. In addition, this technique is able to classify the type of BA that affects the prognosis.[7-13]

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Liver fibrosis can occur in neonatal obstructive cholangiopathy and may result in liver cirrhosis, which is an irreversible pathological process. According to previous studies, choledochal cysts in neonates and infants have a greater chance to develop biliary cirrhosis than older children, but the prognosis is better when compared to that of patients with cystic BA.[14-16] Although the prognosis of cystic BA is likely to be better than that of other patients with BA, there is a high correlation with the age at Kasai operation compared with that among patients with BA[10, 17-19]; however, strong evidence is still lacking. As cystic BA is extremely rare, its clinical features receive little attention. Therefore, our objective was to review the clinical characteristics of cystic BA, based on our 15 years of experience, to better understand this rare form of BA.

Materials and methods

This retrospective chart review study involving human participants was performed in accordance with the ethical standards of the institutional and national research committees and with the 1964 Declaration of Helsinki and its later amendments or comparable ethical standards. The Institutional Review Board (IRB) of Chulalongkorn University reviewed and approved this study (IRB no. 543/61). We retrospectively reviewed the charts of all infants diagnosed with cystic BA between January 2005 and December 2019 at two university hospitals (King Chulalongkorn Memorial Hospital, Thai Red Cross Society, Chulalongkorn University and Srinagarind Hospital, Khon Kaen University). The diagnosis of BA was confirmed in all cases by exploratory laparotomy and intraoperative cholangiography. The clinical data, preoperative laboratory and imaging, intraoperative findings, and post-operative outcomes were reviewed and analyzed. Data were expressed as the mean ± standard deviation (SD). Continuous variables were analyzed using the Student's *t* test when they followed a normal distribution and the Wilcoxon rank-sum test for those with a non-normal distribution. Statistical analysis was performed using STATA version 14.0, and $P < 0.05$ was considered statistically significant.

Results

Clinical data

Out of the 227 patients with BA, there were 14 patients with cystic BA (6.2%) during the studied period. There was no prenatal diagnosis of the cyst in all 14 patients. The number of males and females was equal (male:female, 7:7), and 11 of 14 patients underwent a Kasai operation. The mean age at the time of the Kasai operation was 95.3 ± 37.6 days. For the pre-operative laboratory evaluations, the mean serum total bilirubin (TB), direct bilirubin (DB), aspartate aminotransferase (AST), and alanine aminotransferase (ALT) were 13.2 ± 3.6 mg/dL, 9.6 ± 3.0 mg/dL, 266.1 ± 115.8 IU/L, and 217.1 ± 112.7 IU/L, respectively. Moreover, 13 out of the 14 patients (92.9%) had preoperative ultrasonography, and the mean size at the maximal part of the cyst was 2.1 ± 1.2 cm. A representative ultrasound is shown in **Figure 1**. Eight patients (57.1%) received an additional diisopropyl iminodiacetic acid (DISIDA) scan, with no excretion of the tracer in any of the cases, as shown in **Table 1**. Only 1 in 11 patients (9.0%) underwent a Kasai operation before 60 days of age. In addition, 6 patients (54.5%) underwent a Kasai operation between 61 and 90 days, whereas 4 patients (36.3%) underwent a Kasai operation between 113 and 154 days.

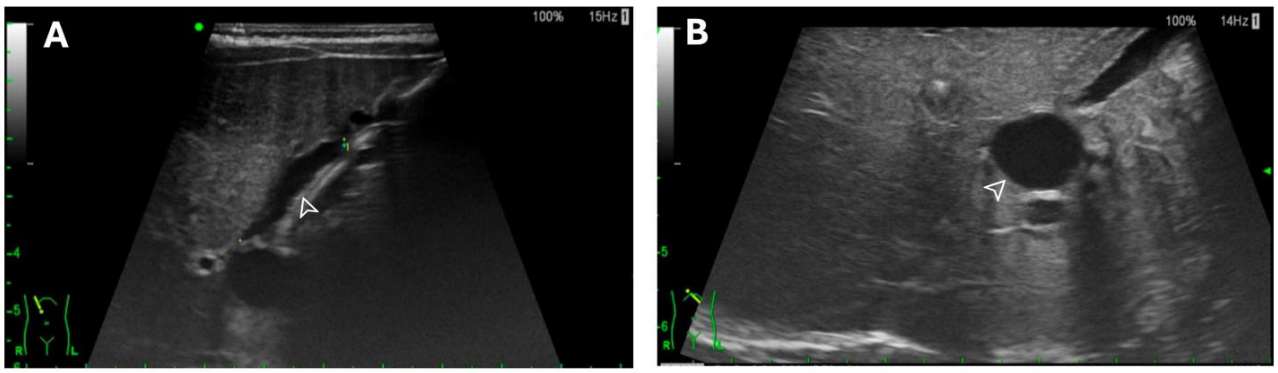

Figure 1. Abdominal ultrasonography of an 87-day-old girl demonstrated **(A)** a gallbladder length 2.6 cm (arrowhead), **(B)** a 1.1 cm diameter cyst (arrowhead) with cirrhotic liver. At latest follow-up, the patient was 1 year 8 months without jaundice.

Table 1. Characteristics and outcome of BA children.

Case number	Gender	Cyst size (maximum part from USG; cm)	DISIDA scan excretion	Pre-operative laboratory						Gross cirrhosis liver	Age at Kasai operation (days)	Jaundice-free after Kasai op.
				TB	DB	AST	ALT	ALP	GGT			
1	Female	2.5	No excretion	12.0	8.6	172.0	109.0	541.0	–	–	70	Yes
2	Male	–	No excretion	13.8	10.1	338.0	286.0	535.0	–	–	79	Yes
3	Male	2.4	–	6.5	4.3	101.0	210.0	–	–	No	30	Yes
4	Female	3.7	–	14.0	9.5	115.0	215.0	–	1054.0	Yes	135	Yes
5	Female	1.1	–	10.2	8.8	192.0	116.0	487.0	1112.0	Yes	87	Yes
6	Female	2.3	No excretion	8.6	8.2	376.0	199.0	265.0	639.0	Yes	154	Yes
7	Female	1.2	–	10.4	9.0	274.0	163.0	657.0	2009.0	No	113	No
8	Male	0.7	–	19.3	14.9	524.0	457.0	479.0	425.0	Yes	No Kasai operation	
9	Male	2.6	No excretion	14.3	12.7	170.0	116.0	352.0	1402.0	Yes	67	Yes
10	Male	4.8	–	15.0	14.3	261.0	106.0	580.0	120.0	Yes	No Kasai operation	
11	Female	1	No excretion	12.3	5.5	311.0	174.0	368.0	–	Yes	86	No
12	Female	2.8	No excretion	19.2	7.9	216.0	438.0	247.0	–	–	No Kasai operation	
13	Male	1	No excretion	13.6	9.5	307.0	271.0	549.0	–	Yes	82	Yes
14	Male	1	No excretion	15.2	11.3	369.0	179.0	477.0		Yes	145	No

ALP, alkaline phosphatase; ALT, alanine aminotransferase; AST, aspartate aminotransferase; BA, biliary atresia; DB, direct bilirubin; DISIDA, diisopropyl iminodiacetic acid; GGT, gamma-glutamyl transferase; TB, total bilirubin; USG, ultrasonography. – indicates missing data.

Operative data and outcome

Intraoperative findings revealed atresia of the right and left hepatic ducts (type III) proximal to the cysts in all cases. Three patients did not receive a Kasai operation, based on their surgeons' decision, either because of advanced age (199.7 ± 13.3 days) or gross cirrhosis; the latter was defined intraoperatively by the macroscopic nodular appearance of the liver, which was pathologically confirmed via liver biopsy. After the Kasai operation, 8 out of 11 patients (72.7%) achieved a jaundice-free state (defined as serum TB < 1.0 mg/dL) within 3 months. The mean age at surgery of jaundice-free patients was 88.0 ± 39.3 days, whereas the mean age at surgery of patients with persistent jaundice was 114.7 ± 29.5 days ($P = 0.28$). Gross cirrhosis was recorded in nine patients, and seven of these nine patients received a Kasai operation. Interestingly, 5 of the 7 patients with gross cirrhosis who underwent surgery (71.4%) were jaundice-free within 3 months, as shown in **Figures 2 and 3.**

In summary, an overall jaundice-free rate was achieved in 72.7% of patients with cystic BA who underwent a Kasai operation. In addition, 71.4% of patients with cystic BA and gross cirrhosis were jaundice-free. Finally, 70% of patients with cystic BA aged > 60 days were jaundice-free.

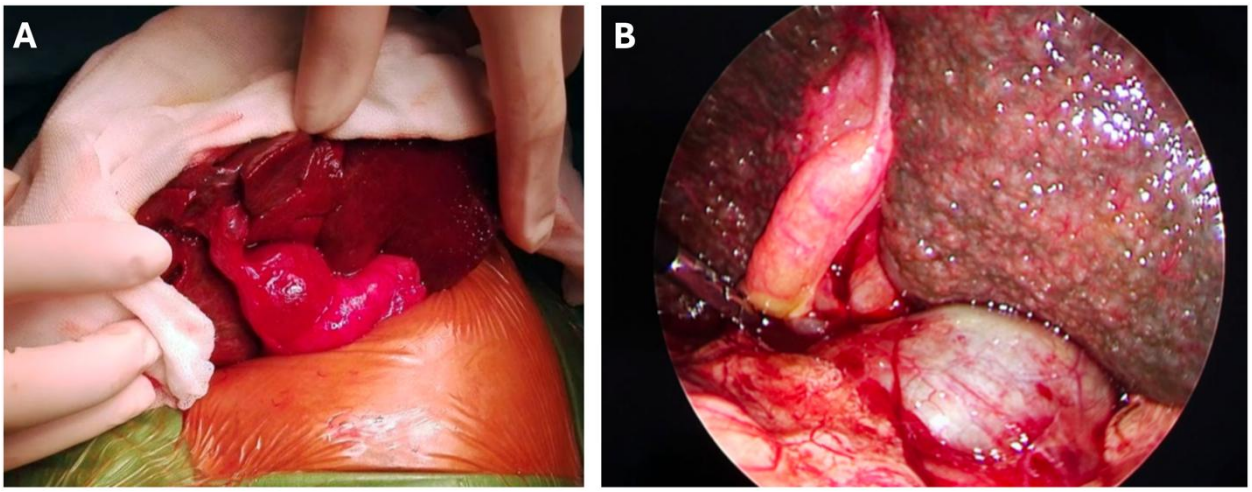

Figure 2. Intraoperative findings: **(A)** a 30-day-old male infant with a 2.38 × 1.69 cm cyst and no cirrhosis, **(B)** a 135-day-old girl revealed a 3.66 × 2.02 cm cyst and gross cirrhosis. Both patients remained jaundice-free at the latest follow-up (2.5 years and 10 months, respectively).

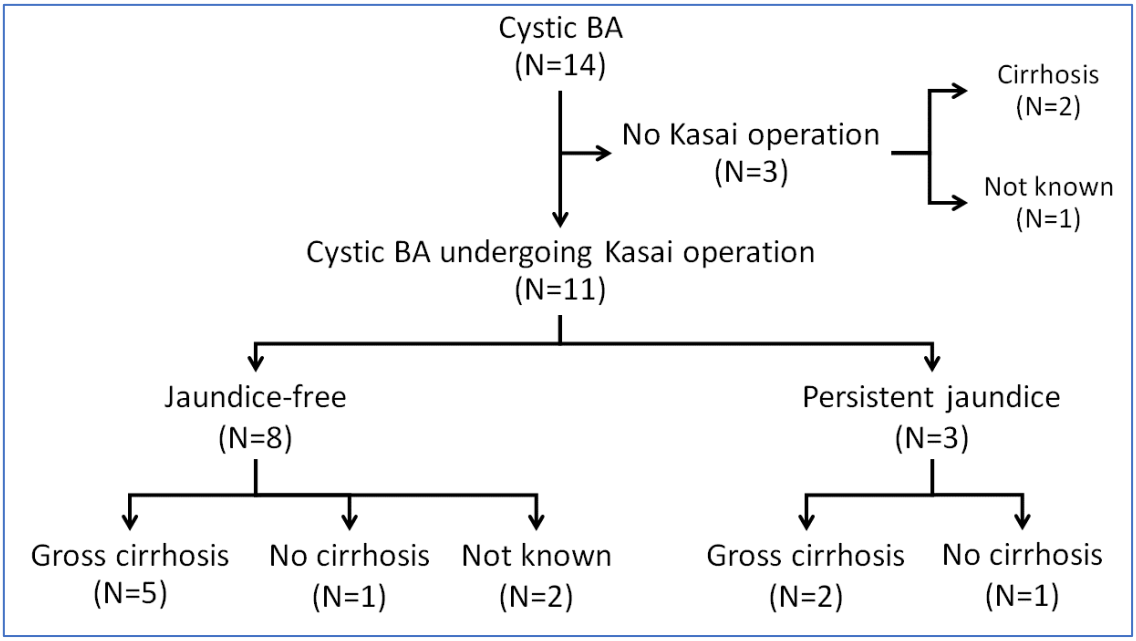

Figure 3. The summary of 14 cystic BA patients according to their outcome and gross cirrhosis. BA, biliary atresia.

Discussion

BA is an obstructive condition of the bile ducts that causes jaundice in newborns and can result in irreversible liver cirrhosis later in life. Anatomic and cholangiography findings are used to classify this condition and are usually divided into three groups depending on the location of the obstruction according to the Japanese Society of Pediatric Surgeons classification, of which the most common is type III atresia at the porta hepatis.[20] The presence of an extrahepatic cyst is one of the variant types of BA[1], which are often referred to as cystic BA. This condition is found in approximately 5%–20% of cases.[10-12, 17, 18] Similarly, the results of our study found that this condition occurred in 6.2% of cases, and all fell into classification type III. The key data points, including pre-operative clinical data, intraoperative findings, and patient outcomes, are summarized in **Figure 4.**

Neonates with clinical features of obstructive jaundice and cystic mass at the extrahepatic bile duct are often similar to those of choledochal cysts and cystic BA. Distinguishing between these two conditions is sometimes challenging. According to several studies, sonographic findings showed the size of the cystic structure at the porta hepatis ranged from 0.4 to 4.7 cm in patients with cystic BA, whereas in patients with choledochal cysts, this was 2–13 cm.[4, 13, 14, 21, 22] This is similar to the result of our study, where the mean size at the maximal part of the cyst from preoperative ultrasonography was 2.1 ± 1.2 cm. However, there were reports of a large cyst and dilated hepatic ducts in cystic BA cases.[11, 21] Therefore, it is necessary to confirm the diagnosis via intraoperative cholangiography.

Generally, BA types I and II have a good prognosis, while type III, the most common type, has a less desirable outcome.[20, 23, 24] Similarly, the outcome of cystic BA type III was comparable to that of BA type III without a cyst.[8, 24] In addition, age at portoenterostomy is an important prognostic factor in patients with BA. Some studies have shown that the prognosis of cystic BA has a high correlation with the age at Kasai portoenterostomy compared with that of patients with BA.[10, 17-19] All our cystic BA patients were classified as type III, and the mean age at the time of surgery was 95.3 ± 37.6 days. Although the patients were considered to be within the poor prognosis category, most did not have jaundice within 3 months after the operation (72.7%). Furthermore,

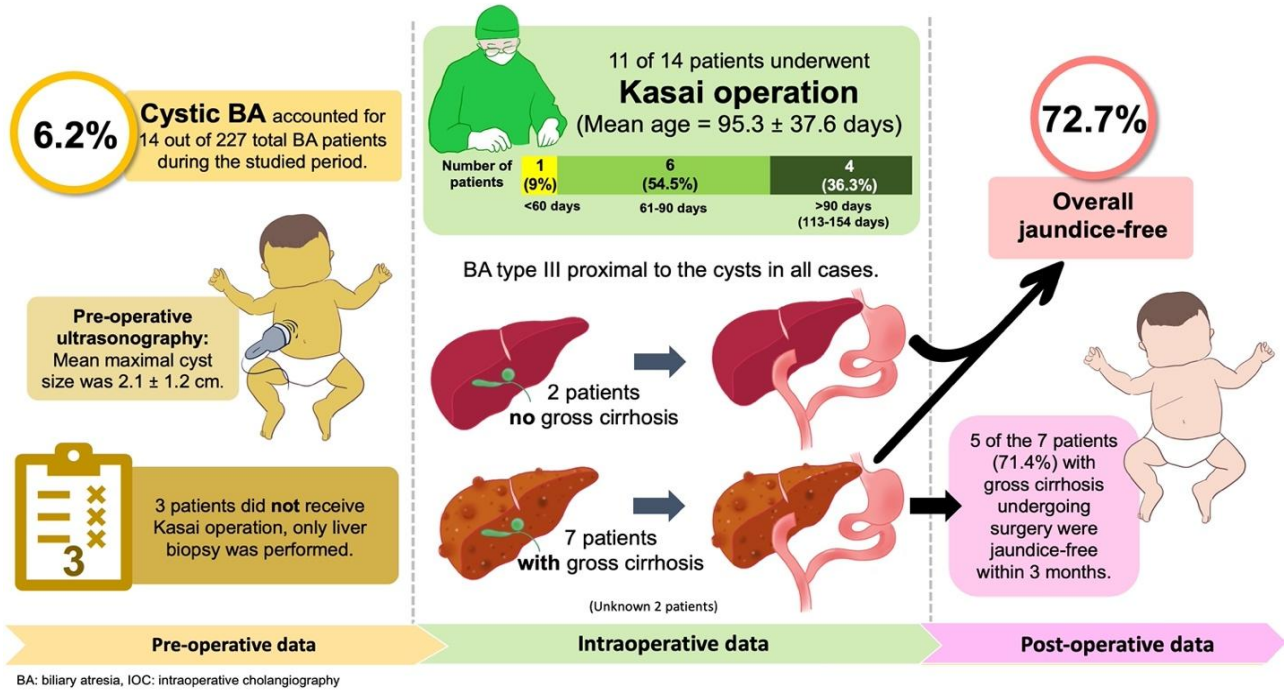

Figure 4. Summary of patient characteristics, management, and clinical outcomes of cystic BA patients. BA, biliary atresia.

the mean age at surgery was not different between the groups with and without jaundice. The age at surgery in this study was quite late compared to other studies, which is probably because of the inability to detect the cyst prenatally. Moreover, the decision to proceed with the Kasai procedure was influenced by the patient's financial constraints and the goal to delay the need for liver transplantation. Consequently, 5 of the 7 patients with gross cirrhosis at the time of surgery remained jaundice-free during the follow-up period. While we acknowledge that intra-abdominal adhesions from the Kasai procedure could potentially make subsequent transplantation more challenging, our previous institutional data demonstrated that a prior Kasai operation did not adversely affect perioperative outcomes (including operative time, intraoperative blood loss, and postoperative complications) or survival rates in patients who eventually underwent liver transplantation.[25] Therefore, the outcome of cystic BA in this study is similar to those of other studies demonstrating better prognosis than BA type III without an extrahepatic cyst.[10, 17, 18, 24, 26-28]

This study has several limitations. First, the small sample size reflects the rarity of cystic BA; however, it may underpower certain statistical analyses. Second, the retrospective design resulted in missing data regarding the postoperative steroid administration and cytomegalovirus infection status, which may have potentially influenced the outcomes. In addition, inconsistent long-term follow-up precluded our assessment of progressive cirrhosis, the requirement for liver transplantation, or long-term survival rates. Thus, future prospective studies with more robust longitudinal tracking are essential.

Conclusion

Cystic BA is a rare and unique pathology that needs to be distinguished from choledochal cyst of the neonate. Awareness of this type of BA may facilitate the diagnosis and dissection around the porta hepatis area, which may increase the chances of more favorable patient results. The Kasai procedure may still be considered to provide an opportunity for bile drainage even when gross cirrhosis is present.

Author contributions

KT and PV contributed to the conception and design of the study, and to data acquisition, analysis, and interpretation. SC, KD, and NS contributed to data acquisition and interpretation. KT drafted the manuscript. All authors critically revised the manuscript and approved the final version for publication.

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Conflict of interest statement

All authors have completed and submitted the International Committee of Medical Journal Editors Uniform Disclosure Form for Potential Conflicts of Interest. All authors declare that they have no conflicts of interest.

Data sharing statement

All data generated or analyzed in the present study are included in the published article. Further details are available for non-commercial purposes from the corresponding author upon reasonable request.

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