

DIAGNOSIS AND MANAGEMENT OF CHOLEDOCHAL CYST IN ADULT : AN EXPERIENCE IN A TERTIARY LEVEL HOSPITAL IN CHINA

A B M Kaiser¹ Subrata Barua¹ Siddartha Biswas¹ Nur Nahar²
Noor Mohammad³ Shamim Ara² Niu Wei⁴ Benjia Liang⁴ Jun Niu⁵

Summary

The cystic dilatation of the biliary tract is a rare disease and uncertain origin. It is recognized more frequently in children; however, its incidence comes increasing in adults, representing 20% of the cases. The aims of this study to demonstrate our experience with choledochal cyst and to evaluate morbimortality rates, discuss the aetiopathogenesis, presentation, management, and outcome with review of the literature. A retrospective study and review of the records of all the patients above 15 years, who underwent therapeutic intervention of choledochal cyst in Qilu Hospital of Jinan, China, was carried out. Ten cases of choledochal cyst were found, 8 female, with mean age 31 years. These included 8 cases of Todani type I and one case each of type II and type III. The predominant symptoms were abdominal pain and jaundice. Abdominal mass and past history of cholangitis and pancreatitis were seen in 2 patients. Investigations included ultrasound in 8 patients, CT in 7, ERCP in 3, and MRCP in 5. Surgical intervention included complete excision of the cyst with hepaticojejunostomy and cholecystectomy (Type I) excision of the diverticulum (Type II) and ERCP sphincterotomy (Type III). Malignancy was not seen in any patients. The long-term postoperative complications included cholangitis in two patients. Biliary tract cystic dilatation is a rare disease. However, its incidence is increasing in the adult population; so, it must be thought as differential diagnosis when facing obstructive jaundice.

Key words : Choledochal malformation; Choledochal cyst; Carolis disease; Congenital hepatic fibrosis.

Introduction

Choledochal Malformation (CDM) is a pathological condition characterized by varying degree of congenital dilatation of the biliary system including the common, intrahepatic, and intrapancreatic bile duct. Presence of significant dilatation constitutes Choledochal Cyst (CCD). This entity occurs more frequently in Asia than in western countries with most reports originating from Japan [1]. The incidence ranges from 1 in 13,000 in Japan to 1 in 2 million in England [1-3]. They usually manifest in children and very few of them present when adults. About 25% of CCD is diagnosed antenatal or within the first year of life, 60% during the first decade of life and 20% go undiagnosed into adulthood [3-5]. A marked female preponderance has been widely recognized (Female to male ratio 3 : 1) [3]. Presence of Anomalous Biliopancreatic Duct Junction (APBDJ) allowing pancreatic juice to reflux into biliary tree is the most widely accepted etiopathogenic concept [2]. CCD is associated with biliary tree stasis and lithiasis and the whole biliary epithelium is considered at risk of malignant transformation. Choledochal cyst was first described by Vater & Ezler in 1723 and resection of the cyst by McWhorter in 1924. In 1959 Alonzo-Lej et al. classified the cysts : A classification that was modified by Todani et al. in 1977 - based on the location of the cyst into five types (Fig 1) [4,5]. Type I is the classic and most common type, representing on average 85-90% of cases and consists of dilation of the bile duct, which may be cystic, focal or fusiform (Sub types A, B and C respectively). The cystic duct thus generally enters the choledochal cyst and the right and left hepatic ducts, while the intra-hepatic ducts are of normal diameter. Type II is the rarest of all choledochal cysts, accounting for less than 5% of cases. It is described as a simple diverticulum of the extra-hepatic biliary tree. Type III or choledochoceles is the cystic dilation of the

1 MS Student of Hepatobiliary Surgery (Thesis Part)
Qilu HOspital, Shandong University, Jinan, China

2 MD Student of Radiology (2nd Part)
Qilu HOspital, Shandong University, Jinan, China

3 MD Student of Cirtical Care Medicine
Qilu HOspital, Shandong University, Jinan, China

4 Consultant of Hepatobiliary Surgery
Qilu HOspital, Shandong University, Jinan, China

5 Professor of Hepatobiliary Surgery
Qilu HOspital, Shandong University, Jinan, China

Correspondence : Dr. A B M Kaiser

Email : dr.abmkaiser@yahoo.com

Cell : 01751 069690

intraduodenal portion of the extrahepatic biliary tree. Type IV, sub type A is the second most common type of choledochal cyst, defined as intra- and extra-hepatic dilation. Sub type B involves multiple dilations of the extra-hepatic biliary tree [6,7]. Finally type V, also known as Caroli's disease, is the cystic dilation of the intra-hepatic biliary system, which may be associated with periportal fibrosis and cirrhosis and may be bilobal or confined to a single lobe [8]. Complete cyst excision with cholecystectomy followed by biliary reconstruction using Roux-en-Y hepaticojejunostomy is the treatment of choice for the extrahepatic component of the disease (Type I and type VCCD). In type VCCD (Caroli's disease), liver resection is tailored to the extent of intrahepatic disease and the presence and severity of underlying chronic liver (Congenital hepatic fibrosis) and the associated kidney disease are taken into consideration. The pathogenesis of choledochal cyst remains uncertain, although it is believed that the lesions are congenital in origin and some are diagnosed by way of antenatal ultrasound [9]. Magnetic Resonance Cholangiopancreatography (MRCP) is currently the most accurate preoperative imaging study to assess cyst anatomy and classify the disease according to standard Todani classification (Fig 1) [3,4,10,11]. The present study aims to demonstrate our experience with choledochal cyst and discuss the aetiopathogenesis, presentation, management and outcome with review of the literature.

Material and methods

A retrospective study and review of records was carried out at the General Surgery Department of Qillu Hospital of Jinan, Shandong Province, China (A tertiary level hospital) in the period of 2012 and 2014.

The aims of this study to demonstrate our experience with choledochal cyst and discuss the aetiopathogenesis, presentation, management, and outcome with review of the literature.

Patients were included if they had been diagnosed with a bile duct cyst and undergone surgery. All the patients below 15 years were excluded from this study.

Demographic characteristics, such as age and sex, symptomology, blood biochemistry and imaging examinations were investigated. The study also covered the type of surgery performed, the classification of the bile duct cyst and post-operative complications and post operative follow up for one year. Statistical analysis were performed with Statistical Package for Social Science (SPSS 19.0 software).

Results

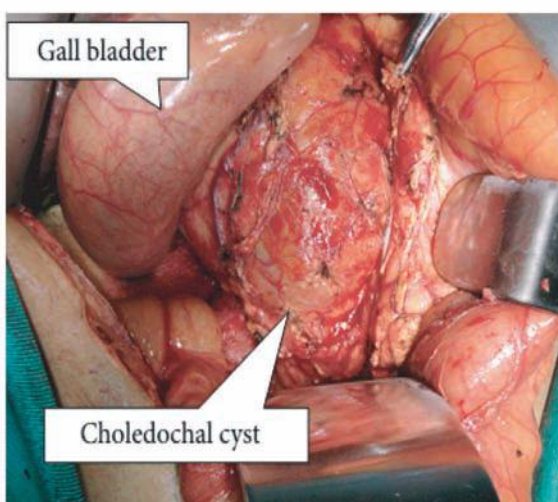
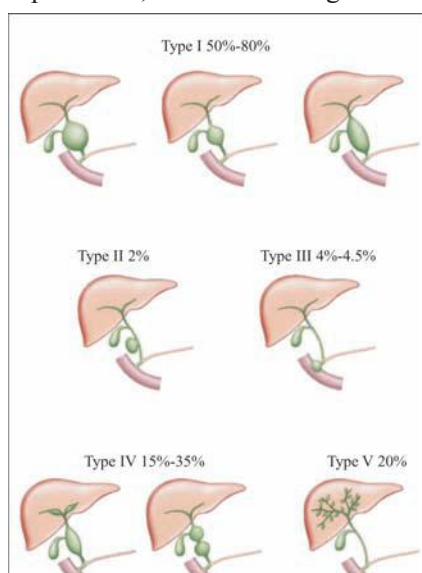
Ten patients were treated for choledochal cyst, of whom eight were women. The median age of presentation was 31 years (16–38 years) and two of them were males with the mean age of 36 years (26 to 48 years). The predominant symptom was abdominal pain occurring in all patients. Two patients presented with history of recurrent cholangitis and another two of them presented with abdominal mass. The symptoms and complications at presentation are summarized in table I. The imaging studies carried out for diagnosis included abdominal ultrasonography in all ten patients, abdominal Computed Tomography (CT) in 7, ERCP in 3, and MRCP in 5 patients (Fig 2a and 2b). The cyst classification by Todani classification revealed eight cases of type I cyst (Fusiform 5 and saccular 3) and one of each of type II and type III cysts (Choledochoceles). None of the patients had undergone preoperative drainage procedure. ERCP was carried out in 3 patients to define the APBDJ. The patient with choledochocoele in addition underwent sphincterotomy. In none of these patients, malignancy was detected. One of the patients had undergone previous cystoduodenostomy (10 years back elsewhere, at the age of 18) and had recurrent cholangitis. Surgical strategy in eight patients with type I choledochal cyst consisted of complete cyst resection, cholecystectomy, and bilioenteric anastomosis (Fig 3, 4, and 5). In one patient with saccular type I cyst, the cyst was opened to define the transaction line of the upper end of the cyst. The operative procedure details are summarized in table I. The mean operating time was 2 hours and 40 minutes (1 hour 40 minutes to 4 hours range). Type II lesion was excised without any reconstruction and type III patient underwent ERCP sphincterotomy. All the resected specimens showed chronic inflammation. However, in one specimen in addition, features of mild dysplasia were noted.

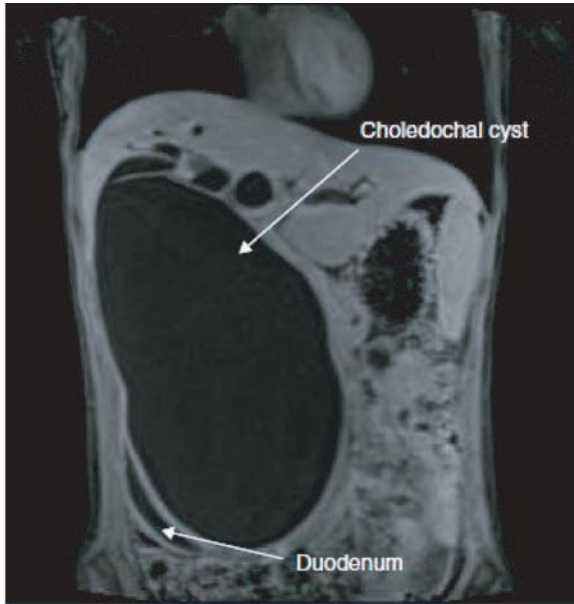
The early outcome was that one of the patients developed mild pancreatitis and anastomotic leak, which was managed conservatively with IV fluids, antibiotics, and ultrasound guided drainage. The median duration of hospital stay was 10 days (Range 3–32). Patients were followed up for a median duration of 6 years (Range 3 months to 12 years). There was no perioperative mortality. All patients were symptom-free, except for two patients who developed 2 episodes of recurrent cholangitis during their follow-up and were managed successfully with antibiotics. No anastomotic strictures or malignancy was noted in any of these patients during follow-up of one year. In addition, no patients developed nutritional abnormalities and they were in good health during the period of follow-up.

Table I : Demographic details, management and outcome of patients

Pt	Age/Sex	Symptoms/ Signs	Type of Cyst	Previous Surgery	Investigation	Procedure	Postopcom. Early	postopcom. Late	Hospital Stay (days)
1	38/F	Abd. pain, acute pancreatitis	Type III	Nil	US, CT, and ERCP	ERCP sphct	Nil		5
2	28/F	Abd. pain, Jn, and cholangitis	Type I-C	Choledocho- duodenostom	US/CT/MRCP	CC/CEx/HJ	Nil		10
3	32/F	Abd. pain, Jn	Type I-A	Nil	US/ERCP/MR	CPCC/CEx/HJ	Nil		10
4	16/F	Abd. pain, Jn, cholangitis, and abd. mass	Type I-A	Nil	US/ERCP/MRCP	CC/CEx/HJ	Mild pancreatitis/ anastomosis leak	Chol.	32
5	22/M	Abd. pain	Type II	Nil	US/MRCP	Excision	Nil		7
6	37/M	Abd. pain, Jn, and acute pancreatitis	Type I-A	Nil	US/CT	CC/CEx/HJ	Nil		7
7	38/M	Abd. pain, Jn, and abd. mass	Type I-C	Nil	US/CT	CC/CEx/HJ	Nil	Chol.	8
8	30/F	Abd. pain	Type I-A	Nil	US/CT	CC/CEx/HJ	Nil		8
9	25/F	Abd. pain, Jn	Type I-A	Nil	US/CT/MRCP	CC/CEx/HJ	Nil		7
10	25/F	Abd. pain, Jn	Type I-c	Nil	US/CT	CC/CEx/HJ	Nil		7

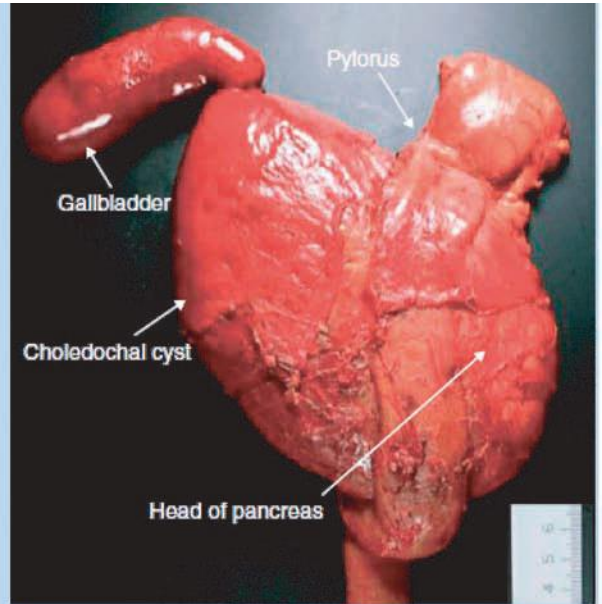
Pt = Patient number, Abd. = Abdominal, Jn = Jaundice, US = Ultrasound, MRCP = Magnetic Resonance Cholangiography, ERCP = Endoscopic Retrograde Cholangiopancreatography, CC = Cholecystectomy, CEx = Complete Excision of Cyst, HJ = Hepaticojejunostomy, Sphct = Sphincterotomy, Comp. = Complications, Chol. = Cholangitis.

**Fig 1 :** Todani's classification of choledochal cyst**Fig 2 :** Intraoperative view of large choledochal cyst dissected out in patient's no. 6



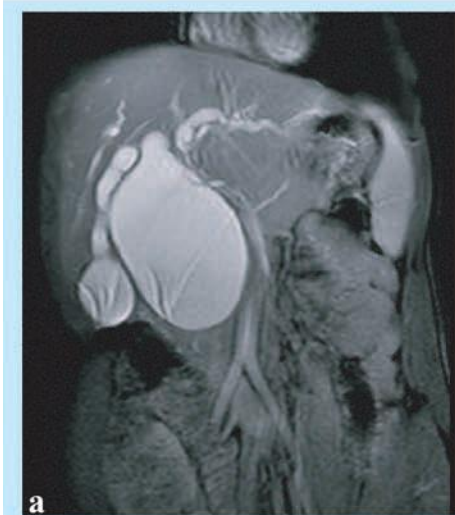
a

Fig 4a : CT scan findings of patient no. 8



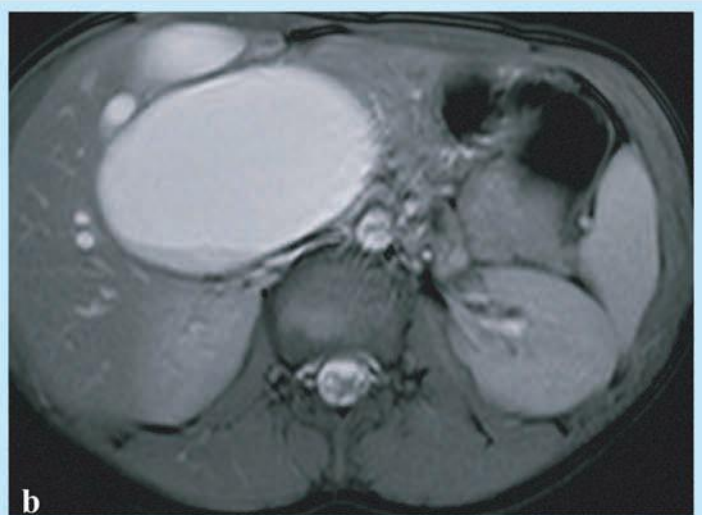
b

Fig 4b : Specimen of completely excised choledochal cyst with gallbladder with head of the pancreas of patient no. 8



a

Fig 5a : MRCP showing type I-A saccular choledochal cyst in patient no. 3 (Table I).



b

Fig 5b : MRCP (Cross section) showing large type I-A saccular choledochal cyst in patient no. 3 (Table I).

Discussion

Cystic disease of the bile duct or choledochal cyst was first described by Vater & Elzer in 1723. However, the first full clinical description was produced by Douglas, 1823 [10,11]. It is considered a low-incidence disease, affecting between 1:13,000 and 1:2,000 people worldwide and most common in Japan, where two thirds of the cases described in the literature occurred.

It can affect people of any age, but is more frequent in children below ten years of age and only 20% of cases occur in individuals aged over 20 years. The distribution by sex is 4:1 (Women: men). The main theory regarding the etiology of cystic disease of the bile duct concerns the reflux of pancreatic enzymes into the common bile duct, owing to an anomalous pancreatobiliary junction, i.e. when the join occurs more than 1.5 cm from

the hepatoduodenal ampulla. However, this theory does not explain the formation of type II and type V cysts, which may have a genetic component [12]. The classical triad (Jaundice, abdominal pain and a palpable mass in the upper right quadrant of the abdomen) is rare in adults, who account for 0-17% of cases. However, in around 85% of children and 25% of adults, at least two elements of the triad are present. Patients may also present initially with cholangitis, pancreatitis or biliary peritonitis through rupture of the cyst. It is important to note the presence of concomitant malignancy. The incidence of this is higher when diagnosis of cystic disease of the bile duct occurs at a more advanced age: 2% at 20 years and 43% at 60 years. Type I and type IV cysts are pose the greatest risk of malignant degeneration. Type V poses the least risk (7%) [13]. Ultrasound is the normal triage imaging examination [14]. Computerized tomography may diagnose a bile duct cyst, but is less accurate than magnetic cholangioresonance and is more useful subsequent to surgery to diagnose stenosis of the bilioenteric anastomosis. Retrograde endoscopic cholangiopancreatography is good for charting the anatomy of the bile duct and thus diagnosing cystic disorders. However, magnetic cholangioresonance provides images that are at least similar, without the risk of complications posed by retrograde endoscopic cholangiopancreatography [15]. Surgical treatment has changed over the years. Previously, the surgical procedure involved a cystoenterostomy. However, in view of postoperative complications, such as cholangitis, pancreatitis and hepatolithiasis (Re-operation rate of 70%) and the risk of malignant degeneration of the cyst this procedure is no longer used. At present, the standard treatment involves excision of the cyst and biodigestive derivation [16]. There is some controversy regarding treatment of type IV and type V cysts [17]. The excision of an extra-hepatic cyst is the rule, but there are doubts as to whether intra-hepatic cysts should be removed or not. The tendency is to carry out a hepatectomy if the cysts are localized. If they are dispersed, this procedure is not possible. Stenosis of the biliodigestive anastomosis is one of the most feared complications and its incidence varies according to the technique employed [18]. All the patients attended emergency services and presented with symptoms on admission to hospital.

All presented with abdominal pain and 60% were jaundiced. In all cases, the Todani classification was definitively diagnosed only during surgery. There was no pre-operative diagnosis because this is a rare disease and the hypothesis was not suggested by the imaging examinations. In all cases, an ultrasound exam was conducted and no report suggested cystic disease of the bile duct. Neither was this disease suggested in the patient who underwent retrograde endoscopic cholangiopancreatography and magnetic cholangioresonance. As three patients presented with obstructive jaundice, exploration of the bile duct was therefore recommended. One other patient underwent exploratory laparotomy as there was a hypothetical diagnosis of a mesenteric cyst or lymphangioma. The other patient presented with suppurative cholangitis with a liver abscess and underwent emergency surgery. In the two patients whose cyst was resected the reconstruction involved a Roux-en-Y hepaticojejunostomy [19,20]. The other patient underwent a hepaticogastrojejunostomy, i.e. joining the anastomosis of the jejunum to the stomach and the hepatic duct to the same flap, in order to clear the way for the endoscope to the intra-hepatic bile duct in the case of recurrence of intra-hepatic calculi. There was no malignant degeneration of the cyst in any of the three cases where histopathological analysis was conducted, nor of the postoperative segment. However, follow-up was of short duration (8-23 months). Early postoperative complications included infection of the surgical wound in two patients and one reoperation owing to intra-peritoneal buildup of bile. The patient with a liver abscess was operated on two more times and died forty days after the first operation of septic shock. None of the surviving patients developed postoperative pancreatitis, cholangitis or intrahepatic lithiasis.

Conclusion

Cystic disease of the bile duct is a rare disease in our setting, although it should be considered in the differential diagnosis of obstructive jaundice. Cyst resection with reconstruction of the bile duct is the standard treatment, with a low prevalence of complications and postoperative malignant degeneration.

Disclosure

All the authors declared no competing interest.

References

1. Kasai M, Asakura Y, Taira Y, "Surgical treatment of choledochal cyst". *Annals of Surgery*. 1970;172(5): 844–851.
2. Woon CYL, Tan YM, Oei CL, Chung AYP, Chow PKH, Ooi LLPJ, "Adult choledochal cysts: an audit of surgical management". *ANZ Journal of Surgery*. 2006;76(11):981–986.
3. Cho MJ, Hwang S, Lee YJ et al., "Surgical experience of 204 cases of adult choledochal cyst disease over 14 years," *World Journal of Surgery*. 2011;35(5):1094–1102.
4. Lenriot JP, Gigot JF, Segol P, Fagniez PL, Fingerhut A, Adloff M, "Bile duct cysts in adults: a multi-institutional retrospective study". *Annals of Surgery*. 1998; 228(2):159–166.
5. de Vries JS, de Vries S, Aronson DC et al., "Choledochal cysts: Age of presentation, symptoms, and late complications related to Todani's classification". *Journal of Pediatric Surgery*. 2002;37(11):1568–1573.
6. Atkinson HDE, Fischer CP, Jong CHC, Madhavan KK, Parks RW, Garden OJ. Choledochal cysts in adults and their complications. *HPB (Oxford)*. 2003; 5(2):105-110.
7. Balbbitt DP. Congenital choledochal cysts: new etiological concept based on anomalous relationships of the common bile duct and pancreatic bulb. *Ann Radiol*. 1969; 12(3):231-240.
8. Benjamin IS. Biliary cystic disease: the risk of cancer. *J Hepatobiliary Pancreat Surg*. 2003; 10:335-339.
9. Chijiwa K, Koga A. Surgical management and long-term follow-up of patients with choledochal cysts. *Am J Surg*. 1993; 165:238-242.
10. Douglas A. Case of dilation of the common bile duct. *Mon J Med*. 1852; 14:97-100.
11. Lipsett PA, Pitt HA. Surgical treatment of choledochal cysts. *J Hepatobiliary Pancreat Surg*. 2003; 10:352-359.
12. Liu DC, Rodriguez JA, Meric F, Geiger JL. Laparoscopic excision of a rare Type II choledochal cyst: Case report and review of the literature. *J Pediatr Surg*. 2000; 35(7):1117-1119.
13. Shimonishi T, Sasaki M, Nakanuma Y. Precancerous lesions of intrahepatic cholangiocarcinoma. *J Hepatobiliary Pancreat Surg*. 2000;7:542-550.
14. Singham J, Schaeffer D, Yoshida E, Scudamore C. Choledochal cysts: Analysis of disease pattern and optimal treatment in adult and pediatric patients. *HPB (Oxford)*. 2007; 9(5):383-387.
15. Todani T, Watanabe Y, Narusue M, et al. Congenital bile duct cysts: Classification, operative procedures and review of thirty seven cases including cancer arising from choledochal cyst. *Am J Surg*. 1977; 134:263- 269.
16. Visser BC, Suh I, Way LW, Kang Sang-MO. Congenital choledochal cysts in adults. *Arch Surg*. 2004; 139:855-862.
17. Yamaguchi M. Congenital choledochal cyst: Analysis of 1433 patients in the Japanese literature. *Am J Surg*. 1980; 140(5):653-657.
18. Yamaguchi M, Sakurai M, Takeushi S, Awazu S. Observation of cystic dilatation of the common bile duct by ultrasonography. *J Pediatr Surg*. 1980; 15(2):207-210.
19. Wiseman K, Buczkowski AK, Chung SW, Francoeur J, Schaeffer D, Scudamore CH. Epidemiology, presentation, diagnosis and outcomes of choledochal cysts in adults in an urban environment. *The American Journal of Surgery*. 2005; 189:527-531.
20. Yamashita H, Otani T, Shioiri T, Takayama T, Kakiuchi C, Todani T, Makuuchi M. Smallest Todani's type II choledochal cyst. *Digestive and Liver Disease*. 2003; 35:498-502.