Case Report

CHOLEDOCAL CYST– A CASE REPORT AND A DETAIL REVIEW OF LITERATURE INCLUDING PATHOGENESIS, COMPLICATION AND TREATMENT

Zahangir Boksh[1], Zahid Boksh[2] & Jamal Ahmed Chowdhury[2]

[1]Kulliyyah of Medicine and Health Sciences, INSANIAH University College, Kedah, Malaysia.

[2]Department of General Surgery, Jahurul Islam Medical College & Hospital, Dhaka, Bangladesh.

ABSTRACT

A case study from department of General Surgery, Jahurul Islam Medical College & Hospital, Dhaka, Bangladesh. A two year old boy presented with abdominal mass. Hematological, biochemical and ultrasonography investigation were performed. A cystic dilatation of about 10 cm noted attached to the liver and a provisional diagnosis of hepatic cyst was made. After laparoscopy a huge cyst was noted attached with the bile duct and a type 1 choledocal cyst was diagnosed. The cyst was removed with a segment of attached duct and a reconstruction surgery was performed. The cyst measured 10.5 cm in maximum dimension with about 0.2 cm of the thickness of the cyst wall. On opening the cyst contained yellowish green bile stained serous fluid. The duct measured 12.0 X 1.0 X1.0 cm. The patient had an unremarkable recovery and is well. A detailed review of literature including pathogenesis, complications and treatment were discussed.

Keyword: *choledocal*

Introduction

Choledocal cyst is a rare anomaly characterized by cystic dilatation of the common bile duct (CBD), and was first described by Vater and Ezler in 1723. Douglas subsequently published a patient with this anomaly in 1853 with a detailed description speculating about the congenital nature of this condition. Alonso-Lej et al in 1959 reviewed 94 cases with two cases of their own[1]. They classified choledochal cysts into 3 types. In 1977, Todani et al further classified this anomaly into 5 types based on the cholangiographic findings[2].

The estimated prevalence of choledochal cysts ranges from approximately 1 case in 13,000 people to 1 case in 2 million people. Again they are more common in Japan and Asia[3]. Yamaguchi et al. reviewed 1433 cases. Of these, 1204 cases were from Japan[4]. Choledochal cysts can occur in persons of any age however two thirds of the cysts are diagnosed before the age of 10 years[5]. 20% of cysts are diagnosed in much older patients[6]. Choledochal cysts have been detected at prenatal ultrasonography as early as 15 weeks' gestation.

Case report

A two year old boy presented with abdominal mass and discomfort, mild jaundice, loss of appetite and failure to thrive. He was admitted to the general surgery department of Jahurul Islam Medical College & Hospital, Dhaka, Bangladesh, with this clinical signs and symptoms. Routine, hematological, biochemical and ultrasonography investigation were performed. A cystic dilatation of about 10 cm noted attached to the liver and a provisional diagnosis of hepatic cyst was made. After laparoscopy a huge cyst was noted attached with the bile duct and a type 1 choledocal cyst was diagnosed. The cyst was removed with a segment of attached duct and a reconstruction surgery was performed. The cyst measured 10.5 cm in maximum dimension with about 0.2 cm of the thickness of the cyst wall. On opening the cyst contained yellowish green bile stained serous fluid. The duct measured 12.0 X 1.0 X1.0 cm. The patient had an unremarkable recovery and is well.

Pathogenesis

The exact cause of choledochal cyst remains unknown. Many authors believe that they are congenital because most of the cysts are diagnosed in infants and children. Since approximately 20% are diagnosed in adults, including elderly patients, several theories have been postulated[7.]

- Weakness of the wall of the bile duct[8]
- Obstruction of the distal choledochus[9]
- Combination of obstruction and weakness[10]
- Reflux of pancreatic enzymes into the CBD secondary to an anomaly of the pancreaticobiliary junction[8]

Figure 1: Gross picture of choledocal cyst

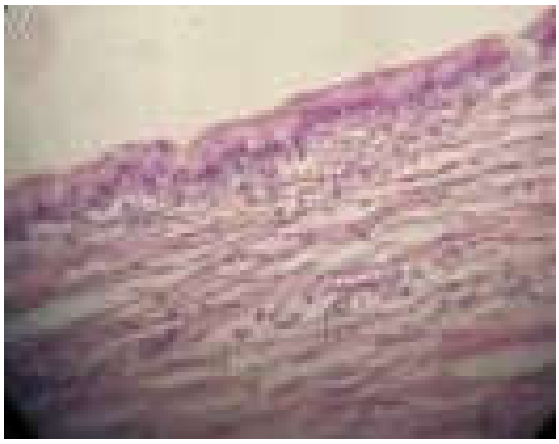

Figure 2: Cyst wall showing columnar epithelial lining

(Microscopy H&E high power)

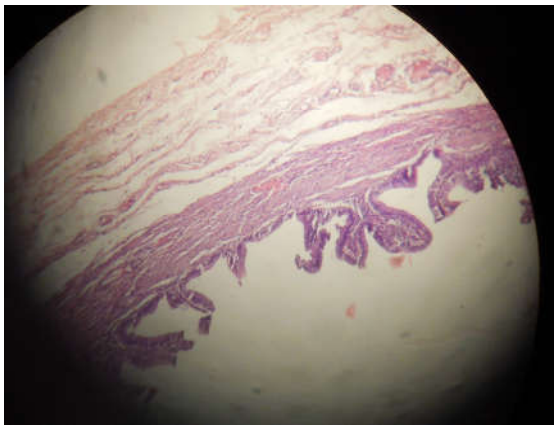

Figure 3: Ductal epithelium showing tufting and hyperplasia (Microscopy H&E Low power)

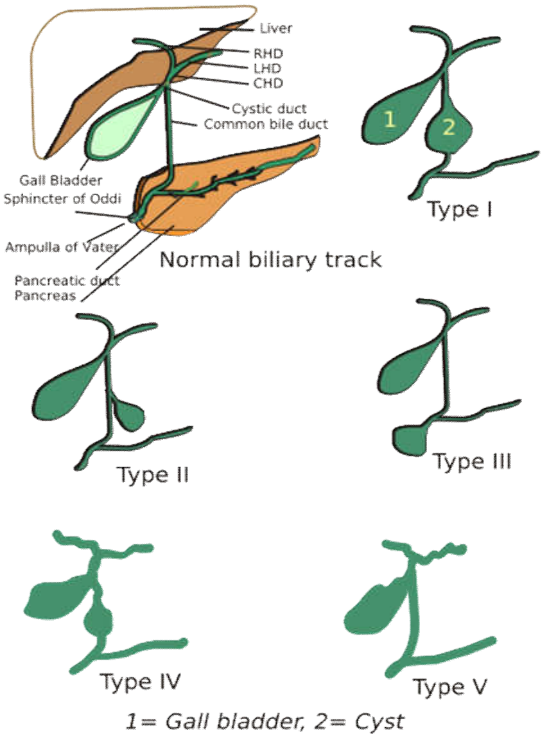

Figure 4: Pancreaticobiliary ductal system theory of choledochal cyst etiology.

In all reported series, five types of choledocal cysts have been described with the most frequent type of choledochal cyst is type I. In types I and IV the choledochal cysts show fusiform dilation of the CBD without or with dilated intrahepatic ducts, respectively, the female-to-male ratio is about 4:1. The types II show diverticulum formation of the CBD, and the type III shows choledochocele, and type V shows dilation of intrahepatic ducts only, where this cysts occur with equal frequency in both sexes.

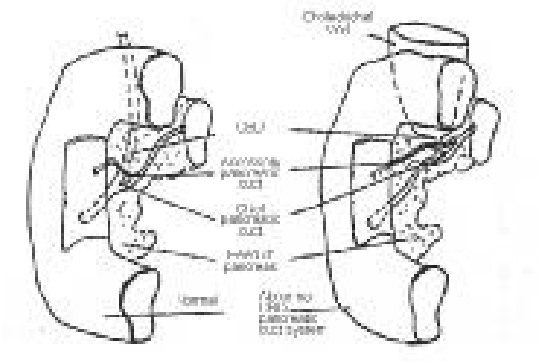

Figure 5

Pathogenesis of choledocalcyst is an enigma. In 1969, Babbitt and colleagues carefully analyzed the cholangiograms of patients with choledochal cysts and found many with an anomaly of the pancreaticobiliary junction[11]. In these patients, a small distal CBD entered the pancreatic duct at 2-3.5 cm from the ampulla of Vater, whereas the normal common channel is 5 mm or less. This may represent failure of normal separation of these two ducts during embryologic development. This proximal junction precludes the proper functioning of the sphincter of Oddi. The pressure in the pancreatic duct (30-50 cm H_2 O) exceeds the pressure in the CBD (25-30 cm H_2 O) favoring reflux of pancreatic secretions into the CBD. They also noted high amylase content in the fluid from the cysts. The reflux of pancreatic juice could lead to weakness and dissolution of the wall of the CBD (Figure 5).

Experimental support for this concept was reported in 1974 by Kato and associates who anastomosed the main pancreatic duct to the gallbladder in dogs[10]. Within 9 days after the anastomosis, all the tested animals had varying degrees of dilatation of the CBD, with edematous changes of the CBD wall. They concluded that proteolytic enzymes were responsible for the damage.

In 1977, Spitz supported the concept of distal CBD obstruction as a cause of choledochal cyst when he demonstrated bile duct dilatation in lambs by means of ligation of the duct near the duodenum[9]. Miyano et al (1981) established an experimental model of an anomalous choledochopancreatic ductal junction by creating a choledochopancreatic end-to-side ductal anastomosis in puppies[12]. They successfully reproduced the dilatation of the CBD in all experimental animals.

In 1984, Todani et al conducted an analysis of endoscopic retrograde cholangiopancreatograms and other cholangiograms and confirmed this long common-channel anomaly[1,3]. The anomaly was found in most patients. Other authors have reported the same findings in their series[14, 22]. Reflux of pancreatic enzymes into the CBD resulting in damage to the ductal wall and the distal portion of the CBD is the most at risk, and with repeated irritation, it can be stenotic.

All of these theories are applicable to choledochal cyst type I, III, and IV anomalies, but they cannot be used to explain type II and V choledochal cysts in which the CBD is normal. Perhaps genetic factors play a role[3,14,15].

Despite this, the two most accepted theories are still reflux of pancreatic enzymes into the CBD secondary to an anomalous pancreaticobiliary junction and obstruction of the distal CBD. Grossly, the size of a type I choledochal cyst widely varies[3,5,14]. The cyst contains a few hundred milliliters of bilious fluid that is rich in pancreatic enzymes. The cyst wall thickness also varies, ranging from very thin to a few millimeters in thickness.

In type IV and V anomalies, intrahepatic cysts can be fusiform or saccular and are continuous with the CBD. Dilatation of the proximal common duct is frequent, particularly just proximal to the long common channel. Sludge and stones are sometimes present within the cyst[3,14]. The bileduct distal to the cyst is usually stenotic. Metaplasia results secondary to repeated destruction and regrowth of the lining of the cyst. Inflammation has been noted to be more severe in intrahepatic cysts. The inflammation is significantly less in younger patients[4,16,17,18].

Carcinoma arising in a choledochal cyst wall or remaining biliary tree after complete cyst excision is well recognized[19]. Malignancy is believed to be the result of chronic inflammation and metaplasia. The typical malignancy is adenosquamous carcinoma or occasional results in small cell carcinoma.

Malignancies in choledochal cyst can arise from the distal CBD or from the intrahepatic bile ducts. The risk of cancer appears to be related to the age of the patient; it is high about 20 times, compared with that of the healthy population [2,20,21,22]. The risk of detecting a biliary tree malignancy in a resected cyst is 0.7% in patients who undergo surgery before age 10 years, 6.8% in patients who undergo surgery at age 11-20 years, and 14.3% in patients who undergo surgery after age 20 years.

More than half of the cancers arise from the cyst wall, even after successful internal drainage. Total cyst excision has not prevented the risk of malignancy in the remaining bile ducts. Malignancy can develop many years after excision of the cyst and can develop in areas of the biliary tree remote from the cyst such as the gallbladder and terminal common duct, which is left behind after excisional surgery.

Any type of cyst is susceptible to malignancy, but the greatest prevalence is observed with types I, IV, and V. Factors thought to contribute to the development of malignancy include prolonged bile stasis and chronic inflammation of the cyst wall. Inflammatory and metaplastic changes increase with patient age, and they are frequently observed in association with carcinoma of the bile duct. The increased risk of biliary tract malignancy, even after surgery, warrants close surveillance in any case

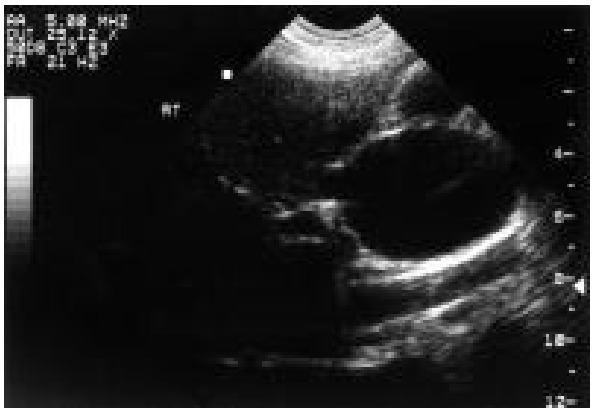

Figure 6: Ultrasonogram shows a large unilocular cyst under the liver.

Endoscopic retrograde cholangiopancreatography (ERCP) remains the standard diagnostic study. In expert hands, ERCP can be performed with a high rate of success, even in small infants. When successful, ERCP clearly shows the anatomy of the pancreaticobiliary junction[2,5]. Magnetic resonance cholangiopancreatography (MRCP) has largely supplanted ERCP as the diagnostic test of choice for choledochal cysts because it offers high resolution detailed images of relevant anatomy, is noninvasive, and does not suffer from complications such as post procedure pancreatitis (Figure 7)[28].

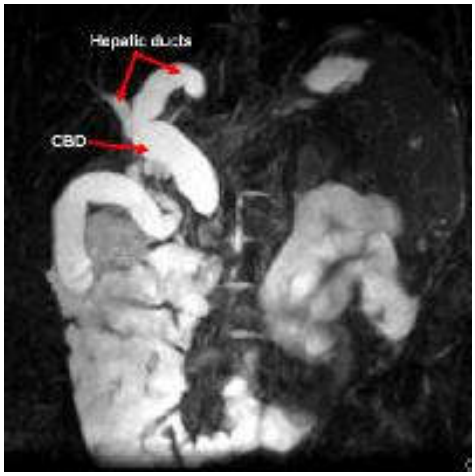

Figure 7: Magnetic resonance cholangiopancreatography shows dilated hepatic ducts and common bile duct (CBD) of a type IV cyst.

MRCP detects most choledochal cysts with sensitivities from 90-100% and specificities from 73-100%, with the exception of small choledochoceles and minor ductal anomalies. MRCP has been shown to be effective in neonates[29], children[30], adults[31] and fetuses[32]. CT scanning may also be useful to delineate the cyst and its relationship to surrounding structures. In older patients, especially adults, CT scanning combined with cholangiography may be useful (Figure 8).

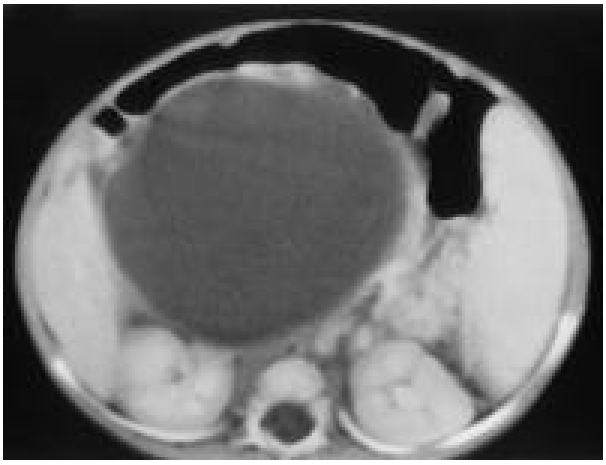

Figure 8: CT scan shows a large cyst with wall thickening.

Treatment of choledochal cysts is surgical, except in type V multiple intrahepatic cysts, which can benefit from medical management for variable periods of time. Internal drainage, either with cystoduodenostomy or cystojejunostomy with Roux-en-Y biliary reconstruction, was used in the past but it resulted in a high incidence of calculi, recurrent cholangitis, anastomotic strictures, and carcinoma arising from the cyst. Of patients treated with either cystoduodenostomy or cystojejunostomy, 65% remained symptomatic, and 40% required repeat surgery at a later date [23,24].Recurrent cholangitis and chronic inflammation in the remaining cyst eventually produces metaplasia that leads to malignant transformation.

Management

Total excision of the cyst in types I, II, and IV followed by reconstruction of the biliary tree with hepaticojejunostomy in a Roux-en-Y fashion has been widely accepted and has been found to be superior to hepaticoduodenostomy[25].This procedure constitutes excision of the distal common bile duct which subsequently blocks the reflux of pancreatic enzymes into the biliary tract, thereby decreasing the incidence of carcinoma of the bile duct.

With type III choledochal cysts, the general approach is one of lateral duodenotomy with unroofing of the choledochocele to drain the bile duct and pancreatic duct directly into the duodenum. The two ductal openings should be carefully examined to determine whether ductoplasty is required[26,27]. With regard to type V choledochal cysts, patients with localized disease may benefit from a hepatic lobectomy. If the disease is diffuse, involving both lobes of the liver, treatment is palliative and liver transplantation may be required[13,26].

Total excision of the cyst is possible in virtually all infants and young children except in older patients with repeated cholangitis and marked pericystic inflammation which is best managed with resection of the anterolateral part of the cyst followed by an endocystic resection of the lining, leaving the back wall adjacent to the portal vein in place, as reported by Lilly in 1977[33]. This technique makes the dissection less hazardous.

Several groups have successfully performed laparoscopic -assisted and laparoscopic total cyst excision with Roux-en-Y hepatoenterostomy with complication rates comparable to those of the open procedure. Li and colleagues (2004) performed laparoscopic cyst excision with laparoscopic-assisted Roux-en-Y hepatoenterostomy in 35 children (33 cyst type, 2 fusiform) without conversion to the open procedure and with postoperative stays of 3-5 days[34]. The procedure is described in detail for both choledochal cyst and biliary atresia surgery by Martinez-Ferro et al (2005)[35] Liuming et al[36] and Liem et al[37] also concluded that laparoscopic excision was as safe as open excision.

Finally, although we reported one case of choledocal cyst and the disease is uncommon as well, the authors believe, that the disease should be taken seriously since the resection depends on the various types of the choledocal cyst. A careful and early detection and classification should be made. A complete resection is indicated to prevent any malignant transformation or recurrence.

References

1. Alonso-Lej F, Rever WB Jr, Pessagno DJ. Congenital choledochal cyst, with a report of 2, and an analysis of 94, cases. *Int Abstr Surg*. Jan 1959;108(1):1-30. [Medline].
2. 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*. Aug 1977;134(2):263-9. [Medline].
3. Flanigan PD. Biliary cysts. *Ann Surg*. Nov 1975;182 (5):635-43. [Medline].
4. Yamaguchi M. Congenital choledochal cyst. Analysis of 1,433 patients in the Japanese literature. *Am J Surg*. Nov 1980;140(5):653-7. [Medline].
5. Katyal D, Lees GM. Choledochal cysts: a retrospective review of 28 patients and a review of the literature. *Can J Surg*. Dec 1992;35(6):584-8. [Medline].
6. Lipsett PA, Pitt HA, Colombani PM, et al. Choledochal cyst disease. A changing pattern of presentation. *Ann Surg*. Nov 1994;220(5):644-52. [Medline].
7. Saing H, Tam PK, Lee JM, Pe-Nyun. Surgical management of choledochal cysts: a review of 60 cases. *J Pediatr Surg*. Aug 1985;20(4):443-8. [Medline].
8. Babbitt DP, Starshak RJ, Clemett AR. Choledochal cyst: a concept of etiology. *Am J Roentgenol Radium Ther Nucl Med*. Sep 1973;119(1):57-62. [Medline].
9. Spitz L. Experimental production of cystic dilatation of the common bile duct in neonatal lambs. *J Pediatr Surg*. Feb 1977;12(1):39-42. [Medline].
10. Kato T, Asakura Y, Kasai M. An attempt to produce choledochal cyst in puppies. *J Pediatr Surg*. Aug 1974;9 (4):509-13. [Medline].
11. Kato T, Hebiguchi T, Matsuda K, Yoshino H. Action of pancreatic juice on the bile duct: pathogenesis of congenital choledochal cyst. *J Pediatr Surg*. Apr 1981;16 (2):146-51. [Medline].
12. Miyano T, Suruga K, Suda K. Choledocho-pancreatic end to side anastomosis in dog as an experimental model of choledocho-pancreatic long common channel disorders. *Jpn J Pediatr Surg*. 1981;13:525-31.
13. Todani T, Watanabe Y, Fujii T, et al. Congenital choledochal cyst with intrahepatic involvement. *Arch Surg*. Sep 1984;119(9):1038-43. [Medline].
14. Oguchi Y, Okada A, Nakamura T, et al. Histopathologic studies of congenital dilatation of the bile duct as related to an anomalous junction of the pancreaticobiliary ductal system: clinical and experimental studies. *Surgery*. Feb 1988;103(2):168-73. [Medline].
15. Miyano T, Yamataka A, Kato Y, et al. Hepaticoenterostomy after excision of choledochal cyst in children: a 30-year experience with 180 cases. *J Pediatr Surg*. Oct 1996;31(10):1417-21. [Medline].
16. Yamaguchi M, Sakurai M, Takeuchi S, Awazu S. Observation of cystic dilatation of the common bile duct by ultrasonography. *J Pediatr Surg*. Apr 1980;15 (2):207-10. [Medline].
17. Todani T, Watanabe Y, Urushihara N, et al. Biliary complications after excisional procedure for choledochal cyst. *J Pediatr Surg*. Mar 1995;30(3):478 -81. [Medline].
18. Suita S, Shono K, Kinugasa Y, et al. Influence of age on the presentation and outcome of choledochal cyst. *J Pediatr Surg*. Dec 1999;34(12):1765-8. [Medline].
19. Kamisawa T, Okamoto A, Tsuruta K, Tu Y, Egawa N. Carcinoma arising in congenital choledochal cysts. *Hepatogastroenterology*. Mar-Apr 2008;55(82-83):329-32. [Medline].
20. Imazu M, Iwai N, Tokiwa K, et al. Factors of biliary carcinogenesis in choledochal cysts. *Eur J Pediatr Surg*. Feb 2001;11(1):24-7. [Medline].
21. Iwai N, Deguchi E, Yanagihara J, et al. Cancer arising in a choledochal cyst in a 12-year-old girl. *J Pediatr Surg*. Dec 1990;25(12):1261-3. [Medline].
22. Todani T, Tabuchi K, Watanabe Y, Kobayashi T. Carcinoma arising in the wall of congenital bile duct cysts. *Cancer*. Sep 1979;44(3):1134-41. [Medline].
23. Chijiiwa K, Koga A. Surgical management and long-term follow-up of patients with choledochal cysts. *Am J Surg*. Feb 1993;165(2):238-42. [Medline].
24. Chijiiwa K, Tanaka M. Late complications after excisional operation in patients with choledochal cyst. *J Am Coll Surg*. Aug 1994;179(2):139-44. [Medline].
25. Shimotakahara A, Yamataka A, Yanai T, et al. Roux-en-Y hepaticojejunostomy or hepaticoduodenostomy for biliary reconstruction during the surgical treatment of choledochal cyst: which is better?. *Pediatr Surg Int*. Jan 2005;21(1):5-7. [Medline].
26. Joseph VT. Surgical techniques and long-term results in the treatment of choledochal cyst. *J Pediatr Surg*. Jul 1990;25(7):782-7. [Medline].
27. Maning P, Polley T, Oldam K. Choledochocele: an unusual form of choledochal cyst. *Pediatr Surg Int*. 1990;5:22-6.
28. Kim SH, Lim JH, Yoon HK, et al. Choledochal cyst: comparison of MR and conventional cholangiography. *Clin Radiol*. May 2000;55(5):378-83. [Medline].
29. Lin JTH YH, Chen YH, Ni HS, et al. Magnetic resonance cholangiopancreatography diagnosed pancreatitis associated choledochal cyst: report of one case. *Acta Paediatr Taiwan*. Nov-Dec 2001;42(6):363 -6. [Medline].
30. Yamataka A, Kuwatsuru R, Shima H, et al. Initial experience with non-breath-hold magnetic resonance cholangiopancreatography: a new noninvasive technique for the diagnosis of choledochal cyst in children. *J Pediatr Surg*. Nov 1997;32(11):1560-2. [Medline].

31. Zhong L, Yao QY, Li L, Xu JR. Imaging diagnosis of pancreato-biliary diseases: a control study. *World J Gastroenterol*. Dec 2003;9(12):2824-7. [Medline].
32. Wong AM, Cheung YC, Liu YH, et al. Prenatal diagnosis of choledochal cyst using magnetic resonance imaging: a case report. *World J Gastroenterol*. Aug 28 2005;11(32):5082-3. [Medline].
33. Lilly JR. Total excision of choledochal cyst. *SurgGynecol Obstet*. Feb 1978;146(2):254-6. [Medline].
34. Li L, Feng W, Jing-Bo F, et al. Laparoscopic-assisted total cyst excision of choledochal cyst and Roux-en-Y hepatoenterostomy. *J Pediatr Surg*. Nov 2004;39 (11):1663-6. [Medline].
35. Martinez-Ferro M, Esteves E, Laje P. Laparoscopic treatment of biliary atresia and choledochal cyst. *Semin Pediatr Surg*. Nov 2005;14(4):206-15. [Medline].
36. Liuming H, Hongwu Z, Gang L, Jun J, Wenying H, Wong KK, et al. The effect of laparoscopic excision vs open excision in children with choledochal cyst: a midterm follow-up study. *J Pediatr Surg*. Apr 2011;46 (4):662-5. [Medline].
37. Liem NT, Pham HD, Vu HM. Is the laparoscopic operation as safe as open operation for choledochal cyst in children?. *J Laparoendosc Adv Surg Tech A*. May 2011;21(4):367-70. [Medline].

Corresponding author: Prof. Dr. Zahangir Boksh
Email address: boksh_zahangir@yahoo.ca

Received: August 2015
Accepted for publication: October 2015