# Surgical Management of Type IA Choledochal Cyst: A Case Report

Hang-Lung Chang[1], Jenny Nai-Wan Chang[2,3]

*Choledochal cyst is a rare congenital abnormality of the bile duct and usually occurs in young ages. We presented a 19 year-old girl who suffered from abdominal pain, fever and jaundice, and was subsequently diagnosed with Todani type IA choledochal cyst. Surgery with choledochal resection, mucosectomy of distal common bile duct, cholecystectomy, and Roux-en-Y hepaticojejunostomy was performed. She enjoyed full recovery after the operation. Early diagnosis of choledochal cyst is important due to its potential for malignant transformation, and thus surgical intervention for total cystic excision is recommended.*

***Key words: choledochal cyst, recurrent cholangitis, cystic dilatation of common bile duct***

## INTRODUCTION

Choledochal cyst has been recognized as a congenital anomaly of the bile duct; it encompasses cystic dilations at various parts of the biliary tree[(1)]. It could be classified as five types according to its anatomic and morphology, referred to as Todani classification[(2)]. An estimated 80% of patients with choledochal cysts were diagnosed by age 10[(3)]. The clinical presentations are varied, including asymptomatic, fever, abdominal pain, palpable mass and/or jaundice. Of note, it could be associated with cholelithiasis, cholangitis, cholecystitis, pancreatitis, hepatic abscesses as well as cholangiocarcinoma, particularly in adult patients[(4,5)]. Herein, we reported a young girl who had choledochal cysts type 1A presented with abdomen pain, fever and jaundice. She received surgical intervention and recovered after surgery.

## CASE REPORT

A 19 year-old girl was admitted due to a 3-day history of persistent dull abdominal pain. She was also noted as having tea-coloured urine and grayish white stool during her admittion. On physical examination, she was conscious with her vital signs showing temperature of 38.3°C, pulse rate of 77 beats per minute, respiratory rate of 18 breaths per minute, and blood pressure of 109/66 mm Hg. Her sclera and skin were icterics. Tenderness over right upper quadrant abdomen without Murphy' s sign or peritonitis was noted. Laboratory tests disclosed aspartate aminotransferase (SGOT) 665 U/L, alanine aminotransferase (SGPT) 999 U/L, alkaline phosphatase (alk-p) 467 U/L, and bilirubin, total type, 1.8 mg/dl, and direct, 1.3 mg/dl. ultrasonography of abdomen showed a spherical hypoechogenic cystic lesion in the hepatic hilum. A CT scan

Received: November 30, 2009 Accepted for publication: December 31, 2009
From the [1]Department of Surgery, [2]Department of Family Medicine, Landseed Hospital, Taiwan
[3]Department of Family Medicine, Landseed Hospital, Shanghai
Address reprint requests and correspondence: Dr. Hang-Lung Chang
Department of Surgery, Landseed Hospital, Taoyuan, Taiwan
77 Kuangtai Road, Pingjen City, Taoyuan County 32449, Taiwan (R.O.C.)
TEL: (03)4941234 ext 2036 FAX: (03)4021057
E-mail: changhl@landseed.com.tw

of abdomen showed fusiform dilatation of common bile duct (CBD) with a compressed gallbladder (Figure 1). Furthermore, magnetic resonance cholangiopancreatography (MRCP) was performed and showed a saccular enlargement of common bile duct of about 8 cm in diameter with elongation of gallbladder and dilation of bilateral proximal intrahepatic bile ducts (Figure 2). The pancreatic duct showed no abnormalities. Therefore, Todani type I choledochal cyst was impressed.

She was scheduled for surgery. During the operation, cystic dilatation of common bile duct and swollen gallbladder without definite stones were noted. Todani type IA choledochal cyst with cholecystitis was confirmed by surgical findings. Total choledochal resection, mucosectomy of distal CBD (Figure 3), cholecystectomy, and Roux-en-Y with end-to-side hepaticojejunostomy were performed. The gross pathology of choledochal cyst was shown in Figure 4. Microscopically, the bile ducts showed choledochocyst characterized with mostly loss of epithelium and chronic inflammatory cell infiltration, and the gallbladder revealed chronic cholecystitis with Rokitansky-Aschoff sinuses. There were no signs of malignancy. The patient recovered well after the operation, and the following SGOT, SGPT, alk-p, and bilirubin levels declined to normal ranges. She was discharged on day 7 following the operation. The patient was followed up at outpatient clinic for 6 months and appeared well.

## DISCUSSION

Five types of choledochal cysts were classified by Todani and colleagues[2]. Among these five types, the type I is the most commonly seen clinically. It accounts for 80% to 90% of all patients with choledochal cysts, and could be further divided into cystic dilation (type 1A), segmental dilation (type 1B) of CBD, and fusiform dilation of both common hepatic duct and CBD (type 1C)[6]. Type II is a form of diverticulum derived from CBD. Type III is a choledochocele, which is found in the intraduodenal portion of the CBD. Being the second most commonly seen in clinical, Type IV accounts for 10% to 15%[3,6]. It could be divided into multiple intra- and extra-hepatic

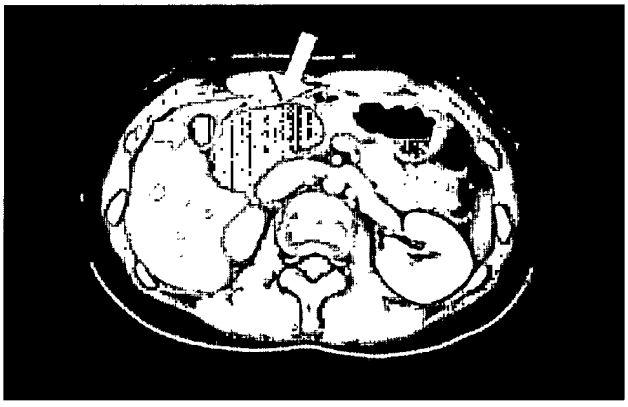

Figure 1 Abdominal CT scan showed dilated common bile duct (long arrow) and a compressed gallbladder (short arrow)

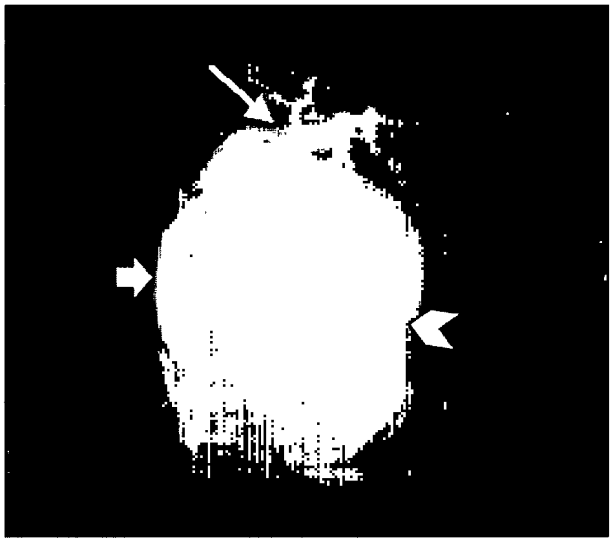

Figure 2 T2–weighted MRCP showed fusiform dilated common bile duct (arrow), dilated common hepatic duct (long arrow), and compressed gallbladder (short arrow)


Figure 3 Mucosectomy was performed to the distal part of choledochal cyst (arrow), which adjacent to pancreas (P)

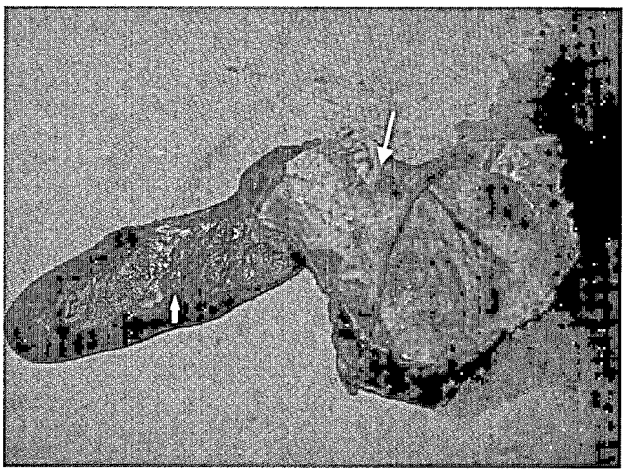

Figure 4 The gross pathology of gallbladder (arrow) and choledochal cyst of common bile duct (long arrow)

biliary tract cystic dilations (type IVA) and multiple extrahepatic biliary tract dilation (type IVB). Type V, also called Caroli's disease, presents as single or multiple diffuse cysts of the intrahepatic biliary tract. However, mixed types of choledochal cyst could occur[7]. The incidence of choledochal cysts was found to be predominant three to fourfold in females[1]. In addition, it seems to be a geographic characteristic. The incident rate in Asia has been found higher than in the United States and other western countries (1: 100,000 versus 1: 150,000)[3]. Of note, more than 33% of all cases were reported from Japan[8].

As our patient's presentation, the most common clinical feature of patients with choledochal cysts was abdominal pain, some of them were accompanied with jaundice, fever, and/or palpable abdominal mass, however, some of them were asymptomatic[4,6]. In addition, it should be considered associated with cystolithiasis, pancreatitis, cholangitis, cholecystitis, hepatic abscess, liver cirrhosis, portal hypertension, and cholangiocarcinoma when the diagnosis of choledochal cyst is made in adults. The exact etiology of choledochal cysts is still unclear. A recent concept has been widely discussed that focused on association between the anomalous union of the pancreaticobile duct and the formation of choledochal cysts[1,9]. The anomalous union of the CBD and the pancreatic duct with a sphincter dysfunction allows refluxing of pancreatic and bile juices, and this will result in inflammation, dilation and scarring of the biliary ducts[1,4]. This finding is similar to the microscopic findings of our patient. Nevertheless, tissues under long-term inflammation will have a high risk for malignant transformation.

Ultrasonography and CT scan could provide valuable information helping in preliminary diagnosis. However, MRCP and endoscopic retrograde cholangiopancreatography (ERCP) further provide a clearly visualized hepatobiliary tract structures including gallbladder, biliary tree, CBD and main pancreatic duct[17,18]. This could help to make a definite diagnosis and to estimate the involved field of choledochal cyst as well as to exclude the possibility of occult lesions, such as tumors[8-10]. Regarding the treatment, internal drainage with biliary bypass for patients with choledochal cyst alone had been utilized in the past. However, among them, 30% to 50% patients

subsequently developed late complications such as recurrent cholangitis, biliary cirrhosis and cholangiocarcinoma[3,11,12]. Therefore, surgery with complete excision of the cyst and reconstruction by hepaticojejunostomy has been recommended in recent years[13], particularly in patients with Todani type I and IV choledochal cysts, which have a higher rate of up to 28% for malignant changes.[2,14] In conclusion, even though choledochal cyst is relative rare, clinician should be alerted when a young patient presents with abdomen pain, especially accompanied with fever and jaundice. Early diagnosis is critical, and surgical intervention should be taken into consideration, because it may complicate further into relevant hepatobiliary diseases, including malignancy.

## REFERENCES

1. Singham J, Yoshida EM, Scudamore CH. Choledochal cysts: part 1 of 3: classification and pathogenesis. Can J Surg 2009;52:434-40.
2. Todani T, Watanabe Y, Narusue M, Tabuchi K, Okajima K. Congenital bile duct cysts: Classification, operative procedures, and review of thirty-seven cases including cancer arising from choledochal cyst. Am J Surg 1977;134:263-9.
3. Singham J, Schaeffer D, Yoshida E, Scudamore C. Choledochal cysts: analysis of disease pattern and optimal treatment in adult and paediatric patients. HPB (Oxford) 2007;9:383-7.
4. Banerjee Jesudason SR, Ranjan JM, Paul MR, Vyas FL, Govil S, Muthusami JC. Management of adult choledochal cysts- a 15-year experience. HPB (Oxford) 2006;8:299-305.
5. Tan SS, Tan NC, Ibrahim S, Tay KH. Management of adult choledochal cyst. Singapore Med J 2007;48:524-7.
6. Uribarrena AR, Raventos N, Fuentes J, Elias J, Tejedo V, Uribarrena ER. Diagnosis and management of choledochal cysts. A review of 10 new cases. Rev Esp Enferm Dig 2008;100:71-5.
7. Agarwal N, Kumar S, Hai A, Parfitt R. Mixed type I and II choledochal cyst in an adult. Hepatobiliary Pancreat Dis Int 2009;8:434-6.
8. Miyano T, Yamataka A. Choledochal cysts. Curr Opin Pediatr 1997;9:283-8.
9. Irie H, Honda H, Jimi M, et al. Value of MR cholangiopancreatography in evaluating choledochal cysts. AJR Am J Roentgenol 1998;171:1381-5.
10. Lee HK, Park SJ, Yi BH, Lee AL, Moon JH, Chang YW. Imaging features of adult choledochal cysts: a pictorial review. Korean J Radiol 2009;10:71-80.
11. Iwai N, Yanagihara J, Tokiwa K, Shimotake T, Nakamura K. Congenital choledochal dilatation with emphasis on pathophysiology of the biliary tract. Ann Surg 1992;215:27-30.
12. Edil BH, Olino K, Cameron JL. The current management of choledochal cysts. Adv Surg 2009;43:221-32.
13. Lipsett PA, Pitt HA. Surgical treatment of choledochal cysts. J Hepatobiliary Pancreat Surg 2003;10:352-9.
14. Kobayashi S, Asano T, Yamasaki M, Kenmochi T, Nakagohri T, Ochiai T. Risk of bile duct carcinogenesis after excision of extrahepatic bile ducts in pancreaticobiliary maljunction. Surgery 1999;126:939-44.

# 第一型膽管囊腫：病例報告

張漢隆[1] 張乃文[2,3]

膽管囊腫是一種罕見的先天性異常膽管疾病，常發生在年幼時期。本病例報告一名19歲女孩因腹痛、發燒和黃疸而至急診就診。檢查後診斷為第一型膽管囊腫。經膽管囊腫、膽總管遠端粘膜、膽囊切除及肝管空腸 Roux - en - Y吻合手術後，恢復良好。膽管囊腫具有潛在惡性轉化的可能性，因此早期診斷很重要，並建議及早以手術切除。

關鍵詞：膽管囊腫，復發性膽管炎，總膽管囊性擴張

收件：98年11月30日 接受刊載：98年12月31日
壢新醫院[1]外科 [2]家庭醫學科 [3]上海辰新醫院家庭醫學科
通訊及抽印本索取：張漢隆醫師 32449桃園縣平鎮市廣泰路77號 壢新醫院外科
電話：(03)4941234轉2036 傳真：(03)4021057
E-mail: changhl@landseed.com.tw