The Korean Journal of Internal Medicine
Vol. 14, No. 2, July, 1999

Choledochal Cyst Associated the with Anomalous Union of Pancreaticobiliary Duct (AUPBD) Has a More Grave Clinical Course Than Choledochal Cyst Alone

Hye-Kyoung Song, M.D., Myung-Hwan Kim, M.D.
Seung-Jae Myung, M.D., Sung-Koo Lee, M.D., Hong-Ja Kim, M.D.
Kyo-Sang Yoo, M.D., Dong-Wan Seo, M.D.
Hyun-Joo Lee, M.D., Byeong-Cheol Lim, M.D., Young-Il Min, M.D.

Department of Internal Medicine, Asan Medical Center, University of Ulsan College of Medicine, Seoul, Korea

Objective : *Since choledochal cyst is frequently associated with the anomalous union of pancreaticobiliary duct(AUPBD), AUPBD has been regarded to be the etiologic factor of choledochal cyst. However, the clinical significance of AUPBD an patients with choledochal cyst has not been clearly defined. Therefore, to clarify the significance of AUPBD in choledochal cyst patients, we compared the clinical features of patients with choledochal cyst according to the presence or absence of AUPBD.*

Methods : *Among 52 cases which were diagnosed as choledochal cyst out of 5,037 ERCP referrals between August 1990 and December 1996, we selected 44 cases, in which the pancreaticobillary junction was clearly visualized on cholangio-pancreaticography. These cases were divided into AUPBD-present group (n=28) and AUPBD-absent group (n=16). Clinical features were compared between the two groups. Furthermore, in AUPBD-present group, clinical data were also analyzed according to Kimura's classification of AUPBD.*

Results : *In our study, AUPBD was associated with choledochal cyst in 28 (64%) cases. AUPBD was found only in type I and IV according to Todani's classification of choledochal cyst. There were no significant differences between the AUPBD-present group and the AUPBD-absent group in the incidence of gallstone disease, while the incidence of acute inflammation was 93% (26/28) in the AUPBD-absent group ($p<0.01$). Carcinoma developed only in the AUOBD-present group (9/28, 32%) ($p<0.05$). Pancreatic disorders (i.e. pancreatic stone, pancreatitis or pancreatic cancer) occurred in 12 of 28 cases in the AUPBD-present group(43%), while only in 1 of 16 cases in the AUPBD-absent group(6%)($p<0.05$).*

Conclusion : *AUPBD associated with choledochal cyst may have implications not only as a possible etiologic factor but also as an important factor that may affect the clinical course, surgical planning and prognosis. In cases with choledochal cyst, we should make an effort to evaluate the presence of AUPBD.*

Key Words : *Anomalous union of pancreaticobiliary duct, Choledochal cyst*

INTRODUCTION

Choledochal cyst is a congenital dilatation of the bile duct. This congenital disease is relatively rare in western countries and more than two thirds of the cases were reported in Japan[1]. Anomalous union of pancreaticobiliary duct (AUPBD) has been regarded to be the etiological factor of the choledochal cyst[2]. However, choledochal cyst is not always associated with AUPBD and the presence of AUPBD without choledochal cyst has been

Address reprint requests to : Myung-Hwan Kim, M.D., Department of Internal Medicine, Asan Medical Center, 388-1 Pungnap-dong, Songpa-gu, Seoul 138-736, Korea

increasingly recognized recently, probably because of the advances in hepatobiliary imaging techniques. Therefore, some authors suggest that these two anomalies should be considered separately[3]. There have been many reports about choledochal cyst or AUPBD, but their cases were mainly infants or neonates. Moreover, cholangiography was obtained by the percutaneous transhepatic route or intraoperatively[4]. Thus, the pancreaticobiliary junction might have been fully evaluated in previous series.

While most of the choledochal cysts were observed in infants, it can also be found in adults[5]. All of our cases had choledochal cyst diagnosed in adulthood and the presence or absence of AUPBD was confirmed by endoscopic retrograde cholangiopancreatography (ERCP). The number of the cases was 44, the largest series having been collected from a single institution, except the ones from Japan. Moreover, there were rare reports in English literature which compared clinical features of choledochal cyst according to the presence of AUPBD. The purpose of our study was to compare the clinical characteristics of 44 cases with adulthood choledochal cyst according to the presence or absence of AUPBD. Furthermore, we tried to clarify the significance of AUPBD in patients with choledochal cyst.

METHODS

From August 1990 to December 1996, 52 cases (1.03%) were diagnosed as having choledochal cyst out of 5,037 ERCP referrals. The diagnosis of choledochal cyst was made as a localized non-proportional dilatation of bile duct after exclusion of tumor, stone or inflammation as a cause of the dilatation[6]. All of the patients in our series were more than 16 years of age. Of the 52 choledochal cyst cases, we selected 44 cases, of which the pancreaticobiliary junction was clearly visualized.

Choledochal cyst was classifled as I, II, III, IVa, IVb, V according to Todani's classification. Type I is cystic or diffuse dilatation of extrahepatic bile duct, type II is a diverticulum in the extrahepatic bile duct, type III is choledochocele, type IVa is multiple cystic dilatation of intra-and extrahepatic bile duct, type IVb is multiple cystic dilatation of extrahepatic bile duct and type V is multiple cystic dilatation of intrahepatic bile ducts (Caroli's disease)[7]. AUPBD was defined as the anomalous union of pancreaticobiliary duct system at a distance > 15 mm from the papilla of Vater[8]. This anomaly was divided into type II (Fig. 1 & 2) and II (Fig. 3) according to Kimura's classification. Type I AUPBD looks as though the pancreatic duct joins the bile duct, which is the major duct, whereas in type II, it looks as though the bile duct joins the pancreatic duct, which is the major duct[8].

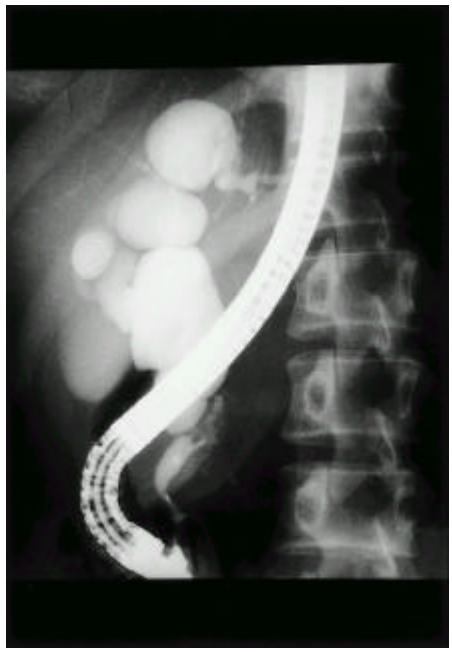

Fig. 1. Endoscopic retrograde cholangiopancreatogram of choledochal cyst (Todani type IVa) and anomalous union of pancreaticobiliary duct (Kimura type I.) Pancreatic duct is partially visualized due to incomplete filling.

The cases with choledochal cyst were divided into those associated with AUPBD (n=28, AUPBD-present group) and those without (n=16, AUPBD-absent group) and clinical characteristics were compared between the two groups. Furthermore, in the AUPBD-present group, characteristics were also compared according to Kimura's classification of AUPBD. The angle of the pancreaticobiliary junction was measured as viewed frontally. Statistical analysis was made by Fisher's exact test and Mann Whitney U test.

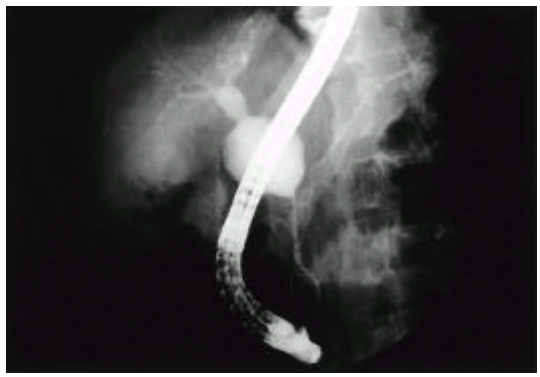

Fig 2. Endoscopic retrograde cholangiopancreatogram of choledochal cyst (Todani type IVb) and anomalous union of pancreaticobiliary duct(Kimura type I).

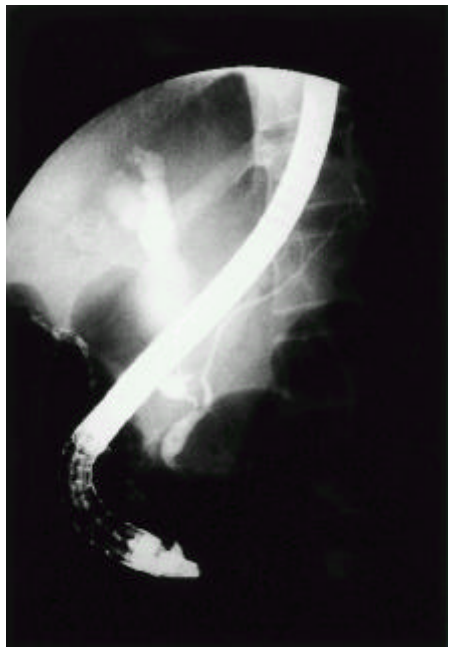

Fig. 3. Endoscopic retrograde cholangiopancreatogram of choledochal cyst (Todani type IVa) and anomalous union of pancreaticobiliary duct (Kimura type II).

RESULTS

Of the 44 choledochal cyst cases, 17 cases had type I cysts, 1 had type II, 6 had type III, 18 had type IVa, 1 had type IVb, and 1 had type V. The AUPBD-present group was 28 (64%), while the AUPBD-absent group was 16 (36%). AUPBD was observed only in type I and IV patients, whereas it was not shown in type II, III, V patients (Table 1). Age distribution of choledochal cyst was 7 in 16-19 years, 7 in 20-29 years, 11 in 30-39 years, 7 in 40-49 years, 5 in 50-59 years and 7 in 60-69 years (Table 2). There were 15 males and 29 females (M:F ratio, 1 :1.9).

Table 1. Type of choledochal cyst by Todani's classification

Type	Cyst+AUPBD (n=28)	Cyst-AUPBD (n=16)	Total
I	12	5	17
II	0	1	1
III	0	6	6
IVa	15	3	18
IVb	1	0	1
V	0	1	1

AUPBD:anomalous union of pancreaticobiliary duct

Table 2. Age distribution of choedochal cyst

Age (Yrs)	Cyst+AUPBD (n=28)	Cyst-AUPBD (n=16)	Total
16-19	5	2	7
20-29	6	1	7
30-39	8	3	11
40-49	4	3	7
50-59	4	1	5
60-69	1	6	7

AUPBD;anomalous union of pancreaticobiliary duct

Comparing the characteristics according to the AUPBD-association, female cases were more observed in both groups, whereas the mean age of the AUPBD-present group was 49.2 years, younger than that of the AUPBD-absent group, although it was not statistically significant (Table 3). Gallstone diseases were associated in 18 (41%) patients with choledochal cyst (n=44). The location of the gallstones was 11 in the cyst, 5 in the gallbladder and 2 in the intrahepatic duct. Pancreatic stones were shown in 2 (5%) patients. Acute inflammation was observed in 31 (70%) cases. They were cholecystitis (n=12), cholangitis (n=8) and pancreatitis (n=11). Malignant neoplasm occurred in 9

Table 3. Differences of clinical characteristics and associated diseases according to the presence of AUPBD

	Cyst+AUPBD (n=28	Cyst-AUPBD (n=16)
Age(mean+SD, yrs)	36.5± 14.1	49.2± 12.2
Sex(M:F)	9 : 19	6 : 10
Stone		
Cystolithiasis	5	6
Gallbladder stone	2	3
Intrahepatic stone	1	1
Pancreatic stone	2	0
	(n=10)	**(n=10)**
Acute inflammation		
Cholecystitis	10	2
Cholangitis	6	2
Pancreatitis#	10	1
	(n=26)*	**(n=5)**
Malignancy		
GB cancer	3	0
CBD cancer	4	0
Pancreatic cancer	2	0
	(n=9)**	**(n=0)**
Pancreatic diseases	**12 (43%)****	**1(6%)**

AUPBD: anomalous union of pancreaticobiliary duct; GB: gallbladder; CBD: common bile duct; *:p<0.01;**:p<0.05, compared to that of cyst-AUPBD, by Fisher's exact test; #:3-fold or more elevation of amylase level associated with acute abdominal pain, two cases with pancreatic stones were included.

(20%) cases: gallbladder in 3, common bile duct in 4 and pancreas in 2 cases. All the cancers in the common bile duct arose from the cyst wall and were adenocarcinoma, pathologically.

The difference in the incidence of associated diseases according to the presence of AUPBD was as follows. The incidence of gallstone disease in the AUPBD-present group did not differ from that in the AUPBD-absent group, while acute inflammation occurred more frequently in the AUPBD-present (26/28, 93%) than in the AUPBD-absent group (5/16, 31%) (p<0.01). Malignant neoplasm developed only in the AUPBD-present group (9/28, 32%), more often than in the AUPBD-absent group (0/16, p<0.05) (Table 3). Pancreatic disorders (pancreatic stone, pancreatitis or pancreatic cancer) occurred in 12 of 28 cases (43%) in the AUPBD-present group, whereas only in 1 of 16 (6%) cases of the AUPBD-absent group (p<0.05, Table 3).

According to Kimura's classification, AUPBD (n=28) was divided into type I (n=12) and II (n=16), but we could not find any difference in associated diseases between the two groups (Table 4). The maximal diameter of the common bile duct was higher in type II AUPBD, but it was not statistically significant. However, the angle between the biliary and pancreatic duct was higher in type II (85.1°) than in type I (45.2°)(p<0.05, Table 4).

DISCUSSION

At the Children's Hospital of Los Angeles, choledochal cyst was diagnosed in 0.5 patients per year[10]. However, in our institution (3rd referral center), we experienced 53 cases during 6 years (incidence of about 9 per year). Our data suggest that choledochal cyst is more prevalent in Korea than in western countries. It also implies that the incidence of choledochal cyst may be higher not only in Japan but also in other oriental countries than in western countries[11]. Since the earlier report by Babbit et al.[12] about the frequent association of AUPBD with choledochal cyst, AUPBD has been regarded as an

Table. 4. Differences of clinical characteristics and associated diseases according to the type of AUPBD

	AUPBD	
	Type I (n=12)	Type II (n=16)
CBD diameter	30.5±3.4mm	42.5±3.7mm
Length of CC	22.7±1.5mm	22.8±1.7mm
Angle*(°)	45.2±15.4	85.1±16.2#
Type of choledochal cyst		
Type I	5	7
Type IVa	6	9
Type IVb	1	0
Stone		
cystolithiasis	3	2
GB stone	1	1
IHD stone	0	1
pancreatic stone	0	2
Acute inflammation		
cholecystitis	5	5
cholangitis	2	4
pancreatitis	4	6**
Malignancy		
GB cancer	2	1
CBD cancer	2	2
pancreatic cancer	0	2
Pancreatic diseaes	4(33%)	8(50%)

AUPBD: anomalous union of pancreaticobiliary duct; CBD: common bile duct; CC: common channel; GB: gallbladder; IHD: intrahepatic duct; *: angle which was formed when common bile duct and pancreatic duct joined; #:$p<0.05$, compared to that of type I, by Mann Whitney U test; **:two cases with pancreatic stones were included.

etiological factor of choledochal cyst[2]. However, the rate of AUPBD association with choledochal cyst was from 33% to 100% according to the reports[4, 13-15]. It might be due to the difference in the characteristics of the selected cases. According to our results, AUPBD was associated only with type I and IV choledochal cyst (Table 1). Therefore, the rate of AUPBD can be affected by the number of cases with type I or IV. Furthermore, Todani et al.[6] sub-classified type I choledochal cyst into type Ia, Ib, Ic, and suggested that AUPBD may not be associated in type I. This also implies that the association rate of AUPBD can be influenced even by the sub-classification in type I choledochal cyst. Moreover, Matsumoto et al.[16] divided choledochal cyst into childhood-type and adulthood-type. Association of AUPBD was observed in almost 100% of childhood-type, whereas less frequently found in adulthood-type. The difference of association rate in many reports, including ours, may be explained by the difference in the characters of the cases included. Although AUPBD was considered as an etiological factor of choledochal cyst[2, 12], the pathogenetic mechanism of choledochal cyst may not be explained solely by AUPBD because AUPBD was not found in all of the choledochal cyst cases. Our data also showed that AUPBD was not always associated with choledochal cyst cases (28/44, 64%). The increasing reports of AUPBD without choledochal cyst support the

notion[17].

In AUPBD, a union of the pancreatic and biliary ducts is located outside the sphincter of Oddi. Therefore, two-way regurgitation occurs. Pancreatic juice refluxes into the common bile duct, or bile regurgitates into the pancreatic duct because the action of the sphincter muscle does not functionally affect the union[18]. Because the intraductal pressure is generally higher in the pancreatic duct than in the bile duct, pancreatic juice regurgitates into the biliary tract resulting in the pancreatic enzyme activation and subsequent recurrent inflammation. These may give rise to metaplastic and, finally, malignant change of the biliary epithelium[19]. Furthermore, after recurrent inflammation of the bile duct, the pressure in the bile duct rises and bile may reflux into the pancreatic duct causing various pancreatic disorders, including acute or chronic pancreatitis, pancreatic stone or pancreatic cancer[18].

The incidence of malignant diseases in choledochal cyst is said to be 2.5%-15%, 15 times greater than the control population without choledochal cyst[20]. In our series, cancer developed in 9 of 44 (20%). The incidence in our series might be higher than in other reports because all of our cases were in adulthood. It is well known that the incidence of cancer in choledochal cyst increases with age[21]. Moreover, cancer developed only in the AUPBD-present group, which implies that AUPBD may be a more important contributing factor than the choledochal cyst itself. Flanigan[22] pointed out that only 57% of cancers occurred in choledochal cyst were located in the cyst wall and the rest of cancers developed in bile duct other than the cyst wall. Moreover, Nagorney et al.[23] also suggested that malignant neoplasm developed in choledochal cyst is not always located in the cyst wall. In one of their cases, cancer developed in the remaining bile duct after complete cyst excision. These data imply that risk factors causing cancer in choledochal cyst are more than the choledochal cyst itself. The cancers developed in our series were located in gallbladder (n=3), common bile duct (n=4) and pancreas (n=2). Only 4 cases occurred in the cyst wall.

Recent reports of gallbladder cancer in AUPBD cases without choledochal cyst suggest that AUPBD is more important for the carcinogenic process than the choledochal cyst itself[24]. In 35 cases of choledochal cyst described by Yoshida et al., 8 cases developed cholangiocarcinoma[25]. AUPBD was associated with all of these 8 cases, which supports our notion that AUPBD may be the major contributing factor for cancer development. Suda et al. examined AUPBD in 34 bile duct cancer patients, 24 gallbladder cancers and 171 controls without biliary disease[26]. They observed AUPBD in 8 of 34 cholangiocarcinoma, 4 of 24 gallbladder cancer, but none in controls. They suggested that AUPBD is one of the pathogenetic factors in biliary malignancy.

In our results, acute inflammatory condition, such as cholecystitis, cholangitis or pancreatitis, was more prevalent in the AUPBD-present than in the AUPBD-absent group. The younger age in the AUPBD-present group suggests that patients in this group might visit hospitals earlier because of more severe symptoms.

One of the factors worth mentioning is pancreatic diseases associated with choledochal cyst. There had been several reports about the association of pancreatitis, pancreatic stone or pancreatic cancer in choledochal cyst[20,27-29]. In the previous reports, however, they did not analyze the data considering AUPBD. In one Japanese report, acute pancreatitis occurred in 30 (17%) of the 176 cases with AUPBD[30]. Activated pancreatic enzymes, after entering the biliary tract, may cause cholangitis, gallstone and cholangiocarcinoma[31]. Likewise, these enzymes may reflux back into the pancreatic duct and cause various pancreatic disorders, such as acute or chronic pancreatitis and pancreatic cancer[32]. In our results, pancreatic disorders developed more frequently in the AUPBD-associated group (Table 3).

These high incidences of malignancy and inflammatory diseases associated with AUPBD also have therapeutic implications for choledochal cyst. In cases of choledochal cyst with AUPBD, cholecystectomy also should be performed in addition to cyst excision because the incidence of gallbladder cancer is very high. Moreover, surgical procedure for correction of AUPBD should be added. Biliary diversion from the pancreatic juice (pancreaticobiliary disconnection) may be needed for prevention of bi-directional reflux of pancreatic and bile juice[23,24]. In this regard, cholecystectomy along with the resection of dilated bile duct and the biliary diversion from pancreatic juice should be performed in cases with choledochal cyst and AUPBD. Komi et al.[35] subdivided

AUPBD associated with choledochal cyst into several categories and suggested that pancreatitis could not be prevented by cholecystectomy, cyst excision and hepaticojejunostomy in certain subgroups. Furthermore, they suggested that, in cases with AUPBD showing dilated common channel or accessory pancreatic duct, sphincteroplasty or pylorus preserving pancreaticoduodenectomy should be needed in addition to the previously mentioned procedures. Schreiber et al.[36] described that AUPBD may be observed as two clinical manifestations. One is the biliary tract disease, such as acute cholecystitis, cholangitis and cholangiocarcinoma. The other one is caused by stasis of pancreatic fluid due to anomalous drainage in the common channels leading to periductal and interlobular fibrosis as a histological sign of chronic pancreatitis. Thus, Schreiber et al. suggested that resection of the anomalous junction and hepaticojejunostomy with a Roux-en-Y anastomosis may resolve both pancreatic reflux into the biliary system and stasis of the pancreatic secretion. The claim by Schreiber et al. has something to do with that of Komi et al. suggesting that hylorus-preserving pancreaticoduodenectomy may be recommended in certain cases of AUPBD.

AUPBD frequently associated with choledochal cyst may have an implication not only as an etiological factor but as an associated disorder leading to a grave clinical course. In this regard, we should make an effort to confirm the presence of AUPBD in patients with choledochal cyst. Moreover, adequate surgery may be required to prevent the occurrence of cancer. Cancer associated with choledochal cyst may often be in an advanced stage when detected. Curative resection may be difficult[20]. Prevention, therefore, may be the best way, if possible. AUPBD associated with choledochal cyst may be a very important factor that affects the clinical course, surgical planning and prognosis.

REFERENCES

1. Yamaguchi M: *Congenital choledochal cyst: analysis of 1,433 patients in the Japanese literature. Am J Surg 1980; 140:653-7.*
2. Babbitt DP, Starshak RJ, Clemett AR: *Choledochal cyst: a concept of etiology. Am J Roentgenol 1973, 119: 57-62.*
3. Anomaly of biliary tree and gallstones. In: Tsuchiya Y, Matsumoto Y, ed. *New trend in treatment for cholelithiasis. Tokyo. Kanehara Co, 1991; 211-30.*
4. Kaneko K, Ando H, Ito T, Watanabe Y, Seo T, Harada T, et al: *Protein plugs cause symptoms in patients with choledochal cysts Am J Gastroenterol 1997; 92:1018-21.*
5. Chijiwa K, Koga A: *Surgical management and long-term follow-up of patients with choledochal cysts. Am J Surg 1993; 165238-42.*
6. Todani T: *Definition and classification of congenital bile duct dilatation. Biliary Tract and Pancreas 1995, 16. 715-7 (in Japanese).*
7. Todani T, Watanabe Y, Narusue M, Tabuchi K, Okajima K: *Congenital bile duct cysts Am J Surg 1977; 134263-9.*
8. Kimura K, Ohto M, Saisho H, Unozawa T, Tsuchiya Y, Morita M, et al: *Association of gallbladder carcinoma and anomalous pancreaticobiliary ductal union. Gastroenterology 1985, 89:1258-65.*
9. Kimura K, Ohto M, Ono T, Tsuchiya Y, Saisho H, Kawamura K, et al: *Congenital cystic dilatation of the common bile duct: relationship to anomalous pancreaticobiliary ductal union. Am J Roentgenol 1977; 128:571-7.*
10. Hays DM, Goodman GN, Snyder WH Jr, Woolley M: *Congenital cystic dilatation of the common bile duct. Arch Surg 1969; 98:457-61.*
11. Komi N, Kin C, Lei M: *Choledochal cyst in the world. Biliary Tract and Pancreas 1995; 16:719-22 (in Japanese).*
12. Babbitt DP: *Congenital choledochal cysts: new etiological concept based on anomalous relationship of the common bile duct and pancreatic bulb. Ann Radiol 1969; 12231-40.*
13. Komi N, Udaka H, Ikeda N, Kashiwagi Y: *Congenital dilatation of the biliary tract: new classification and study with particular reference to anomalous arrangement of the pancreaticobiliary ducts. Gastroenterol Jpn 1977; 12293-304.*
14. Miyano T, Suruga K, Suda K: *Abnormal choledocho-pancreatico-ductal junction related to the etiology of infantile obstructive disease. J Pediatr Surg 1979; 14:16-26.*
15. Bakka A, Bergan A, Soreide O: *Bile duct cysts in adults. Scand J Gastroenterol 1991; 26:197-206.*
16. Matsumoto T, Fujii H, Itakura A, Subekada K: *congenital choledochal cyst-relationship between the gross configuration of the bile duct and the clinical aspects. Biliary Tract and Pancreas 1995; 16:729-34(in Japanese).*
17. Aoki H, Sugaya H, Shimazu M: *A clinical study on cancer of the bile duct associated with anomalous arrangements of pancreaticobiliary ductal system. Biliary Tract and Pancreas 1987; 8:1539-51(in Japanese).*
18. The Japanese Study Group on Pancreaticobiliary Maljunction (JSPBM): *The Committee of JSPBM for*

Diagnostic Criteria:Diagnostic criteria of pancreaticobiliary maJunction. J Hep Bil Panc Surg 1994; 1:219-21.

19. Shimada K, Yanagisawa J, Nakayama F: *Increased lysophospatidylcholine and pancreatic enzyme content in bile of patients with anomalous pancreaticobiliary ductal junction. Hepatology 1991; 13:438-44.*
20. Todani T, Tabuchi K, Watanabe Y, Kobayashi T: *Carcinoma arising in the wall of congenital bile duct cysts. Cancer 1979; 44:1134-41.*
21. Chijiwa K, Nagai E, Makino I, Shimada K: *Are secondary bile acids in choledochal cysts important as a risk factor in biliary tract carcinoma? Aust N Z J Surg 1993; 63:109-12.*
22. Flanigan DP: *Biliary carcinoma associated with biliary cysts. Cancer 1977; 40:880-3.*
23. Nagorney DM, McIlrath DC, Adson MA: *Choledochal cysts in adults: clinical management. Surgery 1984; 96:656-63.*
24. Yamauchi S, Koga A, Matsumoto S, Tanaka M, Nakayama F: *Anomalous junction of pancreaticobiliary duct without congenital choledochal cyst: a possible risk factor for gall bladder cancer. Am J Gastroenterol 1987; 82:20-4.*
25. Yoshida H, Itai Y, Minami M, Kokubo T, Ohtomo K, Kuroda A: *Biliary malignancies occurring in choledochal cyst. Radiology 1989; 173:389-92.*
26. Suda K, Miyano T, Konuma I, Matsumoto M: *An abnormal pancreatico-choledocho-ductal junction in cases of biliary tract carcinoma. Cancer 1983; 52:2086-8.*
27. Toki F, Oi I, Saito S, Tomatsu S, Kozu T, Takeuchi T, et al: *Two cases of congenital choledochal cyst with translucent pancreatic stone. Jpn J Gastroenterol 1979; 76:1146-51 (in Japanese).*
28. Hatayama K, Shibata N, Moden M, et al: *A case of idiopathic choledochal dilatation combined with dilated pancreatic duct and pancreatic calculi. Jpn J Gastroenterol Surg 1979; 12:940-3 (in Japanese).*
29. Munakata H, Hada R, Tohyama S: *Congenital bile duct dilatation with pancreatolithiasis. Jpn J Pediatr Surg 1982; 18:1017-22 (in Japanese).*
30. Funabiki T, Matsubara T, Ochiai M: *Symptoms, diagnosis and treatment of pancreaticobiliary maJunction associated with congenital cystic dilatation of bile duct. J Jpn Surg soc 1996; 97:582-8 (in japanese).*
31. Todani T, watanabe Y, Fujii T, Uemura S: *Anomalous arrangement of the pancreatobiliary ductal system in patients with a choledochal cyst. Am J Surg 1984; 147:672-6.*
32. Okada A: *Pancreaticobiliary maJunction and congenital dilatation of bile duct. J Jpn Surg Soc 1996; 97:589-93 (in Japanese).*
33. Nagata E, Sakai K, Kinoshita H, Kobayashi Y: *The relation between carcinoma of the gallbladder and an anomalous connection between the choledochus and the pancreatic duct. Ann Surg 1985; 202-182-90.*
34. Nagata E, Sakai K, Kinoshita H, Hirohashi K: *Choledochal cyst: complications of anomalous connection between the choledochus and pancreatic duct and carcinoma of the biliary tract. World J Surg 1986; 10:102-10.*
35. Komi N, Takehara H Kunitomo K, Miyoshi Y, Yagi T: *Does the type of anomalous arrangement of pancreaticobiliary ducts influence the surgery and prognosis of choledochal cyst? J Pediatr Surg 1992; 27:728-31.*
36. Schreiber F, Gurakuqi GCH, Schnedl W, Trauner M: *ERCP and balloon dilatation are valuable alternatives to surgical biliodigestive anastomosis in the long, common channel syndrome in childhood. Endoscopy 1996; 28:724-5(Letter to the Editor).*
37. Yamashiro Y Miyano T, Suruga K, Shimomura H, Suda K, Matsumoto M: *Experimental study of the pathogenesis of choledochal cyst and pancreatitis, with special reference to the rate of bile acids and pancreatic enzymes in the anomalous choledocho-pancreatico-ductal junction. Pediatr Gastroenterol Nutr 1984; 2:721-7.*