

# Anomalous pancreaticobiliary junction: image analysis and treatment principles

Ze-Li Yu, Li-Jun Zhang, Jian-Zhu Fu, Jie Li, Qing-Yu Zhang and Fou-Lai Chen

Beijing, China

**BACKGROUND:** Anomalous pancreaticobiliary junction is often associated with biliary tract carcinoma and acute pancreatitis. We assessed the value of image analysis in the diagnosis of patients with anomalous pancreaticobiliary junction (APBJ) and the principles for the treatment of APBJ.

**METHODS:** Sixty-four patients with APBJ were subjected to ultrasound imaging, endoscopic retrograde cholangiopancreatography (ERCP) and magnetic resonance cholangiopancreatography (MRCP) before surgery. The diagnostic accuracy of image analysis and their surgical outcomes were evaluated retrospectively.

**RESULTS:** On ERCP and MRCP, the length of the common channel was calculated to be 15 mm or longer in all patients, and the angle of the junction was more than 75° in 49 (76.6%) of the 64 patients. Of the 64 patients, 28 were defined of pancreatic duct type (P-C) (28/64, 43.75%), 32 bile duct type (C-P) (32/64, 50%), and 4 common channel type (4/64, 6.25%).

**CONCLUSIONS:** Patients with APBJ are often associated with biliary tract and pancreatic diseases, and early detection and correct surgical treatment could avoid serious complications. ERCP and MRCP are accurate in the diagnosis of APBJ.

(*Hepatobiliary Pancreat Dis Int* 2004; 3: 136-139)

**KEY WORDS:** anomalous pancreaticobiliary junction; image examination; surgical treatment

## Introduction

In anomalous pancreaticobiliary junction (APBJ), which is connected with the pancreatic and biliary ducts outside the duodenal wall to form a long common channel. It is often associated with biliary tract carcinoma. Patients with APBJ have also been reported to

frequently develop acute pancreatitis.<sup>[1]</sup> Although clinical and genetic studies of the biliary epithelium in patients with APBJ have been reported,<sup>[3]</sup> clinicopathologic features of APBJ are unknown. To identify the characteristics of APBJ and analyze imaging results, we report our experience in 64 patients with APBJ at our hospital, who had been detected by ERCP, MRCP and operative or postoperative cholangiography.

## Methods

### Patients

From February 1979 through August 2001, we treated surgically 3720 patients with pancreatic and biliary diseases, including 64 patients with APBJ (1.7%) at our hospital. Of the 64 patients, 22 were men and 42 women with a mean age of 46.8 years (range 15 to 70 years). Their clinicopathological findings, associated pancreatic and biliary disorders, diagnostic imaging, treatment and long-term outcomes were reviewed retrospectively, including particularly the cholangiopancreatographic results obtained by preoperative ERCP ( $n=52$ ) and MRCP ( $n=16$ ) as well as operative or postoperative cholangiographic ones ( $n=60$ ).

### Clinical findings

All the 64 patients complained of upper abdominal pain and nausea. Twenty-one (32.8%) of these patients were associated with obstructive jaundice; C-P type (13 patients) and P-C type (8). Sixteen (25%) patients had increased level of amylase, and 39 (60.9%) had abnormal hepatic function.

### Pancreatic and biliary disorders

In the 64 patients, 32 (50%) had congenital choledochal cyst. Their diameter of common bile ducts was measured 32 mm to 79 mm. In 40 patients (62.5%) with cholelithiasis, 17 patients had gallstones and biliary tree stones (the diameter of their common bile ducts was measured 11 mm to 27 mm) and 23 had gallstones. The diameter of the common bile ducts was normal. Twenty-one patients were associated with biliary tract neoplasm (32.8%); carcinoma of the common bile duct (9), carcinoma (8), and adenoma (4) of the gallblad-

**Author Affiliations:** Department of General Surgery, Beijing Tongren Hospital, Capital University of Medical Sciences, Beijing 100730, China (Yu ZL, Zhang LJ, Fu JZ, Li J, Zhang QY and Chen FL)

**Corresponding Author:** Ze-Li Yu, MD, Department of General Surgery, Beijing Tongren Hospital, Capital University of Medical Sciences, Beijing 100730, China (Tel: 86-10-65129911 ext 2551; Email: zhlj1968@126.com)

© 2004, Hepatobiliary Pancreat Dis Int. All rights reserved.

der. In 16 patients (25%) with acute or chronic pancreatitis, 11 belonged to C-P type with a dilated common bile duct and 5 P-C type with gallstones. One of the 64 patients suffered from pancreatic head carcinoma.

### Treatment

In the 64 patients, 32 patients associated with congenital choledochal cyst underwent excision of the dilated extrahepatic bile duct and Roux-en-Y hepaticojejunostomy; carcinoma of the common bile duct was identified histologically in 2 patients. In 40 patients who were associated with cholelithiasis were treated by cholecystectomy, 17 patients were subjected to common bile duct exploration and T-tube drainage during the procedure. Nine of the 64 patients had carcinoma of the common bile duct; advanced carcinoma in 4 patients, who had to receive palliative choledochojejunostomy and the other 5 patients were treated by excision of the extrahepatic bile duct, extensive dissection of lymph nodes, and Roux-en-Y hepaticojejunostomy. Eight patients were associated with carcinoma of the gallbladder; extended cholecystectomy was performed in 7 patients and operative exploration in 1 patient because of advanced tumor. Four patients with adenoma of the gallbladder underwent cholecystectomy alone. Of the 64 patients, one who suffered from carcinoma of the pancreatic head underwent palliative side-to-side choledojejunostomy because of the involvement of the portal vein. During the operation the amylase level of gallbladder bile in the 64 patients was higher than 300  $\mu\text{L}$ , with the highest being 20 000  $\mu\text{L}$ .

### Results

The common channel was measured 17 mm to 49 mm (mean 19 mm) in the 64 patients, and the angle of



Fig. APBJ (C-P type) with congenital choledochal cyst showing the long common channel.

junction was more than 75° in 49 (76.6%) of them. Of all patients, 32 were defined of bile duct type (C-P) (Fig.), 4 common channel type, and 28 pancreatic duct type (P-C). The diameter of the common bile duct was measured 6 mm to 79 mm (mean 18 mm).

### Discussion

APBJ is a rare congenital anomaly, in which the pancreatic and biliary ducts are joined outside the duodenal wall.<sup>[1,2]</sup> It is frequently associated with congenital choledochal cyst and carcinoma in the biliary tract.<sup>[3-6]</sup>

In APBJ, a long common channel of over 15 mm shown by endoscopic retrograde cholangiopancreatography comprises the common bile duct and the major pancreatic duct. It is usually divided clinically into three types: C-P, P-C and common channel.<sup>[7]</sup> In our 64 patients, 32 (50%) belonged to C-P, 28 (43.75%) P-C and 4 (6.25%) common channel.

Usually no specific symptoms or signs are seen in APBJ patients. Clinical diagnosis of APBJ is dependent on its complications. The only significant criterion of diagnosis is the high amylase level of gallbladder bile, in some cases, even higher than 10 000  $\mu\text{L}$ .<sup>[8-11]</sup> Screening for APBJ is valuable in patients with suspected anomaly who show a dilated bile duct, polypoid gallbladder lesions, diffusely thickened gallbladder mucosa on ultrasonography or acute pancreatitis of unknown cause. When ERCP or/and MRCP show that the common channel is longer than 15 mm and the junction is located outside of the duodenal wall, APBJ should be identified. ERCP depicts APBJ most reliably but is invasive, and there is a potential risk of morbidity.<sup>[12-15]</sup> MRCP is a noninvasive and accurate technique for diagnosing APBJ, but is less accurate than ERCP.<sup>[16-19]</sup>

APBJ allows pancreaticobiliary or biliopancreatic reflux.<sup>[20-24]</sup> Refluxed proteolytic pancreatic enzymes and phospholipase A<sub>2</sub> are activated in the biliary tract. These activated enzymes and lysophatidylcholine produced by phospholipase A<sub>2</sub> may induce biliary tract carcinoma. When APBJ presents with no dilated common bile duct, these activated enzymes and lysophatidylcholine would concentrate in the gallbladder, and neoplasm is likely induced. Although the hydropressure in the pancreatic duct is usually higher than that in the bile duct, bile may reflux into the pancreatic duct via APBJ in some situation. The refluxed bile may activate pancreatic enzymes, particularly phospholipase A<sub>2</sub>, which may cause acute pancreatitis.

APBJ is known to be frequently associated with carcinoma in the biliary tract and pancreatic disorders.<sup>[24,25]</sup> The incidence of biliary carcinoma is reported to be as high as 15.6% to 36% in adult patients with APBJ.<sup>[21,26,27]</sup> Hence early diagnosis and treatment of APBJ

before a malignancy arises in the biliary tract is extremely important. The treatment of choice<sup>[28-30]</sup> for APBJ is excision of the extrahepatic bile tract and gallbladder in patients with congenital choledochal cyst, or cholecystectomy alone in patients without congenital choledochal cyst because of high incidence of carcinoma of the gallbladder among them.

In conclusion, patients with APBJ often develop acute pancreatitis (25% to 31%), and they are likely to develop biliary tract neoplasm (32.8%). Clinically, attention should be given to the possibility of APBJ in the patients who experienced acute pancreatitis and acute cholangitis or cholelithiasis. When one is suspected of APBJ, ERCP and MRCP are valuable. Once the diagnosis of APBJ is established, APBJ with choledochal cyst should be treated by cyst excision and hepaticojejunostomy to avoid serious complications of the pancreatic and biliary tracts. And cholecystectomy alone may be adequate for APBJ with or without cholelithiasis in the absence of choledochal cyst.

### Competing interest

No benefits in any form have been received or will be received from a commercial party related directly or indirectly to the subject of this article.

### References

- Komuro H, Makino S, Yasuda Y, Ishibashi T, Tahara K, Nagai H. Pancreatic complications in choledochal cyst and their surgical outcomes. *World J Surg* 2001;25:1519-1523.
- Todani T, Watanabe Y, Toki A, Urushihara N. Carcinoma related to choledochal cysts with internal drainage operation. *Surg Gynecol Obstet* 1987;164:61-64.
- Nagai M, Watanabe M, Iwase T, Yamao K, Isaji S. Clinical and genetic analysis of noncancerous and cancerous biliary epithelium in patients with pancreaticobiliary maljunction. *World J Surg* 2002;26:91-98.
- Babbitt DP. New etiological concept based on anomalous relationships of the common bile duct and pancreatic bulb. *Ann Radil* 1969;12:231.
- Chaudhary A, Dhar P, Sachdev A, Kumar N, Vij JC, Sarin SK, et al. Choledochal cysts differences in children and adults. *Br J Surg* 1996;83:186-188.
- Sugiyama M, Atomi Y. Anomalous pancreaticobiliary junction without congenital choledochal cyst. *Br J Surg* 1998;85:911-916.
- Oguchi Y, Okada A, Nakamura T, Okumura K, Miyata M, Nakao K, et al. Histopathological studies of congenital dilatation of the bile duct as related to anomalous junction of the pancreaticobiliary ductal system. *Clinical and experimental studies. Surgery* 1986;103:168.
- Chen FL, Yu ZL, Li SY, Gong JZ. APBJ and its complications of the biliary tract. *Chin Pract J Surg* 1992;6:317-318.
- Sugiyama M, Baba M, Atomi Y, Hanaoka H, Mizutani Y, Hachiya J. Diagnosis of anomalous pancreaticobiliary junction: value of magnetic resonance cholangiopancreatography. *Surgery* 1998;123:391-397.
- Sugiyama M, Atomi Y, Kuroda A. Pancreatic disorders associated with anomalous pancreaticobiliary junction. *Surgery* 1999;126:492-497.
- Khan TF, Reddy KS, Johnston TD, Ranjan D. Sphincter of Oddi dysfunction and the anomalous pancreaticobiliary junction. *J Hepatobiliary Pancreat Surg* 2003;10:396; author reply 397.
- Sugiyama Y, Kobori H, Hakamada K, Sasaki M. Altered bile composition in the gallbladder and common bile duct of patients with anomalous pancreaticobiliary ductal junction. *World J Surg* 2000;24:17-21.
- Takaya J, Muneyuki M, Tokuhara D, Takada K, Hamada Y, Kobayashi Y. Congenital dilatation of the bile duct: changes in diagnostic tools over the past 19 years. *Pediatr Int* 2003;45:383-387.
- Choi BH, Lim YJ, Yoon CH, Kim EA, Park YS, Kim KM. Acute pancreatitis associated with biliary disease in children. *J Gastroenterol Hepatol* 2003;18:915-921.
- Hu B, Gong B, Zhou DY. Association of anomalous pancreaticobiliary ductal junction with gallbladder carcinoma in Chinese patients; an ERCP study. *Gastrointest Endosc* 2003;57:541-545.
- Okada A, Nakamura T, Higaki J, Fukui Y, Kamata S. Congenital dilatation of the bile duct in 100 instances and its relationship with anomalous junction. *Surg Gynecol Obstet* 1990;171:291.
- Li S, Zhang D, Shi B, Li Z, Chen X. Anomalous junction of pancreaticobiliary duct in congenital bile-duct cystic and cylindrical dilatation. *Zhonghua Wai Ke Za Zhi* 2000;38:349-351, 24.
- Sugiyama M, Abe N, Tokuhara M, Masaki T, Mori T, Atomi Y. Pancreatic carcinoma associated with anomalous pancreaticobiliary junction. *Hepatogastroenterology* 2001;48:1767-1769.
- Yano Y, Tasaka K, Okutani T, Maeda Y, Mori T, Tamura M, et al. A case of undifferentiated carcinoma of the gallbladder with anomalous arrangement of the pancreaticobiliary ductal system. *Oncol Rep* 2001;8:1281-1283.
- Kimura k, Ohto M, Saisho H, Unozama T, Tsuchiya Y, Morita M, et al. Association of gallbladder carcinoma and anomalous pancreaticobiliary ductal union. *Gastroenterology* 1985;89:1258.
- Lipsett PA, Pitt HA, Colombani PM, Boitnott JK, Cameron JL. Choledochal cyst disease; a changing pattern of presentation. *Ann Surg* 1994;220:644-652.
- Zhao L, Li Z, Ma H, Zhang X, Mou X, Zhang D, et al. Congenital choledochal cyst with pancreatitis. *Chin Med J* 1999;112:637-640.
- Tanaka M, Shimizu S, Mizumoto K, Yokohata K, Chijiwa K, Yamaguchi K, et al. Laparoscopically assisted resection of choledochal cyst and Roux-en-Y reconstruction. *Surg Endosc* 2001;15:545-552.
- Sato M, Ishida H, Konno K, Naganuma H, Ishida J, Hirata M, et al. Choledochal cyst due to anomalous pancreaticobiliary junction in the adult; sonographic findings. *Abdom Imaging* 2001;26:395-400.
- Tazuma S, Kajiyama G. Carcinogenesis of malignant lesions of the gall bladder. The impact of chronic inflammation and gallstones. *Langenbecks Arch Surg* 2001;386:224-229.
- 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. *Surge-*

- ry 1999;126:939-944.
- 27 Elnemr A, Ohta T, Kayahara M, Kitagawa H, Yoshimoto K, Tani T, et al. Anomalous pancreaticobiliary ductal junction without bile duct dilatation in gallbladder cancer. *Hepatogastroenterology* 2001;48:382-386.
- 28 Tanaka K, Ikoma A, Hamada N. Biliary tract cancer accompanied by anomalous junction of pancreaticobiliary ductal system in adults. *Am J Surg* 1998;175:218.
- 29 Nakayama K, Konno M, Kanzaki A, Morikawa T, Miyashita H, Fujioka T, et al. Allelotype analysis of gallbladder carcinoma associated with anomalous junction of pancreaticobiliary duct. *Cancer Lett* 2001;166:135-141.
- 30 Nakakubo Y, Kondo S, Omi M, Hirano S, Anbo Y, Morikawa T, et al. Endocrine cell carcinoma of the gallbladder with anomalous pancreaticobiliary ductal junction. *Hepatogastroenterology* 2000;47:1538-1540.

*Received May 16, 2003*

*Accepted after revision December 4, 2003*

---

What we call the beginning is often the end and to make an end is to make a beginning.  
The end is where we start from.

— T. S. Eliot