J Hep Bil Pancr Surg (1994) 1:236–239

Journal of HBP Surgery

Type classification of anomalous pancreaticobiliary junction by ERCP

KAZUNORI YOKOHATA, HIROSHI KIMURA, GEN NARITOMI, HIROYUKI KONOMI, TORAHIKO TAKEDA, YOSHIAKI OGAWA, and MASAO TANAKA[1]

[1] First Department of Surgery, Kyushu University Faculty of Medicine, Maidashi 3-1-1, Higashi-ku, Fukuoka, 812 Japan

Abstract: The role of endoscopic retrograde cholangiopancreatography (ERCP) in the preoperative assessment of anomalous pancreaticobiliary junction was retrospectively evaluated in 74 consecutive patients (19 males and 55 females; aged 0–80 years). Sixty-three patients had congenital biliary dilatation and 11 did not. Type classification of congenital biliary dilatation was possible by ERCP alone in 45 patients (71%). The main causes of classification failure were previous bilio-enteric anastomosis and restriction of postural changes during ERCP due to general anesthesia in pediatric patients. Classification of anomalous junction was possible in 69 patients (93%). Technical difficulty in ERCP caused classification failure in 5 patients. Neoplastic lesions were found in 12 patients (16%) and all but 1 were correctly diagnosed by ERCP. We conclude that ERCP plays an important role in the preoperative diagnosis and type classification of anomalous pancreaticobiliary junction and congenital biliary dilatation.

Key words: anomalous pancreaticobiliary junction, endoscopic retrograde cholangiopancreatography congenital biliary dilatation

Introduction

Anomalous pancreaticobiliary junction (APBJ) is a rare malformation in which the bile duct and a pancreatic duct are joined at the extramural portion of the duodenum. Since the sphincter of Oddi does not exist at that portion, there is free reflux of the pancreatic juice into the bile duct and vice versa.[1]

APBJ is present in most patients with congenital biliary dilatation (CBD)[2] and in some with gallbladder cancer.[1] This condition has been reported to play an important role in dilatation of the bile duct,[3] and in neoplastic changes of the bile duct[4] and gallbladder.[1,5,6,7] Due to the malignant potential of the bile duct in the presence of APBJ, surgical intervention has changed from bilio-enteric drainage procedures to total excision of the extrahepatic dilated bile duct, followed by hepaticojejunostomy. Therefore, to accomplish the excisional procedure safely preoperative evaluation of the relevant anatomy has become important.[8–10]

APBJ was previously diagnosed only by intraoperative cholangiography or at autopsy.[11] With the advent of endoscopic retrograde cholangiopancreatography (ERCP), it has become possible to delineate APBJ preoperatively. The aim of this report was to assess the role of ERCP in the preoperative evaluation of APBJ.

Patients and methods

Records of ERCP examinations performed in our department between 1972 and May 1993 were reviewed. Patients with APBJ and/or congenital biliary dilatation were selected and their medical records were reviewed with respect to the efficacy of ERCP for: (1) the diagnosis and classification of CBD and APBJ, and (2) the diagnosis of accompanying neoplastic lesions and stones.

For grouping the patients with CBD, we employed the classification of Todani et al.[8] According to this classification, type I is a solitary fusiform extrahepatic cyst, type II an extrahepatic supraduodenal diverticulum, type III a choledochocele, type IVa fusiform extra- and intrahepatic cysts, type IVb multiple extrahepatic cysts, and type V multiple intrahepatic cysts or Caroli's disease.

The patients with APBJ were divided into three groups according to the classification proposed by

Offprint requests to: M. Tanaka
Received for publication on Nov. 4, 1993; accepted on Dec. 9, 1993

Komi et al.[12] According to this classification, in type a, the narrowed common bile duct joins the pancreatic duct at a site distant from the papilla, mostly at a right angle. In type b, the pancreatic duct joins the common bile duct at a site distant from the papilla, usually at slightly less than 90°. In type c, the pancreatic duct and terminal portion of the common bile duct form a complex arrangement.

For statistical analysis, the Mann-Whitney test or Chi-square analysis with Yates' correction was used. For multiple comparison, Kruskal-Wallis analysis of variance (ANOVA) by ranks, followed by the Mann-Whitney test with Bonferroni correction was employed. A P value less than 0.05 was considered significant.

Results

The review of the ERCP records yielded a total of 74 patients amenable to analysis. Of these, 63 had biliary dilatation, whereas the remaining 11 had APBJ but no dilatation. Nineteen patients were male, and 55 female. The age when the diagnosis of APBJ or CBD was made ranged from 57 days to 80 years, with a mean of 33.3 years (Fig. 1). The mean age of the 11 patients without CBD was significantly higher than that of the 63 patients with CBD (54.5 vs 29.6 years, $P = 0.0009$).

Of the 63 patients with CBD, 33 had type I dilatation (male:female, 6:27; mean age, 29.4 years), 4 patients had type III (male:female, 3:1; mean age, 55.5 years), 24 patients had type IVa (male:female, 3:21; mean age, 25.6 years), and 2 patients had type IVb (all female; mean age, 30.0 years). There were no patients with type II or V. The 11 patients who had APBJ without CBD, 7 males and 4 females, had a mean age of 54.5 years (range, 17–67 years). This subgroup of patients was significantly older than those with type I ($P = 0.02$) or type IVa ($P = 0.003$). The male to female ratio of the patients with CBD was 12:51; this was significantly different from the 7:4 ratio of those without CBD ($P = 0.006$).

We were able to classify the type of CBD in 45 of 63 patients (71%) with CBD by ERCP alone. Of the remaining 18 patients, 8 had undergone choledochal cyst-jejunostomy previously and the biliary tree was poorly visualized by ERCP. In 4 patients, carcinomas or a stone occluded the bile duct. Three patients were children (2 were 2 months old, and 1 was 6 years old) and postural changes during ERCP were restricted due to intratracheal intubation for general anesthesia. Two patients had huge cysts and the intrahepatic biliary tree was not filled by ERCP. One patient had a history of severe post-ERCP pancreatitis (this had occurred in another hospital). The classification of CBD in these 18 patients was accomplished by percutaneous transhepatic cholangiography (PTC) or intraoperative cholangiography.

Figure 2 shows the proportion of each type of APBJ in each type of CBD. Classification of APBJ types was possible in 69 patients (93%), while technical difficulties caused classification failure in the other 5 patients. Overall, type a APBJ was demonstrated in

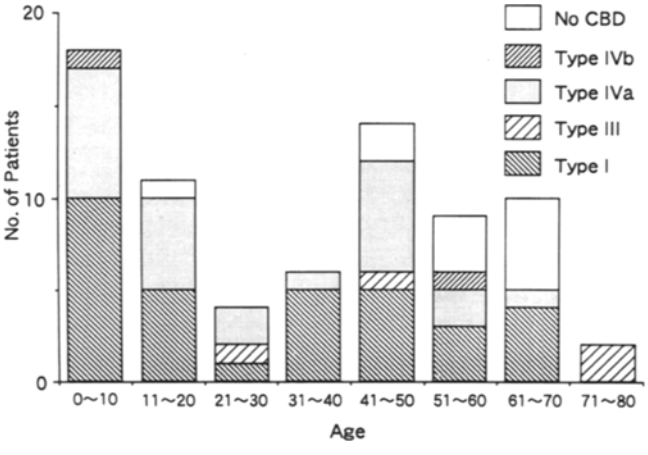

Fig. 1. Age distribution of 74 patients with anomalous pancreaticobiliary junction in relation to type of congenital biliary dilatation (*CBD*) according to the classification of Todani et al.[8] Patients with type I and IVa are widely distributed in almost all age groups. Note that patients with no CBD are in older age groups

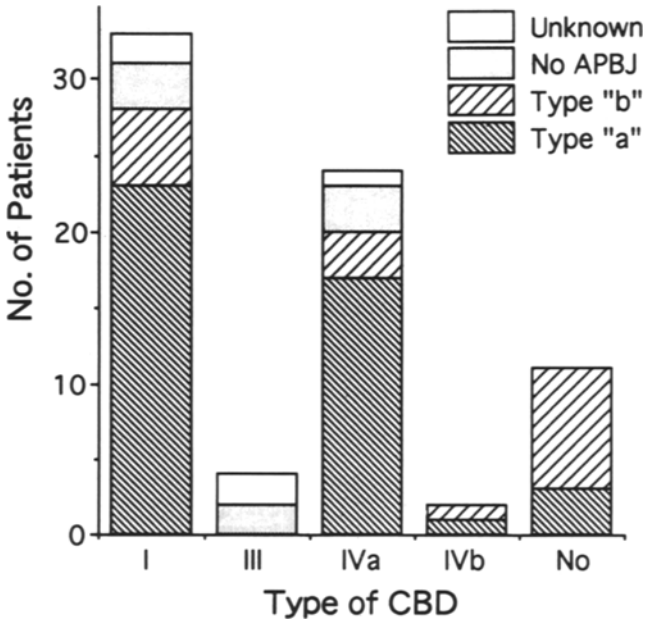

Fig. 2. Type of anomalous pancreaticobiliary junction (*APBJ*; classification of Komi et al.[12]) in each type of CBD. Type a preponderates over type b in the type I and type IVa subgroups. In contrast, type b is dominant in patients without CBD

44 patients, and type b in 17. Eight patients with CBD had no APBJ anomaly. The mean age was not significantly different between the two types of APBJ (31.2 years in type a, 38.2 years in type b). Females were preponderant in both types of APBJ (male to female ratio, 9:35 in type a and 6:11 in type b). The number of patients with type a APBJ was obviously greater than that with the type b anomaly in all CBD subgroups. In contrast, type b APBJ was more common than type a in patients without CBD. The type a to type b ratio was significantly different in patients with and without CBD ($P = 0.01$).

Twelve of the 74 patients had neoplastic lesions; 7 of them had CBD and 5 did not. The frequency of neoplasms in patients without CBD (45.5%) was significantly greater than that in patients with CBD (11.5%) ($P = 0.018$). Seven patients (3 with CBD and 4 without) had gallbladder carcinoma; 3 with CBD had cholangiocarcinoma; 1 patient without CBD had pancreatic carcinoma, and 1 patient with CBD had a carcinoid tumor. All of these patients, except for one with a carcinoid tumor, had the APBJ anomaly. The mean age of the patients with the neoplasms was 53.2 years; this was significantly higher than that of patients without neoplasms ($P = 0.001$).

All the neoplastic lesions but the carcinoid tumor were definitely diagnosed by ERCP. The diagnosis of the carcinoid tumor (maximum diameter 15 mm, height 6 mm) was made only by a macroscopic examination of the dilated bile duct after resection.

Nineteen patients (25.7%) had stones in the biliary tract. In 15 of these patients, the presence of the stones was diagnosed by ERCP. In the remaining 4, the biliary tract was poorly visualized by ERCP due to previous operations, obscuring the presence of the stones. Three patients had false positive ERCP results.

Discussion

This study revealed that ERC played an important role in the preoperative evaluation of CBD and APBJ. In our series of patients with these conditions, there was a preponderance of females and patients with type a APBJ among patients with CBD, while the reverse was true of patients without CBD. Moreover, the incidence of neoplastic lesions in association with APBJ was higher in patients without CBD than in those with CBD, being 45.5% and 11.5%, respectively.

Patients with type I and type IV CBD had a similar mean age and sex ratio. Most of these patients had type a APBJ. A Japanese nationwide survey supports our findings.[13] The type III CBD had peculiar characteristics compared with type I and type IV; patients with type III CBD were predominantly male and their mean age was higher than that of patients with type I and type IV. We could not demonstrate the presence of APBJ in the four patients with type III CBD in our series. These findings suggest that there might be some fundamental difference between type III and other types of CBD.

The 11 patients with APBJ but no CBD showed remarkable differences from those with CBD: (1) They were significantly older than those with CBD. (2) Male patients exceeded females in number, this being inconsistent with several previous studies, which reported female preponderance even in this subgroup.[1,14] The reason for this difference is not known. (3) Type b APBJ was more frequent than type a. (4) This type of anomaly was more frequently associated with neoplasms than APBJ with CBD. Unfortunately, we cannot diagnose this anomaly with high malignant potential by less invasive examinations such as ultrasonography and computed tomography. The presence of APBJ without CBD can be definitely demonstrated only by ERCP. In the 6 patients without neoplasms in our series, epigastric pain due to gallstones or chronic pancreatitis brought them to medical investigations. Otherwise, this subgroup of patients may not have sought medical advice despite their precancerous condition.

The existence of patients with CBD but no APBJ is noteworthy in regard to the etiology of biliary dilatation. Among this group there were three patients with type I CBD, two type III, and three type IVa. The main mechanism underlying biliary dilatation is thought to be reflux of the pancreatic juice into the bile duct due to the presence of APBJ.[3,11] In view of this hypothesis, these patients with a common channel of normal length may possibly have a sphincter mechanism below the pancreaticobiliary junction, thereby allowing the pancreatic juice to flow into the bile duct. Endoscopic biliary manometry[15] or endoscopic ultrasonography[16] would be of value to elucidate whether this is so.

Due to the high incidence of postoperative complications, and the possible development of carcinoma from the dilated bile duct, the operative method for the treatment of CBD has changed from internal drainage to total excision of the dilated extrahepatic bile duct, followed by bilio-enteric anastomosis.[8,9,10] The new surgical procedure is more complicated and requires much more information about the biliary tract and the pattern of the pancreaticobiliary junction, particularly to avoid injury to the pancreatic duct. The present study demonstrated that ERCP provided a satisfactory detailed view in most of the patients (93%). ERCP was employed safely in three 2-month-old babies and one 5-month-old baby in our series. The use of general anesthesia and a proper pediatric duo-

denoscope (model PJF; Olympus Optical Co., Tokyo Japan) was necessary.[17] If the duodenoscope and a practitioner with sufficient expertise are not available, intraoperative selective contact cholangiopancreatography is reported to be useful.[18] Endoscopic ultrasonography may be able to demonstrate the existence of APBJ,[16] but it would be of little help to clearly delineate APBJ for the safe performance of surgery.

Twelve patients in our series had neoplasms in the biliary tract or pancreas. All lesions but a very small carcinoid tumor in the bile duct were diagnosed by ERCP. This would suggest that even ERCP can provide sufficient information about neoplastic lesions only in patients with advanced cancers. Due to recent advances allowing the early detection of CBD, and thus, earlier surgery, we expect that we will be seeing younger patients and that the percentage of those with cancer will decrease.

Detection of stones in the biliary tract by ERCP was possible in 79% of our patients. Insufficient filling of the biliary tract due to previous bilio-enteric anastomosis caused the failure of detection. For more accurate diagnosis, other diagnostic modalities, such as ultrasonography and endoscopic ultrasonography, must be employed in combination with ERCP.

References

1. Kimura K, Ohto M, Saisho H, Unozawa T, Tsuchiya Y, Morita M, Ebara M, Matsutani S, Okuda K (1985) Association of gallbladder carcinoma and anomalous pancreaticobiliary ductal union. Gastroenterology 89:1258–1265
2. Komi N, Takehara H, Kunitomo K (1989) Choledochal cyst: Anomalous arrangement of the pancreaticobiliary ductal system and biliary malignancy. J Gastroenterol Hepatol 4:63–74
3. Miyano T, Suruga K, Suda K (1981) The choledocho-pancreatic long common channel disorders in relation to the etiology of congenital biliary dilatation and other biliary tract disease. Ann Acad Med 10:419–426
4. Komi N, Tamura T, Miyoshi Y, Hino M, Yada S, Kawahara H, Udaka H, Takehara H (1985) Histochemical and immunohistochemical studies on development of biliary carcinoma in 47 patients with choledochal cyst: Special reference to intestinal metaplasia in the biliary duct. Jpn J Surg 15:273–278
5. Mori K, Nagakawa T, Ohta T, Nakano T, Kayahara M, Kanno M, Ueno K, Izumi R, Miyazaki I (1993) Association between gallbladder cancer and anomalous union of the pancreaticobiliary ductal system. Hepatogastroenterology 40:56–60
6. Chijiiwa K, Tanaka M, Nakayama F (1993) Adenocarcinoma of the gallbladder associated with anomalous pancreaticobiliary ductal junction. Am Surg 59:430–434
7. Yamauchi S, Koga A, Matsumoto S, Tanaka M, Nakayama F (1987) Anomalous junction of pancreaticobiliary duct without congenital choledochal cyst: A possible risk factor for gallbladder cancer. Am J Gasroenterol 92:20–24
8. Todani T, Watanabe Y, Naruse M, Tabuchi K, Okajima K (1977) Congenital bile duct cysts: Classification, operative procedures, and review of 37 cases including cancer. Am J Surg 134:263–269
9. Ishida M, Tsuchida Y, Saito S, Hori T (1970) Primary excision of choledochal cysts. Surgery 68:884–888
10. Hata Y, Sasaki F, Takahashi H, Tamura M, Ohkawa Y, Saji Y, Kurauchi N, Manabe K, Uchino J (1993) Surgical treatment of congenital biliary dilatation associated with pancreatico-biliary maljunction. Surg Gynecol Obstet 176:581–587
11. Babbitt DP, Starshak RJ, Clemett AR (1973) Choledochal cyst: A concept of etiology. AJR 119:57–62
12. Komi N, Udaka H, Ikeda N, Kashiwagi Y (1977) Congenital dilatation of the biliary tract: New classification and study with particular reference to anomalous arrangement of the pancreaticobiliary ducts. Jpn J Gastroenterol 12:293–304
13. Aoki H, Sugaya H, Simazu M (1987) A clinical study on cancer of the bile duct associated with anomalous arrangements of pancreaticobiliary ductal system. Analysis of 569 cases collected in Japan (in Japanese). Tan to Sui (Biliary Tract and Pancreas) 8:1539–1551
14. Uchimura M, Waki S, Kida E, Kanda K, Narita K, Maeda S, Soejima H, Okamoto K, Ozaka K (1991) Anomalous arrangement of pancreaticobiliary ductal system and biliary carcinoma (in Japanese). Tan to Sui (Biliary Tract and Pancreas) 12: 397–404
15. Matsumoto S, Tanaka M, Ikeda S, Yoshimoto H (1991) Sphincter of Oddi motor activity in patients with anomalous pancreaticobiliary junction. Am J Gastroenterol 86:831–834
16. Mitake M, Nakazawa S, Naitoh Y, Kimoto E, Tsukamoto Y, Yamao K, Inui K (1991) Value of endoscopic ultrasonography in the detection of anomalous connections of the pancreatobiliary duct. Endoscopy 23:117–120
17. Shirai Z, Toriya H, Maeshiro K, Ikeda S (1993) The usefulness of endoscopic retrograde cholangiopancreatography in infants and small children. Am J Gastroenterol 88:536–541
18. Komi N, Takahara H, Miyamoto H, Hirai T, Kameoka K, Hino M (1982) Pathophysiology of anomalous arrangement of the pancreaticobiliary ductal system: Clinical point of view (in Japanese). J Clin Surg (Tokyo) 37:1775–1780