**Review**

**Variations of the Union between the Terminal Bile Duct and the Pancreatic Duct in Patients with Pancreaticobiliary Maljunction**

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**Abstract:** Background and Purpose:

Pancreaticobiliary maljunction (PBM) is an anomaly with an extramural location of the union between the bile duct and the pancreatic duct system in the duodenum, and is probably caused by disarrangement of the embryonic connection between the two duct systems that presents varied clinical manifestations. The aim of this study was to analyze and categorize various patterns of connection between the two duct systems in PBM patients in order to improve the precision of the diagnosis of PBM and to establish safe surgical procedures to treat it.

Materials and Methods:

We made group classifications of the patterns of connection between the two duct systems in 226 PBM patients based on radiologic or/and anatomical examinations.

Results:

The 226 cases of PBM were divided into three groups: Group A, 2 patients with a complicated connection pattern, Group B, 176 patients with a notch or a small duct segment distal to the terminal bile duct, Group C, 48 patients without a notch or the segment at the terminal bile duct.

Conclusion:

In Group A, the terminal bile duct was probably connected to one of the multi-branched main ducts of the ventral pancreas, while in Group B, it was probably connected to a second branch of the ventral pancreas, and in Group C, directly to the main pancreatic duct.

**Key words:** pancreaticobiliary maljunction, choledochal cyst, embryology

**INTRODUCTION**

The origin of a narrow duct segment distal to the enlarged common bile duct in congenital choledochal cysts is controversial. In 1936, Yotuyanagi described this duct system and termed it “ductus pancreaticobiliosis” based on the anatomical findings of autopsy cases. Bab- bitt detected the narrow duct system using direct cholangiography in 3 children with congenital cystic dilatation of the bile duct (CCBD), Alonso-Lej Type I) and described the narrow duct as a long common channel, defining it as an “anomalous arrangement of the pancreaticobiliary ductal system”. Both of those authors suspected that the narrow duct system was a pre-disposing factor in the occurrence of CCBD, and subsequently this hypothesis became well known throughout the world.

Since Babbitt’s report, further investigations of pancreaticobiliary maljunction (PBM) have been done. The Japanese Study Group on Pancreaticobiliary Maljunction presented extramural location of the union of the terminal bile
duct and the pancreatic duct in the duodenum as a definition of PBM. However, how PBM arises during embryogenesis remains unclear. Recently we proposed an hypothesis, based on radiological and anatomical analyses of PBM, that the narrow duct system was neither a narrowed bile duct nor a common channel of the two duct systems, but was rather part of the pancreatic duct system. That is, the development of PBM during embryogenesis is probably caused by disarrangement of the embryonic connection between the biliary and pancreatic ducts in an early period of gestation.

However, this connection may occur at various locations of the duct system of the ventral pancreas, and moreover, PBM is often accompanied by other malformations of the hepatobiliary system and pancreas such as CCBD, congenital stricture of the intrahepatic bile duct and pancreas divisum. Consequently, the connection pattern between the two duct systems may be extremely complicated, and making a precise diagnosis of PBM will continue to be very difficult in PBM patients with a short common duct if the embryo genetic etiology of PBM is not clarified.

The aim of this study was to analyze the anatomical variations in the location of the union and in the patterns of connection of the terminal bile duct with the pancreatic duct system in patients with PBM, in order to improve the precision of the diagnosis of PBM and to establish safe surgical procedures. We believe that making a precise diagnosis of PBM, which carries a high risk of bile duct cancer, is important for preventing the development of bile duct cancer.

**Patients and Methods**

**Patients**

Of 2,613 patients who underwent ERCP in our institute, 226 had PBM. The 226 PBM patients consisted of 70 males and 156 females, 158 of whom had benign biliary or/and pancreatic diseases and 68 who had a malignant tumor of the biliary system, while 146 patients had CCBD and 80 did not. The age of the 226 patients ranged from 6 to 81 years and the mean age of the patients with benign biliary disease was 60 + 11 years in 54 males and 53 + 12 years in 104 females. The mean age of the PBM patients with biliary malignancy was 66 + 13 years in 16 males and 63 + 12 years in 52 females.

**Diagnostic Criteria and PBM Detection**

The Japanese Study Group on Pancreaticobiliary Maljunction has defined PBM as a congenital anomaly in which the junction of the pancreatic and biliary ducts is radiologically and/or anatomically detected outside of the duodenal wall (extramural location). We made a diagnosis of PBM by means of ERCP, PTC, and/or anatomical confirmation based on operative findings during transduodenal papilloplasty or examination of pancreateoduodenectomized specimens or autopsy materials. Radiological detection of the extramural location of the union of the two ducts in the duodenum was performed according to the methods described previously.

**Results**

The 226 PBM patients were classified into three groups according to whether or not they had the following characteristics: 1) a compli-
cated connection of the terminal bile duct or the cystic dilatation of the bile duct with the pancreatic duct system, 2) evidence of a notch or a small duct distal to the bile duct (narrow duct segment), and 3) no evidence of a notch or small duct at the union of the bile duct with the ventral pancreas duct on direct cholangiopancreatograms. Here “notch” means a V-shaped indentation on the terminal bile duct near the union of the bile duct and the main pancreatic duct seen on the cholangiopancreatogram, and “a small duct” means a much narrower duct distal to the terminal bile duct or to the biliary cyst. PBM was frequently associated with pancreas divisum, especially partial pancreas divisum in which there was only communication between a small branch of the ventral pancreas and one of the dorsal pancreas. We encountered no case in which there was a union of the bile duct and the dorsal pancreatic duct.

1. Group A: PBM patients in whom the connection of the terminal bile duct (or the bottom of the biliary cyst) with the pancreatic duct showed a complicated pattern. Two of the 226 patients were classified into this group. In these patients, ERCP revealed multi-branching of the main pancreatic duct of the ventral pancreas and communication of the biliary cyst with one of the branches, as shown in Fig. 1a and 1b.

2. Group B: PBM patients in whom a notch or a small duct distal to the terminal bile duct was detected at the point of union between the two duct systems. Of the 226 patients, 176 were classified into this group. Direct cholangiograms of patients in

Fig. 1. ERCP of a 37-year-old woman with Group A PBM, CCBD, pancreas divisum, and a biliary cyst, discovered by AUS in a mass survey screening.

a) The terminal bile duct can be seen to join with a pancreatic duct (long thin arrow). However, the main pancreatic duct branches away into at least three branches, and one of them is connected with the biliary cyst at the bottom of the cyst.

b) The dorsal pancreatic duct (arrow) is visualized by the flow of contrast medium through the major papilla. These images show multi-branching of the main duct of the ventral pancreas and the connection between the ventral and the dorsal pancreatic duct (partial pancreas divisum).
this group showed a notch or a small duct segment in the distal portion of the terminal bile duct (or the biliary cyst). In Fig. 2, a T-tube cholangiogram shows a notch in the distal portion of the terminal bile duct. The terminal bile duct is seen to be suddenly reduced in diameter, showing an irregular shape at the distal end and connecting with the small radicle arising from the main pancreatic duct. This small radicle was a second branch of the ventral pancreatic duct.

Fig. 2. T-tube cholangiogram of a 31-year-old woman with Group B PBM, after cholecystectomy.

The terminal bile duct is seen to join with a small radicle (2 small thin arrows) arising from the main pancreatic duct (arrow) near the duodenal wall and a notch is formed in the distal portion of the bile duct. The terminal bile duct is seen to be suddenly reduced in diameter, showing an irregular shape at the distal end and connecting with the small radicle arising from the main pancreatic duct. This small radicle was a second branch of the ventral pancreatic duct.

Fig. 3. ERCP of a 65-year-old woman with Group B PBM, CCBD and gallbladder cancer.

A narrowed duct segment (2 small thin arrows) distal to the biliary cyst can be seen. This segment is joined with the main pancreatic duct (long thin arrow). A small radicle (arrowhead) arising from the narrow duct segment can be seen. The narrowed duct segment is thought to be a second branch of the ventral pancreatic duct.

This type of connection is considered to indicate that the bile duct is anotomically joined with the main duct of the ventral pancreas, accompanied by an interposition of the second or third branch of the ventral pancreatic duct between the two ducts.

3. Group C: PBM patients in whom a notch or resection of a biliary cyst in a case of CCBD. The main pancreatic duct appears to enter a large U-shaped duct, which seems to be the intrapancreatic portion of the common bile duct. But the U-shaped duct is considered to correspond to the “common duct” which occurs in PBM patients and consists of a narrow duct segment and part of the main pancreatic duct. This type of connection is considered to indicate that the bile duct is anotomically joined with the main duct of the ventral pancreas, accompanied by an interposition of the second or third branch of the ventral pancreatic duct between the two ducts.
a small duct distal to the terminal bile duct was not detected at the union of the two ducts.

Group C consisted of 48 patients with PBM. An ERCP (Fig. 5) of a representative PBM patient in this group demonstrated that the terminal bile duct was joined with the main pancreatic duct near the duodenal wall, without a notch or a small duct on the terminal bile duct. Figure 6 shows that the terminal bile duct was
Fig. 7. ERCP of a 55-year-old woman with Group C PBM, partial pancreas divisum and gallbladder cancer. The whole extrahepatic bile duct, part of the intrahepatic ducts, and the dorsal pancreatic duct (via the small duct indicated by a small arrow) were revealed by the flow of contrast medium through the major papilla. The terminal bile duct was directly joined (arrow) with the proximal end of the main duct of the ventral pancreas and a small pancreatic radicle (the high density line is indicated by a small arrow) was connected with the large duct from the major papilla (ventral pancreatic duct) and the dorsal pancreatic duct from the minor papilla to the pancreatic tail (partial pancreas divisum). Small pancreatic radicles (arrowhead) arose from the large duct from the major papilla, which is thought to be the main duct of the ventral pancreas.
joined with the main pancreatic duct far from the duodenal wall without a notch or a small duct segment. Figure 7 shows that the extrahepatic bile duct and the dorsal pancreatic duct via a small duct arising from the main duct of the ventral pancreas (partial pancreas divisum\(^7,8\)). In this case, the terminal bile duct was considered to be joined directly with the proximal end of the main duct of the ventral pancreas.

**DISCUSSION**

Extramural location of the union of the terminal bile duct and the pancreatic duct in the duodenum was not recognized in the past as an anomaly in the pancreaticobiliary ductal system, but was rather thought to be a normal variations\(^9,12-14\). However, in several reports that described detailed analyses of the location of the junction of the terminal bile duct and the pancreatic duct based on anatomical and/or radiological examinations, a long common channel\(^15\) or extramural location of the union in the duodenum\(^15-17\) was not found in normal cases, but was found in cases of CCBD, congenital biliary atresia (CBA)\(^18\) or acute pancreatitis\(^12,19\).

Since Babbitt’s key report about PBM\(^2\), our knowledge about “a long common channel” has increased, and diagnostic criteria for PBM were presented by the Japanese Study Group on Pancreaticobiliary Maljunction\(^4\). However, the mechanism of embryonic development of PBM was not clarified at that time. We hope that our hypothesis about the mechanism of PBM development will aid progress in making a precise diagnosis of PBM and the establishment of safe surgical procedures for PBM. In this report, we presented various cholangiopancreatograms of PBM patients with various locations or/and anomalous connections due to an embryonic disarrangement between the bile duct and the pancreatic duct, and investigated the essential patterns of the disarrangement with the aim of establishing safe and appropriate surgical procedures for this disease. Moreover, a notch or a small duct segment was noted to be a key point of detecting PBM. We showed that the anatomical location of the union could exist at any site in the duct system of the ventral pancreas. In our series, 136 of the 226 PBM patients had a short common duct less than 1 cm in length, and these 136 patients consisted of 113 patients in Group B and 23 in Group C. The remaining 90 of the 226 patients had a long common duct over 1 cm in length, and they consisted of the 2 patients in Group A, 63 patients in Group B and 25 patients in Group C. A short common duct was apparently formed due to the location of the union near the duodenal wall in Group C patients and also in patients with a notch (not a small duct) in Group B. On the other hand, a long common duct was seen in Group B patients with a small duct segment and in Group C patients with the location of the union far from the duodenal wall, and also in Group A patients. Figure 8 shows a schematic diagram showing the possible anatomical points of the union.

Clinical manifestations in PBM patients, such as acute pancreatitis\(^12,19\), intermittent abdominal pain, obstructive jaundice, acute cholangitis, primary bile duct stones\(^20\) and bile duct cancer\(^21,22\) did not correlate with the presence of a long or a short common duct, with accompanying CCBD\(^5\) or the connection patterns. These clinical features do not correlate with the length of the common duct in PBM patients, so it is difficult to detect the extramural location of the union in the duodenum, especially in patients with a short common duct.

Concerning the relationship between PBM
and the development of gallbladder cancer, PBM was found in 45 of 112 patients with gallbladder cancer treated in our institute during the study period. Since PBM is considered to be a high risk condition for the development of gallbladder cancer, we make it a rule that PBM patients receive cholecystectomy as soon as possible even if the patients have no clinical manifestations, in order to prevent the development of gallbladder cancer.

Moreover, since CCBD with PBM is also known to be associated with a high-risk condition for the development of bile duct cancer, preventive surgery is recommended when the disease is discovered. It is well known that an extirpation of the biliary cyst is the most appropriate procedure, but details of the optimal approach for the intrapancreatic portion of the biliary cyst, as well as those for the proximal portion of the cyst, remain controversial. It is important to establish a practical and safe procedure for treatment of the anomaly with the goals of avoiding intraoperative injury of the main pancreatic duct and preventing the development of cancer originating from a remnant cyst wall. The classification of the patterns of connection between the bile duct and the pancreatic duct into the groups described here should be helpful for the optimization of the surgical approach. A complete resection of the bile duct or the biliary cyst is possible only for patients with a small duct segment in Groups A and B. Resection of the terminal bile duct or the biliary cyst in patients with a notch in Group B and in patients in Group C must be performed in a manner that retains part of the terminal bile duct in order to avoid injury of the main pancreatic duct.

REFERENCES

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