J Hep Bil Pancr Surg (1994) 1:219–221

Journal of HBP Surgery

Special topics

Diagnostic criteria of pancreaticobiliary maljunction

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THE COMMITTEE OF JSPBM FOR DIAGNOSTIC CRITERIA[1]*

Key words: pancreaticobiliary maljunction, diagnostic criteria, junction outside duodenal wall

Introduction

Pancreaticobiliary maljunction first reported by Kozumi and Kodama[1] in 1916, did not attract the attention of pathologists and physicians for many years. In Japan, this condition is now regarded as the possible source of the pathogenesis and carcinogenesis of choledochal cysts, since Komi and colleagues[2] introduced Babbitt's 1969 concept based on the anomalous arrangement of the pancreaticobiliary ductal system that was frequently observed in patients with choledochal cyst.[3,4] However, the diagnostic criteria, terms used, and classification of the pancreaticobiliary maljunction are extremely controversial. In response to the continuing debate on this subject, the Japanese Study Group on Pancreaticobiliary Maljunction (JSPBM) and the Committee for Diagnostic Criteria were established in 1983 by surgeons, pediatric surgeons, physicians, and pathologists interested in this subject. The committee published the proposed diagnostic criteria of the maljunction in 1987 after extensive discussions, and revised these criteria in 1990, as follows.

Offprint requests to: T. Todani
Received for publication on Oct. 27, 1993; accepted on Dec. 8, 1993

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Definition

Pancreaticobiliary maljunction (anomalous arrangement of the pancreaticobiliary ductal system) is a congenital anomaly defined as a union of the pancreatic and biliary ducts that is located outside the duodenal wall.

Pathophysiology

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 (sphincter of Oddi) does not functionally affect the union. Accordingly, various pathological conditions occur in the biliary tract and the pancreas.

Note 1 *Other complicated abnormal connections between the pancreatic and common bile ducts in the extraduodenal region are included in this category.*

Note 2 *Apparently abnormal connections caused by acquired factors such as tumors, cholelithiasis, and papillitis are excluded.*
Note 3 *The pathological conditions may be termed "maljunction syndrome." However, both the naming and the solution of the relationship between the maljunction and diseases of the biliary tract (cholangitis, bile duct dilatation, cholelithiasis, and biliary cancer) and the pancreas (acute pancreatitis, chronic pancreatitis, pancreatolithiasis, and pancreatic neoplasms) are topics for future research.*

Diagnostic criteria

At present, the maljunction may be diagnosed by either radiographic or anatomical examination.

Radiological diagnosis

The lack of effect of the sphincter of Oddi on the pancreaticobiliary ductal junction should be verified. When the sphincteric action cannot be determined, the following findings must be confirmed by endoscopic retrograde cholangiopancreatography (ERCP), percutaneous transhepatic cholangiography (PTC), or operative cholangiography:

(a) The pancreatic duct and choledochus connect with an obviously long common channel, or
(b) They unite in an apparently anomalous form.

Note 1 *Functional observation of the ductal union by cine cholangiography, biliary manometry, and cholangioscopy, including percutaneous transhepatic cholangioscopy, is useful for determining whether or not the sphincteric action is affecting the pancreaticobiliary junction.*
Note 2 *The length of the common channel is not included in the diagnostic criteria for the maljunction because various factors such as the form of the union, the direction, the magnification of the film, and the age and stature of the patient must be considered.*

Anatomical diagnosis

The pancreaticobiliary junction should be confirmed by surgery or autopsy to determine whether or not it exists outside the duodenum, and whether or not its shape is anomalous.

Supplementary diagnosis

The following findings strongly suggest the existence of a pancreaticobiliary maljunction.

Elevated amylase levels in bile

Pancreatic enzymes, especially amylase, in the bile within the bile duct and gallbladder obtained percutaneously or immediately after laparotomy are generally at extremely high levels. However, levels close to or below the normal serum value are occasionally observed in patients with maljunction, and temporarily high amylase levels in the bile are found in some patients with normal pancreaticobiliary junctions.

Extrahepatic ductal dilatation

Maljunction is usually found in patients with so-called choledochal cysts. When cystic, fusiform, or cylindrical dilatation is detected at the extrahepatic bile duct, careful investigations should be made to determine whether pancreaticobiliary maljunction is present. In contrast, however, maljunction is occasionally seen in patients without bile duct dilatation.

Comments on "Diagnostic Criteria of Pancreaticobiliary Maljunction (PBM)" (Takuji Todani)

The pancreaticobiliary maljunction indicates an anatomical abnormality showing extraduodenal union, and not a functionally abnormal junction. This can be commonly observed in patients with congenital choledochal cyst, especially type I, showing cystic or diffuse dilatation, and type IV, in which there is intrahepatic involvement. However, type II, indicating diverticulum, type III, showing choledochocele, and type V, indicating intrahepatic ductal dilatation alone, are not usually associated with the maljunction, and show a normal pancreaticobiliary junction.

Pancreaticobiliary maljunction is occasionally observed in infants and children with such hepatobiliary disorders as biliary atresia, biliary hypoplasia, and infantile hepatitis. Moreover, carcinoma of the gallbladder in young adults who have the maljunction without bile duct dilatation has recently been reported with increasing frequency in Japan.

Although the term "long or elongated common channel," implying that the union is located outside the duodenum, is occasionally preferred to the term "pancreaticobiliary maljunction", the normal length of the common channel generally tends to be less than 10.0 mm in adults and 4.0 mm in infants.[4] Therefore, measurement of the length of the common channel is probably inadequate because of variations in patients' ages, and physical constitution, and also because of differences in the magnification of X-ray films. Pan-

type a	type b	type c
C-P type	P-C type	complex type
choledochal type	pancreatic type	complex type
right angle type	acute angle type	complex type
type II	type III	type I
type I	type II	type III
subtype a (type I)	subtype b (type I)	type II

Fig. 1. Classification of pancreaticobiliary maljunction (see text for definitions)

creaticobiliary maljunction does not mean a long common channel.[5] Short common channels are rarely observed in choledochal cyst, and probably make it difficult to determine whether there is a maljunction.[6] An accurate diagnosis of maljunction require a manometric study of the junction in the case of a doubtful extraduodenal union, although high amylase levels in bile imply the presence of pancreaticobiliary maljunction.

Dilated common channels with a protein plug are often observed in choledochal cyst with maljunction. Dilatated common channel possibly has to do with the development of protein plugs in the pancreatic duct and chronic pancreatitis with pancreatolithiasis.[7]

The term "maljunction syndrome" may not exactly represent the present conditions and signs, since it includes various pathologic conditions of the liver, biliary tract, and pancreas. "Long common channel syndrome"[8–10] also seems to be unsatisfactory for the same reason. The term "syndrome" might be inadequate at present.

Cystic dilatation of the common bile duct cannot be produced by experimentally creating a maljunction and the maljunction itself may not be the primary cause of congenital cystic dilatation of the bile duct. However, the length and grade of strictures in the terminal bile duct seem to determine the type of choledochal dilatation.[11]

The maljunction has been classified by various authors according to the types of confluence between the terminal choledochus and the pancreatic duct, and the classification appears to be divided into three types (Fig. 1). However, several expressions are used for the same type; a uniform definition for the terms describing the types of maljunction has not yet been established. "Type a," "C-P," and "right angle," as shown in Fig. 1, are all used to indicate that the common bile duct is likely to join the pancreatic duct, this type is usually observed in patients with a cystic dilatation of the choledochus. Whereas "type b," "P-C," and "acute angle" all indicate that the pancreatic duct seems to join the choledochus; this type is often seen in patients with diffuse or cylindrical dilatation of the choledochus. "Types c, I, II, and III" imply complicated union of the pancreaticobiliary ductal system. These various terms for the same type should be standardized as soon as possible to prevent confusion.

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