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Review article

Congenital choledochal dilatation: Classification, clinical features, and long-term results

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Abstract: Choledochal cyst is generally accompanied by pancreatobiliary malunion. Pancreatic juice usually refluxes into the bile duct via the malunion, and bile juice occasionally regurgitates into the pancreatic duct. As a result, various pathological conditions occur in the biliary tract, pancreas, and liver. We have revised our classification of choledochal cyst on the basis of malunion. Abdominal mass, pain, and jaundice are regarded as the triad of choledochal cysts, but symptoms are quite non-specific. In neonates and infants less than 12 months of age, choledochal cyst is usually huge and no cylindrical dilatation is observed. Vomiting and jaundice with acholic stool are often observed. No hyperamylasemia is seen, despite the presence of a malunion. Whereas children over 13 months of age tend to show diffuse dilatation, and an abdominal mass is rarely palpable. Epigastralgia with hyperamylasemia, fever, vomiting, and slight jaundice are often observed. Cyst excision is the treatment of choice to prevent the development of ascending cholangitis and cancer in the cyst. Late complications of excisional surgery include recurrent cholangitis, pancreatitis, and, rarely, biliary malignancy. These complications usually develop 5 years or more after initial surgery and become the object of reoperation. Complete removal of the extrahepatic bile duct from the hepatic hilum to the intrapancreatic duct and a hilar hepaticoenterostomy with wide anastomosis after ductoplasty, including the coexisting primary stricture, are essential at the primary surgery. Provided a large anastomosis, favorable results can be obtained with either duodenostomy or Roux-Y jejunostomy. Surgical enlargement of the hilar ducts and careful lifelong follow-up should be routine in all patients who have undergone cyst excision. When ductal stricture with intrahepatic gallstones is confirmed, reoperation at an early stage is indicated to obtain a good quality of life with short hospitalization.

Key words: choledochal cyst, classification, symptoms, long-term result

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Introduction

Congenital choledochal dilatation (choledochal cyst) is a congenital abnormality of the intra- and extrahepatic ducts characterized by cystic or diffuse dilatation of the choledochus.[1,2] It shows a female predominance, and a higher prevalence in Orientals. It occurs in both children and adults, and more than half are seen within the first decade of life. Choledochal cyst is usually associated with pancreatobiliary malunion and has a potential danger of leading to biliary malignancy.[3] Therefore, lately it has been regarded as one component in the spectrum of pancreatobiliary malunion.[4] For the treatment of choledochal cysts, therefore, primary cyst excision with biliary reconstruction has recently become the procedure of choice to interrupt malunion and to prevent various complications.[1,5–8] The postoperative course of excisional surgery is smooth, and the prognosis is usually excellent. However, postoperative complications occasionally develop in the early and late stages. In this paper, the classification, clinical features, and long-term results of excisional procedures in choledochal cyst will be discussed.

Classification of choledochal cysts

Alonso-Lej et al.[9] classified choledochal cyst into three types in 1959. We refined this into six types, due to the recognition of intrahepatic involvement, in 1977,[1] but this classification does not include the concept of pancreatobiliary malunion. Types Ia, Ic, and IV-A of our classification are often observed, and are generally accompanied by pancreatobiliary malunion. Types Ib, II, III, IV-B, and V, however, are much less frequent and a malunion is rarely observed. Therefore, we again revised the classification on the basis of pancreatobiliary malunion (Fig. 1). The revised classification follows.

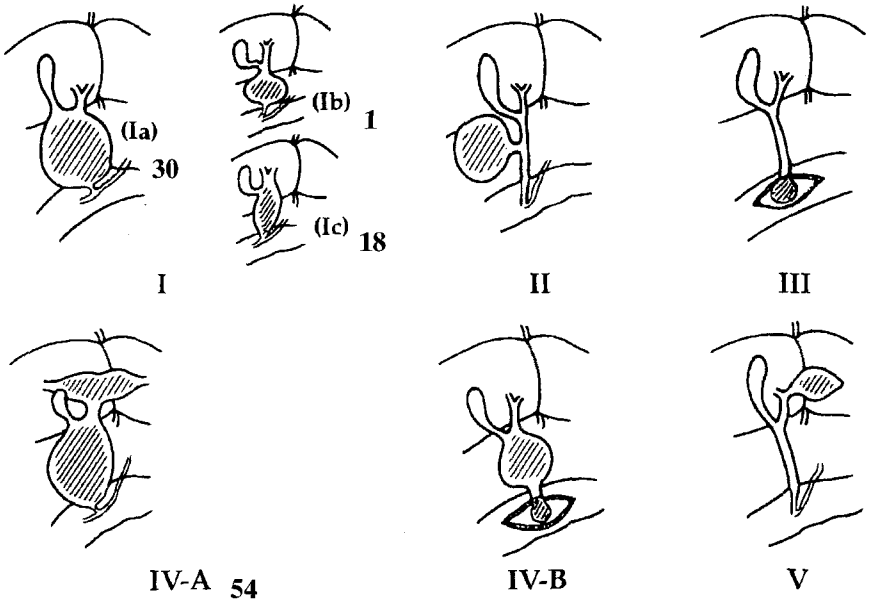

Fig. 1. Revised classification of choledochal cyst with pancreatobiliary malunion. *Numbers* indicate patients undergoing cyst excision in our series

Type I

Type I is choledochal dilatation alone, and the common hepatic duct proximal to the cyst is usually normal. It is divided into the following three subtypes; type Ia showing cystic dilatation with pancreatobiliary malunion, type Ib showing segmental dilatation without malunion, and type Ic showing diffuse dilatation with malunion.

Type II

Type II shows diverticular dilatation of the bile duct without malunion, and similar cysts are occasionally observed in any part of the extrahepatic bile duct.

Type III

Type III is a choledochocele located in the duodenal wall, and is not associated with malunion.

Type IV-A

Type IV-A is multiple cysts containing both intra- and extrahepatic bile duct cysts, and is usually accompanied by malunion. The incidence of type IV-A cyst among choledochal cysts is nearly 50%, higher than previously thought. Abdominal pain and fever are more frequent than in patients with type I, and a mass is often not palpable.[2] Cystic dilatation of the intrahepatic duct may be congenital, whereas diffuse dilatation probably develops secondarily, because the size is reduced after surgery.[2]

Type IV-B

Type IV-B is also multiple cysts, confined to the extrahepatic bile duct alone. The presence of malunion, however, still remains uncertain due to the lack of data.

Type V

Type V cyst shows single or multiple dilatations of the intrahepatic duct, and usually does not accompany malunion. It causes biliary infection and bile stasis resulting in intrahepatic gallstones or hepatic cirrhosis. Some may belong to Caroli's disease.

Pancreatobiliary malunion

Pancreatobiliary malunion located outside the duodenum is often observed in types Ia, Ic, and IV-A cysts, but types II, III, and V cysts are usually not accompanied by malunion.[3,10] Recently, gallbladder cancer has been detected with increasing frequency in patients with the pancreatobiliary malunion with slight or no biliary dilatation.[11]

The common channel in the malunion is simply classified into three types: non-dilated, dilated, and complex (Fig. 2). A dilated long common channel is found in one-third of the malunions, and probably causes protein plugs or stones, leading to the future development of pancreatitis.[12,13]

Malunion causes two-way regurgitation: pancreatic juice refluxes into the bile duct, or bile juice regurgitates into the pancreatic duct, and various pathological con-

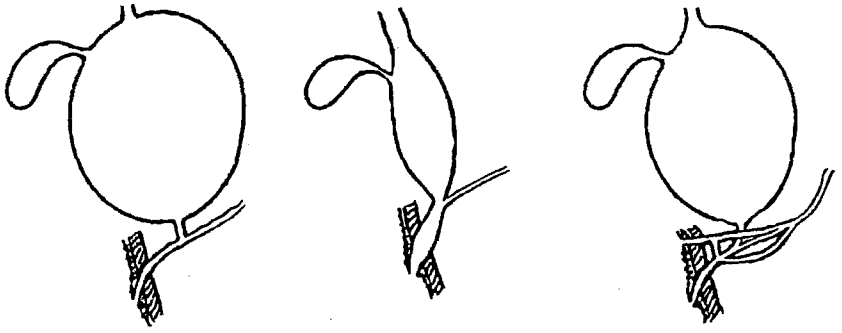

Fig. 2. Classification of the common channel in pancreatobiliary malunion

ditions occur in the biliary tract (biliary dilatation, cholangitis, biliary perforation, and biliary malignancy), the pancreas (pancreatitis and pancreatic cancer), and the liver (intrahepatic abscess, cirrhosis, and hepatic rickets).[3,4,14–16]

Clinical features of choledochal cyst

Abdominal mass, pain, and jaundice have been regarded as the classical triad of choledochal cysts, but presenting symptoms are usually quite non-specific. Epigastralgia is recurrent over several months, and slight jaundice and hyperamylasemia are often observed. In neonates and infants less than 12 months of age, however, the clinical features are often entirely different from those in children over 13 months old and young adults.[17,18]

Infants under 12 months of age

Infants under the age of 12 months usually have a large choledochal cyst, and some are accompanied by an intrahepatic component. No cylindrical dilatation of the choledochus is observed, but exceptionally, biliary perforation may lead to cylindrical dilatation due to shrinkage of the cyst. Vomiting caused by compression of the gastrointestinal tract and jaundice with acholic stools are frequent.

No hyperamylasemia is seen despite the presence of a malunion. Serum amylase shows low levels due to physiologically incomplete growth of pancreatic acinar glands. Pancreatic amylase level gradually increases with age, and usually reaches normal values in children over 1 year of age. Therefore, amylase in bile shows a low level in infants, while other pancreatic enzymes in bile, such as trypsin, elastase I, and lipase, show abnormally high levels.[17] For the proof of a reflux, enzymes other than amylase in bile should be examined.

Children over 13 months of age and adolescents

Children over 13 months of age and adolescents tend to show diffuse or cylindrical dilatation, and an abdominal mass is rarely palpable.[17,18] Ultrasonography performed at the time of abdominal pain or discomfort, however, may detect biliary dilatation. Symptoms are frequently non-specific, but epigastralgia with hyperamylasemia, fever, vomiting, and slight jaundice, as in patients with acute pancreatitis, are often observed.

Surgical treatment (excisional procedure)

Primary cyst excision and biliary reconstruction is the treatment of choice for preventing complications and avoiding the reflux of pancreatic juice by interrupting the malunion.[1,3,6] Previous internal cyst drainage should be converted to cyst excision as early as possible. In biliary reconstruction, Roux-Y hepaticojejunostomy has been widely and conventionally employed, but it does not always protect against cholangitis.

The free drainage of bile, rather than the reconstruction method employed, is the essential factor for preventing ascending cholangitis. This can be achieved only by a wide anastomosis performed at the hepatic hilum. Type IV-A cyst requires more selective management for the intrahepatic component. Plasty of the intrahepatic ductal stricture, to achieve a wide anastomosis, is necessary, and hepatic segmentectomy to remove the complicated lobe is occasionally feasible.

Biliary reconstructions used commonly are classified into two types (Fig. 3):

Hepaticoduodenostomy

Distal hepaticoduodenostomy (performed below the hilum) tends to cause anastomotic stricture, resulting in ascending cholangitis. Hilar hepaticoduodenostomy (performed at the hilum) with wide anastomosis[19] is our preferred procedure because of the physiological state, simplicity, and rarity of complications.

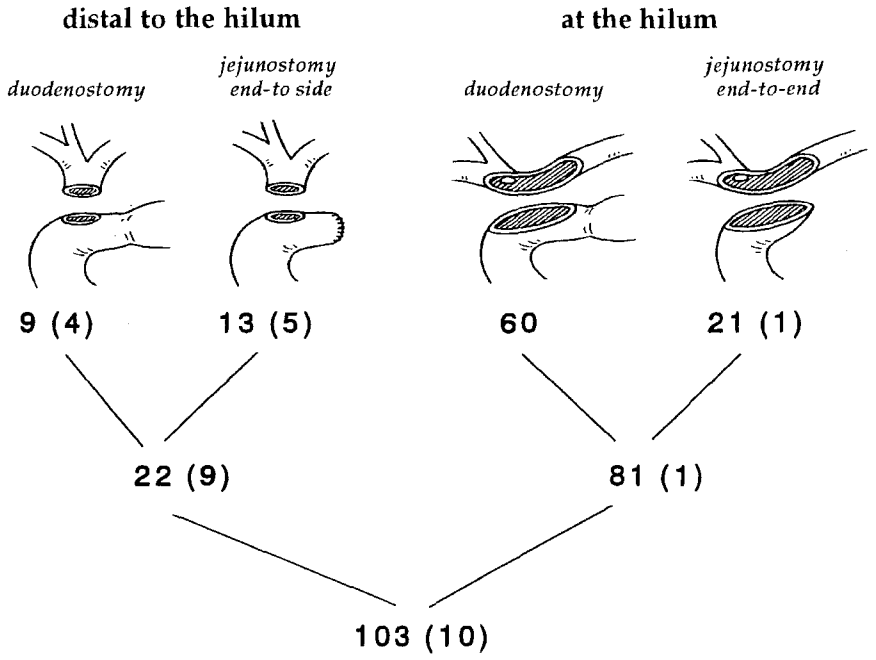

Fig. 3. Cyst excision and biliary reconstructions performed in 103 patients from 1969 to 1996. *Numbers* indicate the number of patients; *parentheses* indicate the number who underwent reoperation

Roux-Y hepaticojejunostomy

Distal hepaticojejunostomy with end-to-side anastomosis is generally done to adapt to the size of the common hepatic duct. However, it may be unsuitable because the common hepatic duct often shows a relatively small caliber, receives poor blood supply, and has denuded mucosa with fibrosis, resulting in stricture.[20] End-to-end anastomosis at the hilum should be performed after hilar ductoplasty to fit the jejunal stoma.[20]

Results of excisional surgery

The postoperative course of excisional surgery is usually smooth, and the prognosis is excellent. Since 1969, we have employed cyst excision with no operative death in 103 patients without cancer. Ages ranged from 27 days to 37 years and 97 patients underwent the excision as a primary surgery and 6 as a secondary procedure for previous internal drainage. The first 22 patients, until 1975, underwent distal hepaticoenterostomy, and the subsequent 81 received hilar hepaticoenterostomy to provide a wider anastomosis. Postoperative complications occasionally developed in the early and late stages, as described below.

Early complications

Minor leakage of the anastomosis.
This is effectively managed by Penrose or silicon drains placed around the anastomosis.[20] Continuous nasogastric decompression and long-term parenteral nutrition are also necessary.

Bleeding from the dissected layer.
Careful electrocoagulation and several purse-string sutures in the dissected outer layer around the pancreatic head are effective to avoid bleeding and pancreatic fistula.[20]

Gastrointestinal bleeding.
Peptic ulcer occurs in patients who have undergone Roux-Y hepaticojejunostomy, and is treated successfully with H2 blockers.

Acute pancreatitis and pancreatic fistula.
These complications are caused by injury of the pancreatic duct or tissue at the time of dissection of the terminal bile duct. Continuous nasogastric decompression and parenteral nutrition for several weeks are mandatory.[20]

Intestinal obstruction.
In patients with Roux-Y jejunostomy, adhesive ileus tends to occur more often compared the rate in patients with duodenostomy, because jejunostomy involves manipulation of the abdominal cavity below the transverse mesocolon.[19] Nasogastric tube decompression for several days is occasionally effective. Otherwise, relaparotomy is required.

A transient duodenal obstruction may occur in hilar hepaticoduodenostomy due to kinking of the mobilized duodenum caused by lifting it up to the hilum.[19,20] Transduodenal tube feeding is necessary for a week.

Table 1. Complications after excisional surgery

Early complications
Anastomotic leakage
Bleeding from dissected outer layer
Gastrointestinal bleeding
Acute pancreatitis
Pancreatic fistula
Intestinal obstruction
Late complications
Cholangitis and/or biliary calculi
Biliary carcinoma
Pancreatitis
Pancreatic carcinoma

Late complications: Long-term results

Cyst excision usually provides excellent long-term results, but late complications, such as recurrent cholangitis, cholelithiasis, biliary cancer, pancreatitis, or pancreatolithiasis occasionally develop several years after surgery (Table 1). The number of patients with long-term complications has been increasing as the number of patients undergoing cyst excision with long-term follow-up increases. Therefore, lifelong follow-up is necessary to avoid or manage potential problems of the liver, bile duct, and pancreas.[21–23]

In our 103 patients, the follow-up period ranged from 7 months to 28 years. Ten patients (9.7%) required reoperation 3–21 years (mean: 9.1 years) after cyst excision, due to recurrent cholangitis caused by anastomotic and primary ductal strictures, as described below.[22]

Recurrent cholangitis.
Postoperative cholangitis associated with abdominal pain and high fever is the most serious complication in biliary reconstruction, and often requires reoperation several years after surgery. It leads to intrahepatic stones, particularly in type IV-A cyst,[8,22] and occasionally results in biliary cirrhosis, hepatic failure, or biliary cancer, with fatal consequences.

Antibiotics and percutaneous transhepatic cholangiodrainage with bougie may be effective temporarily for complicated intrahepatic stones.[24,25] Complete removal of stones by percutaneous transhepatic cholangioscope often demands a long hospital stay and frequently has an unsatisfactory outcome.[26] Reoperation should be performed at an early stage to improve the quality of life with short hospitalization.

Recurrent cholangitis and stones result from bile stasis, caused mainly by anastomotic stricture or intestinal obstruction, and occasionally by primary intrahepatic ductal stricture.

Anastomotic stricture. Distal hepaticoenterostomy tends to produce anastomotic stricture,[20,27] especially in type IV-A cyst, because the stoma of the common hepatic duct often shows relative stricture, the duct being smaller than the size of the intrahepatic ductal dilatation. Anastomotic stricture disturbs free bile drainage from the intrahepatic duct, and causes bile stasis. Reoperation is often required 5 years or more after the initial surgery.[22] Hilar hepatico-enterostomy, therefore, should be primarily performed to achieve a wide stoma.[20,26] No significant difference has been observed in the occurrence rate of recurrent cholangitis between hilar duodenostomy and Roux-Y jejunostomy.[20,22]

Primary stricture. Primary stricture in the hilar or intrahepatic bile ducts is occasionally found in the bifurcation of the hilar and intrahepatic ducts,[20,28,29] and causes intrahepatic ductal dilatation and stones. Plasty of strictures around the hilum or umbilical portion to obtain a large anastomosis can be achieved only by portal dissection of the liver. In strictures of the upper ducts, partial hepatectomy may be feasible. Primary stricture is classified into two types, simple and membranous: *(1) Simple stricture* of the hilar duct is usually observed around the hilum, and is easily corrected by hilar ductoplasty. However, a slight hilar stricture may occasionally be overlooked on cholangiograms, and may become more severe as inflammatory attacks often develop. *(2) Membranous stricture* is seen in the umbilical portion,[24] and can be repaired by portal dissection in the liver, and a long, wide cholangio-jejunostomy incorporating hilar anastomosis is recommended for free drainage of intrahepatic bile. If the left hepatic lobe is involvement of complications, lateral segmentectomy may be indicated.[2,30]

Biliary carcinoma.
Cancer of the anastomotic site[31] or intrahepatic ducts[2,32,33] infrequently develops even in patients who have undergone an excisional surgery. It tends to occur in patients with jaundice due to long-standing obstructive cholangitis caused by stricture. Bile stagnation in the intrahepatic duct is possibly responsible for producing cancer.

Cancer also arises from the retained bile duct embedded in the pancreas.[34,35] Reflux of pancreatic juice has a hazardous effect on the retained epithelium and may result in cancer. Complete removal of the distal duct is desirable at the primary surgery.

Pancreatitis.
Pancreatitis occasionally develops several years after cyst excision in patients with the retained distal bile duct, protein plugs or stones in the dilated common channel, complex malunion, and/or pancreas divisum:[12,36]

Remnant intrapancreatic bile duct. The residual duct may cause not only carcinoma but also pancreatitis or stone.[37] In patients with recurrent epigastralgia even after cyst excision, an endoscopic retrograde pancreatogram should be performed to detect the remnant duct. Total excision of the extrahepatic bile duct, including intra-pancreatic bile duct, is essential.[2,13,22] In diffuse choledochal dilatation, a cholangiogram using a hemoclip placed on the distal bile duct near the malunion is beneficial to determine the excisional line for providing complete duct removal and avoiding damage to the pancreatic duct.[38]

Dilated common channel and malunion. Non-dilated common channel accompanied by malunion generally does not cause pancreatic disorders, whereas a dilated channel often accompanies protein plugs or stones, and may cause relapsing pancreatitis after surgery. Intraoperative cyst endoscopy is useful for removal of stones or plugs in the dilated channel.[13] Protein plugs, however, if left in the common channel, tend to be dissolved spontaneously by complete excision of the terminal duct.

In some patients, the orifice of the sphincteric papilla may be stenotic, and transduodenal or endoscopic papilloplasty may be indicated to achieve free drainage of pancreatic juice into the duodenum.[12] In selected patients with a complex malunion, a pylorus-preserving pancreatoduodenectomy may be indicated.[12]

Pancreatic carcinoma.
Pancreatic carcinoma rarley develops after surgery. Pancreatic disorders may occur because of bile influx into the pancreatic duct through the malunion. The relationship between pancreas cancer and pancreatobiliary malunion, however, still remains unclear.

Hepatic failure.
Patients may die of progressive hepatic dysfunction because of cirrhosis and portal hypertension caused by delayed surgery for choledochal cyst. Progressive hepatic dysfunction can be effectively managed by liver transplantation.

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