

Pancreatitis After a Primary and Secondary Excision of Congenital Choledochal Cysts

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Abstract

Purpose. Pancreatitis has been reported long after total choledochal cyst excision. The aim of this study was to determine if the disease process of postoperative pancreatitis differs between a primary and secondary cyst excision in a long-term follow-up.

Methods. Among 53 postoperative patients who underwent a total cyst excision and were followed up, 44 patients underwent a primary cyst excision (primary excision group), while 9 patients underwent a secondary cyst excision after a previous cyst-duodenostomy for internal drainage (secondary excision group). The long-term clinical course, including the pancreatographic findings after a total cyst excision, was compared.

Results. In the primary excision group, six patients had mild pancreatitis. Endoscopic retrograde pancreatography demonstrated ductal dilatation that was limited to the common channel in two patients, concurrent with the ventral duct in three, and extended the duct of Santorini in three. Conservative treatments were carried out in three patients, and endoscopic irrigation in one patient with protein plugs in the ventral duct. A resection of the choledochal remnant in the pancreas was performed in two patients with choledochal remnant-associated pancreatitis. From the secondary excision group, 5 of the 9 patients had chronic pancreatitis. Endoscopic retrograde pancreatography showed entire pancreatic ductal dilatation. Two of these patients underwent duodenal papiloplasty at the same time as secondary surgery; however, the disease progressively worsened.

Conclusion. In patients undergoing a secondary total excision after internal drainage, it is difficult to halt the ongoing aggravating process in pancreatitis.

Key words Congenital choledochal cyst · Pancreatic complications · Pancreatitis · Total cyst excision

Introduction

For the treatment of congenital choledochal cysts (CCC), a total cyst excision with a hepaticojejunostomy is now widely established as the treatment of choice. Although the results of a total cyst excision have been generally considered to be satisfactory, pancreatitis as a complication has been reported even long after a total choledochal cyst excision.¹⁻⁴

A total cyst excision has also been secondarily performed for patients who have previously undergone an internal drainage operation. However, in the case of this secondary operation, it remains unclear as to how total a cyst excision influences morbidity in which an internal drainage has been previously performed. Particularly in pancreatitis, little is known about the long-term disease process after a secondary cyst excision has been performed.

In the present study we divided the patients into two groups, consisting of patients who underwent a primary cyst excision and those with secondary cyst excision after previous internal drainage. We compared the long-term clinical findings between these groups of patients, including the pancreatographic findings after total cyst excision, to determine whether the disease process of pancreatitis differed.

Patients and Methods

Among 137 patients who underwent operations for CCC at the Division of Pediatric Surgery, Nihon University Itabashi Hospital, from January 1964 to December 2004, medical records of 53 postoperative patients

(37 females and 16 males) who had been periodically followed up were retrospectively reviewed in this study. The age of the patients ranged from 6 to 30 years old (mean: 18.9 years).

As a surgical procedure, total cyst excision and Roux-en-Y hepaticojejunostomy is a standard procedure for CCC. This procedure was performed as an initial operation in 44 of the patients (primary excision group) and was done after the initial cyst-duodenostomy in the remaining 9 patients (secondary excision group).

The duration of the postoperative follow-up was 5 years or less in 8 patients, 6–10 years in 8 patients, 11–15 years in 22, 16–20 years in 9, and 21 years or more in 6, with the longest follow-up period being 29 years.

The postoperative clinical course and pancreaticographic findings were evaluated.

Diagnoses of acute pancreatitis and chronic pancreatitis were made based on the standardized criteria for clinical diagnosis of acute pancreatitis⁵ and the clinical diagnostic criteria for chronic pancreatitis of the Japanese Society of Gastroenterology,⁶ respectively. Patients with other pancreatic diseases and acute abdomen due to different causes were excluded.

The image of pancreatic ductal morphology was obtained by endoscopic retrograde pancreatography (ERP). The morphological categories of the pancreatic ductal system were classified into four types, including a usual type, an ansa pancreatica type, a loop type, and a divisum type.¹ For the convenience of the measurements at each site of the pancreatic duct, the pancreatic ductal system was subdivided into the common channel (from the duodenal major papilla to the pancreaticobiliary junction), the ventral duct (from the pancreaticobiliary junction up to the junction of the main and accessory ducts), the duct of Santorini, and the ducts of body and tail.¹

The diameters of the largest point of each part of the duct were measured. In the present study, a positive finding of dilatation was recorded if the diameter of the

common channel or the ventral duct exceeded 4 mm, or the diameter of the duct of Santorini or the duct of the pancreatic body exceeded 3 mm.

Results

Of the 53 patients who underwent a primary and secondary total cyst excision, 11 developed pancreatitis postoperatively during the routine checkups on an out-patient basis. These patients comprised 6 from the primary excision group and 5 from the secondary excision group.

Clinical Course in the Primary Excision Group

In the six patients (5 females and 1 male) who underwent a primary cyst excision, age at surgery ranged from 1 to 6 years of age, and the age of onset of pancreatitis after surgery was 5–18 years (mean: 11.3 years) (Table 1). All had recurrent abdominal pain that was aggravated by consuming oily foods. All had high amylase levels in their blood and/or urine. Three patients (patients 4, 5, 6) presented with a mild increase in the white blood cell count and were positive to C-reactive protein in the blood. No other significant abnormal values were found in the blood chemistry, which indicated the severity of pancreatitis. An abdominal ultrasonographic and computed tomographic (CT) scan revealed no swelling or parenchymal heterogeneity. With a diagnosis of acute mild pancreatitis, the patients were given nothing orally, but only intravenous fluid with gabexate mesilate and antibiotics.

After the acute phase of inflammation subsided, ERP was performed (Table 2). The pancreatic ductal type was found to be the usual type in five patients and the loop type in one patient. All had ductal dilatations in various portions of the pancreatic duct. However, no entire pancreatic duct dilatation was observed in this

Table 1. Profiles of patients with pancreatitis in the primary excision group

Patient no.	Sex	Age (years) at primary cyst excision	Age (years) at onset of pancreatitis	Serum/urine amylase level (mIU/l)	Treatment of pancreatitis	Outcome
1	F	4	8	410/860	Conservative treatment	Improved
2	F	3	14	390/1010	Conservative treatment	Improved
3	M	1	5	470/1100	Conservative treatment	Improved
4	F	6	13	310/1200	Protein plug removal by endoscopy	Improved
5	F	4	10	780/1320	Resection of choledochal remnant	Improved
6	F	5	18	680/1235	Resection of choledochal remnant	Improved

Primary excision group, the group of patients who underwent a primary cyst excision

Table 2. Pancreatograms of patients with pancreatitis in the primary excision group

Patient no.	Pancreatic ductal types	Diameter of the duct (mm)			
		Common channel	Ventral duct	Duct of the body	Duct of Santorini
1	Usual	8	2	2	Not patent
2	Usual	5	2	1	Not patent
3	Loop	4	10	1	6
4	Usual	3	5	1	4
5	Usual	4	Protein plug 3	2	3
6	Usual	4	3	2	Not patent

Table 3. Profiles of patients with pancreatitis in the secondary excision group

Patient no.	Sex	Age (years) at			Additional procedures to cyst excision	Outcome	Follow-up period (years)
		Internal drainage	Onset of pancreatitis	Secondary cyst excision			
7	F	10	11	14	Duodenal papiloplasty	Recurrent pancreatitis	18
8	F	12	12	13	None	Recurrent pancreatitis	20
9	F	6	8	11	None	Recurrent pancreatitis	21
10	F	9	10	11	Duodenal papiloplasty	Chronic pancreatitis	25
11	F	4	5	12	Duodenal papiloplasty	Chronic pancreatitis	22

Secondary excision group, the group of patients who underwent a secondary cyst excision after internal drainage

**Fig. 1.** A postoperative pancreatogram demonstrating protein plugs (arrows) in the ventral duct (patient 4)

group. The dilatation of pancreatic duct was limited to the common channel in two patients, and was concurrent with the ventral duct in three. In addition, ductal dilatation extended to the duct of Santorini in three patients (patients 3, 4, 5).

A protein plug developed in the dilated ventral duct in one patient (patient 4, Fig. 1). Endoscopic irrigation

was performed to remove the protein plug. The patient has experienced no sign of abdominal pain since then.

In two patients (patients 5 and 6), pancreatitis associated with choledochal remnant in the pancreas developed. The choledochal remnant was dilated where a protein plug was found in one patient (patient 5). The choledochal remnant in the pancreas was resected by a laparotomy. The postoperative clinical course was uneventful; no sign of abdominal pain was noted. The six patients who underwent a primary cyst excision have experienced no other late complications, such as intrahepatic gallstones.

Clinical Course in the Secondary Excision Group

Among the nine patients who had undergone internal drainage, five occasionally experienced intermittent abdominal pain after a previous internal drainage operation (Table 3). The pain sometimes became persistent, especially after eating oily food. Abdominal pain was occasionally misdiagnosed as acute enterocolitis by local physicians. Finally, they were referred to the hospital and all underwent a total cyst excision as a reoperation; however, they continued to have the same type of abdominal pain as before. Five patients had recurrent abdominal pain with high amylase levels in

Table 4. Pancreatograms of patients with pancreatitis in the secondary excision group

Patient no.	Pancreatic ductal types	Diameter of the duct (mm)					Duct of Santorini
		Common channel	Ventral duct	Duct of the body	Duct of the tail		
7	Usual	6	2	4	3	Not visualized	
8	Usual	Protein plug 5	2	4	3	Not visualized	
9	Usual	5	10	4	2	Not visualized	
10	Usual	6	5	4	3	Not visualized	
11	Usual	Calculi 6	Calculi 6	4	3	Not visualized	
		Calculi					

the blood and urine, thus suggesting the occurrence of pancreatitis. Endoscopic retrograde pancreatography showed pancreatic ductal dilatation in all patients (Table 4). An abdominal CT scan revealed pancreatic parenchymal calcification in two patients (patients 10, 11), thus indicating chronic pancreatitis. No other late complications were detected.

Patient 10

This 28-year-old woman, who had undergone a secondary cyst excision 7 years after previous internal drainage, was admitted to our hospital because of recurrent abdominal pain. According to her past medical history, she had undergone a cyst-duodenostomy at 9 years of age at a local hospital. Recurrent abdominal pain developed a year after the internal drainage operation. She was admitted to a neighboring hospital for further evaluation. An abdominal CT scan demonstrated a dilated pancreatic duct containing protein plugs and calculi, thus leading to a diagnosis of chronic pancreatitis. She was subsequently transferred to our hospital and underwent a laparotomy at 11 years of age. Intraoperative pancreatography showed dilatation extending from the common channel to the duct of tail. These dilations were 6 mm (in diameter) in the common channel, 5 mm in the ventral duct, 4 mm in the duct of the body, and 3 mm in the duct of the tail. Duodenal papilloplasty was finally performed after a vigorous removal of the calculi, in addition to a total cyst excision.

Four years later, however, the abdominal pain reappeared, and amylase levels in the blood (826 mIU/l) and urine (1638 mIU/l) also increased. An abdominal CT scan found pancreatic calcification and extreme dilatation of the main duct remained. Endoscopic retrograde pancreatography showed more calcification in the parenchyma and calculi, which obstructed the main duct (Fig. 2). She is now a candidate for surgery due to chronic pancreatitis.

Patient 11

This 22-year-old woman also underwent a secondary cyst excision. She had a long history of recurrent ab-



Fig. 2. A postoperative pancreatogram demonstrating calcification (arrows) in the parenchyma and calculi leading to be an obstruction in the main duct (patient 10)

dominal pain after a cyst-duodenostomy at 4 years of age. Endoscopic retrograde pancreatography 8 years after the initial operation demonstrated dilatation of the entire pancreatic duct, with no significant parenchymal calcification. Papilloplasty was also performed with a total cyst excision; however, abdominal pain recurred again shortly thereafter. She recently presented with a recurrent abdominal and transient elevation of the serum and urine amylase levels, and was thus treated with conservative medical treatment.

Discussion

This study shows that the pancreatitis observed in patients undergoing primary cyst excision was mild with no particular treatment required, although it improved after a complete resection of the remaining choledochus in the pancreas in two particular patients undergoing an inadequate cyst resection. However, with a total cyst excision after internal drainage, it was difficult to halt the ongoing aggravating process in pancreatitis once chronic pancreatitis developed.

Internal drainage is a procedure that is contraindicated for congenital choledochal cyst at present. A total of nine pediatric patients undergoing internal drainage, such as a cyst-duodenostomy, were followed up in the current study. We finally performed total secondary cyst excisions in all patients, regardless of whether or not they had pancreatitis. Internal drainage itself influences morbidity after a total cyst excision.

In view of the pancreatographic findings, patients with pancreatitis after a primary cyst excision were grouped into four categories. The first category contained two patients demonstrating a usual type with mild dilation in the common channel (patients 1 and 2). Pancreatitis was mild and self-limiting with no particular treatment required. No other morphological abnormalities were demonstrated.

In the second category, a patient demonstrated a loop type with dilations in the ventral duct and the duct of Santorini (patient 3) which were both extremely dilated (10 and 6 mm, respectively). This case belonged to a type of patient in which pancreatitis and calculi are very likely to develop.⁴ This patient has thus far not developed calculi postoperatively.

The third category contained a patient with the usual type of dilation in the ventral duct containing a protein plug (patient 4). Protein plug formation is of clinical significance, but the etiology of the pancreatic protein plug remains uncertain. Some studies have indicated that the increased viscosity of the pancreatic juice is the result of elevated protein concentrations, thus leading to protein plug formation and a temporary blockage of the pancreatic duct by a protein plug, pancreatic calculi, or dysfunction of the sphincter of Oddi.^{1,7,8} The blockage of the pancreatic output may cause dilatation of the pancreatic duct and lead to abdominal pain;⁹ however, the symptoms were of a transient nature because of the small size of the plug in this patient. Endoscopic irrigation is thus the preferred treatment to remove the plug.

Finally, in the patients with a dilated choledochal remnant (patients 5 and 6), the etiology of dilatation remains uncertain. There might be an incomplete resection of the cyst at the primary operation. As the pancreatic duct lies close to the intrapancreatic cyst wall, there

is a risk of injury to the pancreatic duct when one attempts to remove the bottom of the cyst buried in the pancreatic parenchyme during an operation. Some authors recommended a partial cyst excision, thus leaving behind the intrapancreatic portion of the cyst, to avoid pancreatic injury.¹⁰⁻¹² However, others have proposed a complete excision of the cyst including the portion buried in the pancreas.¹³ Although it is unclear as to how much of the cyst buried in the pancreas should be resected, our cases proved the necessity of a complete cyst resection close to the pancreatic duct.

It is well known that internal drainage should no longer be the treatment of choice for congenital choledochal cysts. A total excision is now the definite treatment of choice, also for patients who have previously undergone internal drainage. It is true that internal drainage has been performed when the patient is a small infant or in poor condition during a certain period in the past, thus producing many various complications associated with this inappropriate surgical procedure. Recurrent cholangitis, anastomotic stricture, biliary calculi, and biliary carcinoma tend to occur as complications after internal drainage.^{14,15} These biliary complications have been reportedly emphasized; however, few studies on cases of chronic pancreatitis after internal drainage have been reported to date.^{14,16}

Whether or not a total secondary cyst excision after previous internal drainage truly improves the pathological process of pancreatitis, or relieves the pancreatic symptoms during a long-term follow-up, still remains to be elucidated. This study showed that a secondary cyst excision fails to stop the aggravating process in pancreatitis once it becomes chronic.

In the patients with a secondary cyst excision, pancreatitis developed relatively earlier than in those with a primary excision. Internal drainage causes not only pancreatic juice and bile, but also intestinal fluids to mix together, thus causing pancreatic enzymes to be easily activated. This is why pancreatitis tends to become more severe in the early stage in such cases.

Once the entire pancreatic duct has become extremely dilated, extending to the pancreas tail, a total cyst excision fails to improve this progressive pathology, as does additional papilloplasty. These findings indicate that the changes in the pancreas become irreversible in pancreatitis. Other surgical procedures for pancreatic drainage should also be considered.

Therefore, in patients undergoing a total secondary excision after internal drainage, it is difficult to halt the ongoing aggravating process in pancreatitis. Special care should therefore be taken with pancreatic complications in patients who have undergone a total cyst excision after previously undergoing internal drainage.

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