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ORIGINAL ARTICLE

Retrospective Study

Impact of previous cyst-enterostomy on patients' outcome following resection of bile duct cysts

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Abstract

AIM: To analyze the impact of previous cyst-enterostomy of patients underwent congenital bile duct cysts (BDC) resection.

METHODS: A multicenter European retrospective study between 1974 and 2011 were conducted by the French Surgical Association. Only Todani subtypes I and IVb were included. Diagnostic imaging studies and operative and pathology reports underwent central revision. Patients with and without a previous history of cyst-enterostomy (CE) were compared.

RESULTS: Among 243 patients with Todani types I and IVb BDC, 16 had undergone previous CE (6.5%). Patients with a prior history of CE experienced a greater incidence of preoperative cholangitis (75% *vs* 22.9%, $P < 0.0001$), had more complicated presentations (75% *vs* 40.5%, $P = 0.007$), and were more likely to have synchronous biliary cancer (31.3% *vs* 6.2%, $P = 0.004$) than patients without a prior CE. Overall morbidity (75% *vs* 33.5%; $P < 0.0008$), severe complications (43.8% *vs* 11.9%; $P = 0.0026$) and reoperation rates (37.5% *vs* 8.8%; $P = 0.0032$) were also significantly greater in patients with previous CE, and their Mayo Risk Score, during a median follow-up of 37.5 mo (range: 4-372 mo) indicated significantly more patients with fair and poor results (46.1% *vs* 15.6%; $P = 0.0136$).

CONCLUSION: This is the large series to show that previous CE is associated with poorer short- and long-term results after Todani types I and IVb BDC resection.

Key words: Bile duct cyst; Congenital; Biliary disease; Cyst-enterostomy; Long-term outcome

Core tip: Previous cyst-enterostomy is associated with more severe clinical presentation, including increased incidence of synchronous cancer, as well as poorer short- and long-term results in patients undergoing operations for Todani types I and IVb bile duct cysts.

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INTRODUCTION

In contrast to Asian countries[1], congenital bile duct cysts (BDC) are rare in the United States and in Europe[2] , with an incidence between 1 in 13000 and 1 in 2 million live births[3], respectively. whereas the incidences vary from 1 in 1000 in Japan[4], to 1 in 13500 in United States[5] and 1 in 15000 in Australia[6]. For this reason, there are few published series concerning the Western experience with this disease process[7]. The optimal surgical procedure for the management of BDC is still debated with cyst-enterostomy (CE) having been preferred in the past owing to its technical simplicity[8,9]. However, CE has been reported to be associated with increased morbidity (especially biliary complications) and higher reoperation rates during short- and long-term follow-up in comparison to primary BDC resection[9-11]. Additionally, a few series have emphasized the negative impact of previous CE on results of secondary BDC resection[2]. Lastly, CE has been shown to be associated with an increased risk of cholangiocarcinoma[12,13]. Consequently, primary resection has been considered as the treatment of choice for BDC[9,14,15]. The purpose of the present study was to investigate the role of a previous CE on short and long-term outcomes after secondary cyst resection in a large European multicenter cohort of patients.

MATERIALS AND METHODS

Study population and patients' data collection

A multicenter retrospective study was conducted under the auspices of the Association Française de Chirurgie. During a 37 years period (between 1974 and 2011), 33 centers (including 24 academic centers) from 6 different European countries have included a total of 505 patients operated from Todani type I, IVb, II, III, IVA BDC.

The patients' medical records were included on a website database (http://www.chirurgie-viscerale.org) using an online computerized standardized questionnaire. Patients' demographic data, previous history of surgical and/or endoscopic interventions for hepatobiliary and pancreatic (HBP) disease, clinical symptoms, biochemical and imaging studies, details of surgical procedures, pathological data, duration of follow-up and short- and long-term postoperative outcome were collected.

Table 1 Patients' demographic data, todani bile duct cyst subtypes and imaging studies

	Patient with previous CE	Patient without previous CE	Total	*P* value
Patients	16	227	243	-
Median age (yr) at the time of BDC resection	47.8 (26-60)	29.2 (0-81)	30.8 (0-81)	0.0074
Sex ratio F/M	7	3.73	3.8	0.5359
Adults/children	16/0	148/79	164/79	0.0041
ASA score[1]				
1 or 2	15 (93.75%)	217 (95.6%)	232 (95.0%)	0.5350
⩾ 3	1 (6.3%)	10 (4.4%)	11 (4.5%)	
Types of imaging studies				
None (incidental detection)	1 (6.3%)	2 (0.9%)	3 (1.2%)	0.1855
Percutaneous ultrasound	8 (50.0%)	188 (82.8%)	196 (80.7%)	0.0042
Computed tomography	7 (43.8%)	119 (52.4%)	126 (51.9%)	0.5022
MR-cholangiography	9 (56.3%)	138 (60.8%)	147 (60.5%)	0.7194
ERCP	7 (43.8%)	40 (17.6%)	47 (19.3%)	0.0186
Endoscopic ultrasound	8 (50.0%)	36 (15.9%)	44 (18.1%)	0.0026
Percutaneous cholangiography	0	17 (7.5%)	17 (7.0%)	0.6115
Intravenous cholangiography	2 (12.5%)	13 (5.7%)	15 (6.2%)	0.2583
Intraoperative cholangiography	0	6 (2.6%)	6 (2.5%)	1.0000
Todani bile duct cyst types				
Type I	16	221	237	
Type IVB	0	6	6	1.0000
MBC adequately analyzed	12 (75%)	196 (86%)	208 (85.5%)	
Cyst involvement of MBC	5/12 (41.6%)	47/196 (24.0%)	52 (21.4%)	0.3173
MBC-1	3	39	42	
MBC-2	2	8	10	0.2420
Presence of PBM adequately explored	10	180	190	
Presence of anomalous PBM	8 (80.0%)	141 (78.3%)	149 (78.4%)	1.0000

[1]The American Society of Anesthesiology physical status score; CE: Cyst-enterostomy; ERCP: Endoscopic retrograde cholangio-pancreatography; PBM: Pancreato-biliary malunion; MBC: Main biliary confluence.

Additional data were obtained from e-mail exchanges or phone calls with the referral centers. Operative reports, pathology reports and imaging studies were systematically reviewed by the 3 senior authors (Jean-Yves Mabrut, Reza Kianmanesh and Jean François Gigot) in order to establish homogeneous disease classifications. Patients' operative risk was evaluated by using the American Society of Anesthesiology physical status score (ASA)[16]. A pediatric patient was defined as under 15 years of age.

Complicated clinical presentation was defined by the presence of severe episodes of cholangitis or pancreatitis, portal hypertension, biliary peritonitis or coexistent synchronous carcinoma. Disease involvement of the main biliary confluence (MBC) was also classified according to the classification reported by two of the co-authors[17]. Postoperative morbidity and mortality were defined at 3 mo or during hospital stay. Postoperative morbidity was graded according to Dindo-Clavien' classification[18]. Long-term outcome was evaluated according to the dedicated evaluation score reported by the Mayo Clinic[2]. Complete cyst excision was defined as without macroscopic dilatation of bile duct after resection.

BDC subtypes were defined based on imaging studies in accordance with the Todani classification[14]. Todani BDC subtypes included type Ⅰ in 47.3% (n = 239), type Ⅱ in 3.7 % (n = 19), type Ⅲ in 2.5% (n = 13), type Ⅳa in 14.4% (n = 73), type Ⅳb in 1.2% (n = 6) and type Ⅴ in 30.7% (n = 155). Only patient with isolated, extrahepative BDC disease (Type Ⅰ and Ⅳb) were included (n = 245). Two additional patients that did not undergoing resection were excluded and thus 243 were ultimately analyzed.

A pediatric patient was defined as being aged under 15 years of age. Patients' operative risk was evaluated by using the ASA[16]. Complicated clinical presentation was defined by the presence of severe episodes of cholangitis or pancreatitis, portal hypertension, biliary peritonitis or coexistent synchronous carcinoma. Disease involvement of the MBC was also classified according to the classification reported by two of the co-authors[17]. Postoperative morbidity and mortality were defined at 3 mo or during hospital stay. Postoperative morbidity was graded according to Dindo-Clavien' classification[18]. Long-term outcome was evaluated according to the dedicated evaluation score reported by the Mayo Clinic[2].

Statistical analysis

Comparisons between patient with and without CE were performed using the χ^2 test (or the Fisher's exact test when conditions for the χ^2 test were not fulfilled) for categorical variables and using the Student t test (or the Mann-Whitney nonparametric rank sum test in case of non-normality) for continuous variables. Predictive factors of poor or fair result during long-term follow-up were analyzed by multivariate statistical analysis. Significant variables at the 0.15 level in univariate analysis were introduced in the multivariate logistic regression model. Kaplan-Meier analysis was used to predict the postoperative survival rate at 1 year and at 3 years. The log-rank test was used to compare subgroups of patients at 1 year and at 3 years. Statistical analysis was performed using SAS® version 9.2 (SAS Institute Inc, Cary, North Carolina, United States).

RESULTS

Patients were stratified based on a history of a previous CE. Demographic data, Todani BDC subtypes, types

Table 2 Patients' clinical presentation

	Patients with previous CE *n* = 16	Patients without previous CE *n* = 227	Total *n* = 243	*P* value
Median delay between symptoms and diagnosis (mo) (range)	2 (0-486)	2 (0-360)	2 (0-486)	0.8848
Type of symptoms and signs				
Asymptomatic	2 (12.5%)	31 (13.7%)	33 (13.6%)	1.0000
Isolated pain	2 (12.5%)	104 (45.8%)	106 (43.6%)	0.0094
Cholangitis	12 (75%)	52 (22.9%)	64 (26.3%)	< 0.0001
Pancreatitis	2 (12.5%)	49 (21.6%)	51 (21.0%)	0.5344
Abdominal mass	2 (12.5%)	22 (9.7%)	24 (9.9%)	0.6631
Jaundice[1]	6 (37.5%)	59 (26.0%)	65 (27.2%)	0.3843
Pruritus	1 (6.2%)	2 (0.9%)	3 (1.2%)	0.1855
Weight loss	2 (12.5%)	6 (2.6%)	8 (3.2%)	0.0903
Complicated presentation	12 (75%)	92 (40.5%)	104 (42.8%)	0.0071
Number of patients with > 2 symptoms	9 (56.25%)	92 (40.5%)	102 (42.0%)	0.2942

CE: Cyst-enterostomy; [1]without fever.

Table 3 Coexistent hepato-biliary and pancreatic diseases

	Patients with previous CE *n* = 16	Patients without previous CE *n* = 227	Total *n* = 243	*P* value
Biliary disease	9 (56.2%)	52 (22.9%)	61 (25.1%)	0.0059
Common biliary atresia	0	1 (0.4%)	1 (0.4%)	1.0000
Stones	4 (25%)	37 (16.3%)	41 (16.9%)	0.3216
Gallbladder	0	16 (7.0%)	16 (6.6%)	-
Cyst	3 (18.8%)	9 (4.0%)	12 (4.9%)	-
Common bile duct	1 (6.3%)	22 (9.7%)	23 (9.5%)	-
Intra hepatic duct	0	4 (1.8%)	4 (1.6%)	-
Synchronous cancer	5 (31.3%)	14 (6.2%)	19 (7.8%)	0.0043

CE: Cyst-enterostomy.

of imaging studies (Table 1), clinical presentations (Table 2), coexistent HBP diseases (Table 3), types of surgical procedures (Table 4), and short- and long-term postoperative outcomes (Table 5) were then compared between the two cohorts. Finally, predictive variables of poor/fair long-term outcome are reported in Table 6.

Patients' demographic data, todani BDC subtypes and types of imaging studies

During a 37-year period (1974-2011), 243 patients underwent resections for Todani types Ⅰ and Ⅳb congenital BDC at 33 centers (including 24 academic centers). The patients' gender ratio was largely female (193/50, *i.e.,* 3.86%). Median age was 30.8 years old (range: 0.1-81 years) and 79 patients were classified at pediatric. Patients' characteristics are detailed in Table 1. Sixteen patients had undergone previous CE (16/243, *i.e.,* 6.58%), they were all adults (100% *vs* 65.2%; *P* = 0.0041) and significantly older (47.8-year-old *vs* 29.2-year-old; *P* = 0.0074) than those without a history of CE. Imaging studies used for the diagnosis of BDC were different between the 2 groups: Patients without previous history of CE were offered more non-invasive studies, whereas those patients with previous CE had been submitted to significantly more endoscopic techniques. There was no difference concerning the BDC sub-types, the MBC involvement and the presence of an anomalous pancreato-biliary malunion (PBM) (Table 1).

Table 4 Preoperative treatment and types of surgery

	Patients with previous CE *n* = 16	Patients without previous CE *n* = 227	Total *n* = 243	*P* value
Preoperative biliary drainage	2 (12.5%)	14 (6.2%)	16 (6.6%)	0.2842
Types of surgical procedures				
Absence of resection	1 (6.3%)	5 (2.2%)	6 (2.5%)	0.3384
Elective surgery	14 (87.5%)	214 (94.3%)	228 (93.8%)	0.2583
Emergency surgery	1 (6.2%)	9 (4.0%)	10 (4.1%)	0.5007
Complete cyst excision	13 (81.2%)	199 (87.6%)	212 (87.2%)	0.6631
Incomplete cyst excision	2 (12.5%)	23 (10.1%)	25 (10.3%)	
Superior excision	1 (6.3%)	13 (5.7%)	14 (5.8%)	
Inferior excision	1 (6.3%)	9 (4.0%)	10 (4.1%)	-
Unknown	0	1 (0.4%)	1 (0.4%)	
Associated procedures				
Hepatectomy	2 (12.5%)	4 (1.8%)	6 (2.5%)	0.0523
Pancreaticoduodenectomy	4 (25.0%)	4 (1.8%)	8 (3.3%)	0.0008
Stone extraction	2 (12.5%)	10 (4.4%)	12 (4.9%)	0.2072
Synchronous cancer excision	5 (31.3%)	13 (5.7%)[1]	18 (7.8%)	0.0043
Biliary reconstruction				
Hepatico-jejunostomy	13 (81.2%)	208 (91.6%)	221 (90.9%)	0.1658
Hepatico-duodenostomy	2 (12.5%)	7 (3.1%)	9 (3.7%)	0.1116
Choledoco-duodenostomy	0	1 (0.4%)	1 (0.4%)	1.0000
MBC anastomosis	8/14 (57.1%)	96/180 (53.3%)	104/194 (53.6%)	0.7831

CE: Cyst-enterostomy; MBC: Main biliary convergence; [1]secondary central hepatectomy was performed after good response of chemotherapy.

Clinical presentation

The median delay between the disease's first symptoms and the diagnosis of BDC was 2 mo (0-486) and was similar between the 2 groups of patients. Abdominal pain was the most common presenting symptom (43.6%) for the whole population and significantly more frequent in patients without previous CE (12.5% *vs* 45.8%; *P* = 0.009). The group of patients with previous CE was observed to have a significantly increased prevalence of cholangitis (75.0% *vs* 22.9%; *P* < 0.0001) and complicated presentation (75.0% *vs* 40.5%; *P* = 0.007) (Table 2).

Coexistent HBP diseases

Coexistent biliary disease (liver disease, pancreatic disease, and biliary disease) occurred in 25.1% of the present cohort and was significantly more frequent in

Table 5 Early postoperative and long term outcome of patients

	Patients with previous CE *n* = 16	Patients without previous CE *n* = 227	Total *n* = 243	*P* value
Median postoperative hospital stay (d) (range)	16 (9-110)	10 (2-150)	10 (2-150)	0.0014
Mortality rate	0 (0.0%)	1 (0.4%)	1 (0.4%)	1.0000
Overall complications	12 (75.0%)	75 (33%)	87 (35.8%)	0.0018
Grade I-II	5 (31.3%)	47 (20.9%)	52 (21.4%)	0.3454
Grade III-IV	7 (43.8%)	28 (12.3%)	35 (15.4%)	0.0031
Surgical complications	7 (43.8%)	45 (19.8%)	52 (21.4%)	0.0509
Biliary	0 (0.0%)	20 (8.8%)	20 (8.2%)	0.3750
Mixed pancreatic and biliary fistula	1 (6.3%)	5 (2.2%)	6 (2.5%)	0.3384
Pancreatic	1 (6.3%)	17 (7.5%)	18 (7.4%)	1.0000
Bleeding	5 (31.3%)	3 (1.3%)	8 (3.3%)	< 0.0001
Reoperation rate	6 (37.5%)	20 (8.8%)	26 (10.7%)	0.0032
Long-term outcome of patients				
	Patients with previous CE *n* = 13	Patients without previous CE *n* = 185	*n* = 198	
Median follow-up (mo) (range)	59 (12-175)	36 (4-372)	37.5 (4-372)	0.2482
Mayo clinic score				
Excellent	5 (38.5%)	138 (74.6%)	143 (72.2%)	
Good	2 (15.4%)	18 (9.7%)	20 (10.1%)	0.0099
Fair	0	5 (2.7%)	5 (2.5%)	
Poor	6 (46.2%)	24 (13.0%)	30 (15.1%)	
Mayo clinic score				
Excellent + good	7 (53.8%)	156 (84.3%)	163 (82.3%)	0.0136
Fair + poor	6 (46.2%)	29 (15.7%)	35 (17.7%)	

CE: Cyst-enterostomy.

patients with previous CE (56.2% *vs* 22.9%; *P* = 0.006). The occurrence of associated calculus disease (16.9%) was similar between the two groups. Synchronous biliary cancer occurred in 7.8% of the total cohort and was significantly more frequent in patients with previous CE (31.3% *vs* 6.2%; *P* = 0.004) (Table 3).

Preoperative treatment and type of surgery

Preoperative biliary drainage was performed in 6.6% of the whole cohort. All patients underwent surgical exploration. However, 5 adult patients without CE did not undergo BDC resection, one for no specified reason, the others respectively for locally advanced gallbladder cancer, peritoneal carcinomatosis, severe inflammation of the hepatic pedicle, performance of simple cholecystectomy. Only one patient with previous CE had severe hepatic pedicle inflammation prohibiting cyst resection. Associated bile duct stone extraction rates were similar between the two subgroups. Complete cyst excision was accomplished in 93.8% of all patients and there was no significant difference between the two groups of patients. Associated hepatectomy was more frequently performed in patients with previous CE but

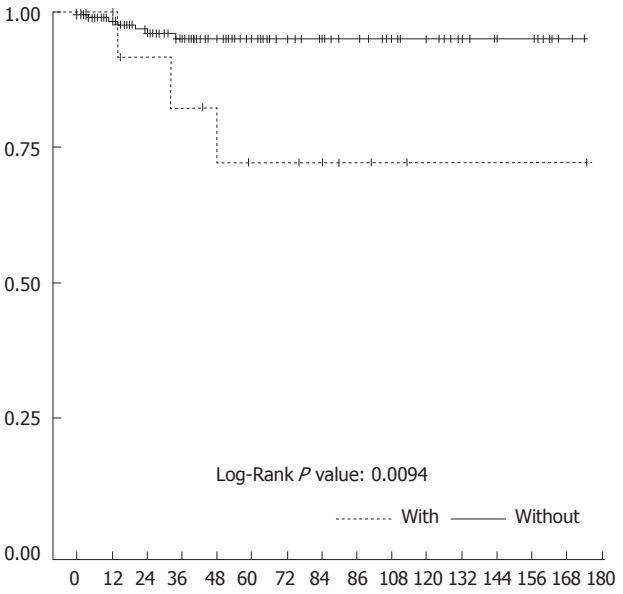

Figure 1 Comparison of overall survival between patients operated from bile duct cysts with and without previous history of cyst-enterostomy.

the difference was not significant (12.5% *vs* 1.8%; *P* = 0.0523). Associated pancreaticoduodenectomy was significantly more frequent in patients with previous CE (25% *vs* 1.8%; *P* = 0.008) (Table 4).

Postoperative mortality, morbidity and early postoperative outcome (within 3 mo)

Postoperative death due to cardiac arrhythmia, occurred in 1 patient (0.4%). Overall morbidity and severe complications rates were 36.2% and 14%, respectively. Patients with previous CE had significantly higher morbidity rates (75.0% *vs* 33.5%; *P* < 0.0008), more severe complications (Grade Ⅲ-Ⅳ) (43.8% *vs* 11.9%; *P* = 0.003), more hemorrhagic complications (31.3% *vs* 1.3%; *P* < 0.0001), a greater reoperation rate (37.5% *vs* 8.8%; *P* = 0.003) and a longer median length of stay (16 *vs* 10 d; *P* < 0.001) than those without previous CE (Table 5).

Postoperative long-term outcome

A total of 44 patients were lost for follow-up at 3 mo, only 3 of whom belonged to the subgroup of patients with previous CE. The median follow-up in the 198 remaining patients was 37.5 mo (range: 4 to 732 mo) for the whole cohort, without any difference between both subgroups. According to the dedicated Mayo Clinic Risk score evaluating long-term results, there were significantly more patients with fair and poor results in the subgroup of patients with previous CE (*P* = 0.001). The overall 3-year survival rate was significantly lower in patients with prior CE (82.5% *vs* 95.9%; *P* = 0.01) (Figure 1) (Table 5).

Univariate and multivariate analysis

Predictive variables of poor and fair long-term results (according to the Mayo Clinic clinical score) were

Table 6 Predictive factors of poor and fair long-term outcome according to Mayo clinic score in 198 patients with a follow-up > 3 mo, including patients suffering from synchronous cancer

Covariate	Univariate			Multivariate		
	OR	95%CI	*P* value	OR	95%CI	*P* value
Adult patient *vs* child	3.587	1.322-9.735	0.0084	-	-	-
Previous cyst-enterostomy; Yes *vs* No	4.611	1.445-14.712	0.0136	3.165	0.829-12.077	0.0918
Synchronous cancer; Yes *vs* No	18.462	4.687-72.71	< 0.0001	16.612	3.999-69.013	0.0001
Post-operative complications; Yes *vs* No	2.397	1.143-5.028	0.0186	-	-	-
Postoperative biliary complications; Yes *vs* No	4.356	1.669-11.367	0.0038	4.597	1.635-12.925	0.0038

OR: Odds ratio.

evaluated in the 198 patients surviving surgery and with a follow-up over 3 mo. Univariate statistical analysis indicated that predictive variables of poor or fair long-term results were to be an adult patient, with prior CE, postoperative complications, postoperative biliary complications and coexistent synchronous cancer. By multivariate analysis, predictive variables of poor or fair long-term results were previous CE, synchronous cancer and postoperative biliary complications (Table 6).

DISCUSSION

The present retrospective series shows that patients submitted to secondary resection of congenital BDC following a previous cyst-enterostomy suffered more complications before, during and after surgery with poorer results during long-term follow-up. Strengths of the present series include the relatively large cohort of patients issued from a multicentric European series, and the central revision of radiological, operative and pathological data, thereby ensuring a homogeneous classification of patients and their symptoms and signs both prior and after surgery, with the use of a dedicated clinical score for long-term results. Furthermore, the analysis was performed in a homogeneous group of BDC with only extrahepatic biliary involvement, namely patients suffering from Todani types I and IVb BDC. Indeed, patients with Todani type IVA were excluded from the present series so that poor results could not be due to residual non-resected intrahepatic biliary disease.

According to the results of the present study, primary complete excision of BDC, with the construction of a wide bilio-digestive anastomosis to a healthy proximal bile duct should be the "gold standard" surgical management of patients suffering from BDC[2,14,19]. Cyst-enterostomy must definitively be abandoned as a treatment option. Limitations of the study include its retrospective design, its long period (37 years) of patients inclusion and the number of patients without available long-term follow-up. The comparison between patients with previous cyst-enterostomy and the control group should also be considered with caution due to the small number of patients in the CE group.

However, despite cyst-enterostomy being no longer the primary approach for the surgical management of BDC, its prevalence was 17.2% in 186 patients operated on after 1980[2,7,20-22] and previous cyst-enterostomy was still observed in 7.3% of 354 patients operated on after 1990[23-26]. Practically, this means that previous CE still remains a challenge during the management of BDC.

Indeed, the consequences of having undergone a cyst-enterostomy, regardless of the technique used, easily explain the more complicated clinical presentation of previously operated patients encountered in the present series: Possible mechanisms include reflux of digestive juice through the entire biliary tract, activation of pancreatic juice by enterokinases linked to pancreatico-biliary malunion[27] or even anastomotic stricture of the CE on diseased cystic biliary tissue[19,28,29]. Any of these can lead to severe biliary inflammation[15], cholangitis, hepatolithiasis[10,19,30,31] and increase the risk of carcinogenesis[12,13,19,27]. The latter is estimated to be over 50% by Todani *et al*[27]. Indeed, malignant degeneration of BDC occurs more than 15 years earlier in patients with previous CE than in patients with primary carcinomas, with a median delay of 4 years in a series by Flanigan *et al*[13] to 10 years (range: 1-53 years of delay) in yet another by Todani *et al*[27] and, overall, is associated with a very poor prognosis. Finally, the reoperation rate after previous CE is high, ranging from 15.7% to 87.5%[1,9,30,32].

The present series also shows that more complex surgical procedures had to be used for patients with a previous CE, mainly due to an increased need for a pancreaticoduodenectomy. Such an extensive procedure was linked, for half of these patients, with the presence of coexistent carcinoma, thereby, requiring tumor resection with a wide tumor-free margin. The other pancreaticoduodenectomies were mainly performed because of severe inflammation within the hepatic pedicle, probably as a result of repeated episodes of cholangitis. This feature observed during secondary BDC resection in patients with previous CE has not been reported until now.

According to several surgical series which compared primary CE with primary BDC resection with Roux-en-Y hepaticojejunostomy have reported an increased mortality rate (range: 8.3%-10% *vs* 0%-7%), morbidity rate (range: 17%-93% *vs* 0%-17%) and reoperation rate (range: 29.7%-87.5% *vs* 0%) in patients with primary CE[9,10,30]. At the time of writing, there are only

3 studies that compared the morbidity and mortality rates between primary BDC resection and secondary BDC resection with previous CE, and these concerned series with only small numbers of patients[2,15,19]. For patients with previous CE, Chijiiwa *et al*[19] and Gigot *et al*[2] observed significantly more early postoperative complications, whilst Kaneko *et al*[15] reported significantly increased operative blood losses, operative time, early postoperative complications and wound infections. The present series demonstrates similar findings of an increased rate of overall and severe postoperative complications as well as hemorrhagic complications and reoperation rates for patients operated following previous CE.

The key-messages of this first large European multicenter study are that BDC patients who have undergone previous CE have more complications including carcinoma, that long-term results and survival rate are worse and that the reoperation rate is greater. It should be emphasized that these results cannot be attributed to intrahepatic disease alone as only Todani type I and IVb lesions were studied in this analysis. Late complications were also increased in the series reported by Kaneko *et al*[15], especially regarding late development of hepatolithiasis and pancreatic stones, though this was not reported in the series by Chijiiwa *et al*[19]. Finally, multivariate statistical analysis confirmed in the present series that previous CE was representing an independent predictive factor of fair or poor prognosis after secondary BDC resection. Weaknesses of the present study include its retrospective nature, the number of patients excluded from long-term follow-up after 3 mo (18.1% of the whole series), the limited median follow-up duration of 37.5 mo and the small numbers of patients with previous CE.

In conclusion, this European retrospective series showed that previous CE was associated with a more complicated presentation, more coexistent HPB diseases including synchronous cancer, more complex surgery and worse early and long-term postoperative outcomes. These features confirmed the abandonment of cyst-enterostomy for the surgical management of congenital bile duct cysts.

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COMMENTS

Background

Congenital bile duct cysts (BDC) are rare in the United States and in Europe. Cyst-enterostomy has been reported to be associated with increased morbidity (especially biliary complications) and higher reoperation rates during short- and

long-term follow-up in comparison to primary BDC resection. Consequently, primary resection has been considered as the treatment of choice for BDC. The purpose of the present study was to investigate the role of a previous cyst-enterostomy (CE) on short and long-term outcomes after secondary cyst resection in a large European multicenter cohort of patients.

Research frontiers

Limitation of the study includes its retrospective design, its long period (37 years), included patients and the number of patients without available long-term follow-up. The comparison between patients with previous cyst-enterosotmy and the comtrol group should also be considered with caution due to the small number of patients in the CE group. However, despite cyst-enterostomy being no longer the primary approach for the surgical management of BDC, its prevalence was 17.2% in 186 patients operated on after 1980 and previous cyst-enterostomy was still observed in 7.3% of 354 patients operated on after 1990. Practically, this means that previous CE still remains a challenge during the management of BDC.

Innovations and breakthroughs

This European retrospective series showed that previous CE was associated with a more complicated presentation, more coexistent HPB diseases including synchronous cancer, more complex surgery and worse early and long-term postoperative outcomes. These features confirmed the abandonment of cyst-enterostomy for the surgical management of congenital bile duct cysts.

Applications

These features confirmed the abandonment of cyst-enterostomy for the surgical management of congenital bile duct cysts.

Peer-review

The results of this European multicenter study are very interesting and the manuscript is well-written.

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