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Bile Duct Cysts in Adults

A Multi-institutional Retrospective Study

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Objective

To review the features of adult patients undergoing surgery for bile duct cysts, focusing on the anatomy of the biliary tree as well as the long-term outcome.

Summary Background Data

Bile duct cysts (BDCs) are uncommon in Western countries, and the majority of reported cases originate from Asia. Japanese authors have emphasized the frequent association of extra- and intrahepatic bile duct dilatations, but grading of patients based on Todani's classification is often hindered by the absence of an accurate definition of types IC and IVA cysts. Moreover, despite the increasing use of extrahepatic cyst excision, little is known about the long-term outcome in patients with intrahepatic bile duct involvement.

Methods

Forty-two adult patients with BDC were treated between 1980 and 1992 in 17 institutions of the French Associations for Surgical Research. Clinical presentation, radiologic presurgical evaluation, and surgical procedures were analyzed. The long-term postsurgical outcome was derived from patient charts, attending physicians, or direct patient contact.

Results

Twelve patients (30%) had recurrent abdominal pain or jaundice from childhood. Seven (17%) had undergone prior cystenterostomy. Twenty-one (50%) had a Todani-type IVA cyst with extra- and intrahepatic bile duct involvement. Of these, nine had segmental, exclusively left-sided intrahepatic bile duct dilatation. Biliary carcinoma was encountered in five patients (12%). Extrahepatic cyst excision with a Roux-en-Y hepaticojejunostomy was performed in 34 patients with type I or IV cysts. The overall operative mortality rate was 2.4%. Long-term results were clearly correlated with cyst type: during a mean follow-up of 8.4 years, 11 of 12 patients (92%) treated by cyst excision for type I cyst remained free of symptoms, whereas 31% of patients who underwent surgery for type IV cyst had episodic or severe cholangitis with intrahepatic stones.

Conclusions

In patients with BDC, particular attention must be given to the associated intrahepatic bile duct dilatations. We propose a modification of Todani's classification to distinguish cystic, segmental, and fusiform dilatations of the intrahepatic biliary tree in type IV cysts. In patients with segmental left intrahepatic cystic dilatations, combined left liver lobectomy and extrahepatic cyst excision is suggested to decrease late postsurgical biliary complications.

Bile duct cyst (BDC), also called choledochal cyst, is rare,[1–3] with <3500 cases reported, mainly in the Japanese literature.[4] The incidence has been estimated to range from 1 in 50,000 to 1 in 200,000 live births in Western countries but is much more frequent in Asia.[1,5,6] In contrast to pediatric cases, which are treated in specialized institutions,[7,8] adult patients with BDC present sporadically to general hospitals, and some require emergent procedures for acute complications,[3,9–11] underlining the need for a greater awareness of this condition.

This study was designed to assess the anatomic features of BDC in adults and to evaluate the long-term results of extrahepatic cyst excision. Particular attention was given to patients with intrahepatic bile duct dilatations.

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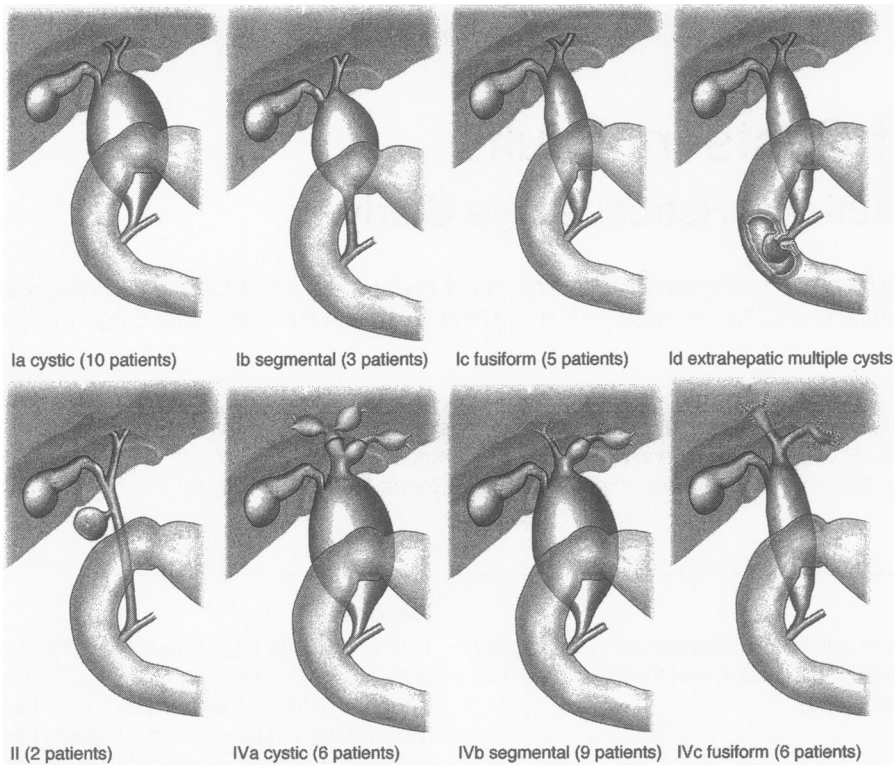

Figure 1. Modified Todani's classification of BDCs. Types IA, IB, IC, and ID are extrahepatic BDCs; type II is a diverticulum of the common bile duct; types IVA, IVB, and IVC are combined intra- and extrahepatic BDCs. Type III, or choledochocele (one patient), and type V (Caroli's disease) remained unchanged, like type II, and are not illustrated in this figure.

METHODS

The medical records of 47 patients who underwent surgery for BDC between 1980 and 1992, collected from 17 institutions of the French Associations for Surgical Research, were reviewed. Presenting signs and symptoms, radiologic data, and treatment were analyzed. Presurgical and surgical radiologic evaluations were reviewed to determine the type of cyst and to identify anomalies of the pancreaticobiliary ductal junction (APBDJ). BDCs were classified according to a modification of Todani's classification[12] (Fig. 1), and the APBDJ was classified according to Kimura et al.[13] (Fig. 2). Patients with type V cyst (Caroli's disease) were not included. After reexamination of the medical records and radiologic data, five patients with fusiform dilatation of the extrahepatic common bile duct, a normal pancreaticobiliary junction, and previous surgery for biliary lithiasis were excluded from this study because they were considered to have secondary and not congenital biliary dilatations.

Data regarding postoperative courses were derived from patient charts and primary physician and patient interviews. Four patients (9%) were lost to follow-up in the short or medium term (18 months to 4 years); all were clinically well when seen last. Patient groups were compared using the chi square test with Yate's correction when the minimal expected value was <5.

Demographics and Clinical Presentation

Forty-two patients with BDC were included in this study; 33 were female, for a ratio of female to male of 3.5:1. The mean age was 33.4 $\pm$ 16 years (range 16 to 70 years). One to seven patients were collected from each institution.

The clinical presentation was nonspecific. Thirteen patients (31%) had right upper or epigastric abdominal pain; 23 patients (55%) had jaundice, associated with cholangitis in 17 and mild acute pancreatitis in 3. The classical triad of

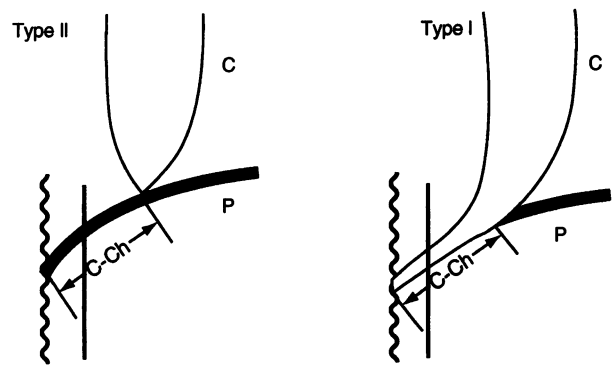

Figure 2. Classification of APBDJ. Type II or CP junction: the common bile duct enters the main pancreatic duct. Type I or PC junction: the main pancreatic duct joins the common bile duct. C, common bile duct; P, main pancreatic duct; C-Ch, common channel.

Table 1. PATIENTS WITH PRIOR CYSTENTEROSTOMY

Age and Sex	Prior Surgery	Age Prior Operation (yr)	Type of Cyst	Cancer
20 yr. F	Cyst duodenostomy	8	IVB	–
17 yr. F	Cyst duodenostomy	1	IVC	–
43 yr. F	Cyst duodenostomy*	27	IVA	Cyst wall
27 yr. F	Cyst duodenostomy†	2	IVA	–
40 yr. F	Cyst jejunostomy	18	IVA	Hepatic ducts
37 yr. F	Cyst jejunostomy	14	IVB	–
32 yr. M	Cyst jejunostomy	24	IA	Cyst wall

* Reoperation for revision.
† Reoperations 1/for gastrojejunostomy, 2/for cholecystojejunostomy.

abdominal mass, jaundice, and pain was encountered in only two patients (5%). Four patients (9.5%) had isolated acute pancreatitis, whereas two had no symptoms.

The duration of symptoms before diagnosis was known for 40 patients: <1 month in 9 cases (22%) and >1 year in 25 cases (62.5%). Twelve patients (30%) had recurrent abdominal pain or jaundice from infancy. Seven patients with cholangitis had undergone prior cystenterostomy (Table 1).

Presurgical Diagnostic Evaluation

In all 37 patients who underwent ultrasonography, the scan showed the cyst; pancreatic pseudocyst was suggested in 7. Intravenous cholangiography was used in 22 nonicteric patients, with two failures from dilution of contrast material. Computed tomography scanning demonstrated a large cystic mass in the porta hepatis of probable biliary origin in 19 of 20 patients.

Endoscopic retrograde cholangiopancreatography (ERCP, 28 cases) and percutaneous transhepatic cholangiography (PTC, 3 cases) were used to assess biliary and pancreatic duct anatomy, with one technical failure in each method. No septic complications occurred after retrograde or transhepatic biliary opacification. An accurate presurgical diagnosis was obtained in 33 of the 35 patients (94%) presenting as new cases.

Cyst Type and APBDJ

BDCs were classified according to the results of ERCP and PTC in 29 patients and surgical cholangiography in the others. In five cases, contrast material was too diluted to define the intrahepatic biliary tree by ERCP, and comparison with surgical cholangiography was necessary to define the cyst type. There were 18 type I (10 IA cystic, 3 IB segmental, and 5 IC fusiform), 2 type II, and 1 type III cysts. In 9 (43%) of the 21 patients with type IV cyst, intrahepatic dilatations were exclusively located on the left side of the intrahepatic biliary tree (Table 2). These patients were classified as type IVB in the modified Todani's classification illustrated in Figure 1. No patients had exclusively right-sided intrahepatic bile duct involvement. Twelve other patients had bilateral intrahepatic dilatations, either cystic (type IVA, six patients) or fusiform (type IVC, six patients) (see Fig. 1).

The pancreaticobiliary ductal junction was visualized by ERCP in 27, PTC in 2, and surgical cholangiography in 13 of the 39 patients with type I or IV cysts. Thirty-five results could be analyzed correctly: 3 had a normal junction, 30 (86%) had a type II or CP junction with a long common channel (mean length 25 ± 6 mm), and 2 (5.7%) had a type I or PC junction (see Fig. 2).

Associated Hepatobiliary and Pancreatic Diseases

At the time of initial surgery or resurgery, 28 of the 42 patients (66%) had associated hepatobiliary and pancreatic disease (Table 3). Biliary lithiasis was observed in 26 patients (62%). All four patients with common channel lithiasis also had intracystic stones. Five other patients (12%)

Table 2. HEPATIC BILE DUCTS INVOLVEMENT IN TYPE IV-A CYSTS

		Hepatic Ducts		Intrahepatic Ducts	
Type of Cyst	No. of Patients	Left	Right	Left	Right
IV-A cystic	10	10	7	7(4)*	3
IV-A fusiform	11	11	8	8(5)*	3

* Number of patients with exclusively left-sided intrahepatic bile duct dilatations.

Table 3. ASSOCIATED HEPATOBILIARY AND PANCREATIC DISEASES (AT THE TIME OF INITIAL OPERATION OR REOPERATION)

Biliary lithiasis (n = 36)	Intracystic stones	18
	Intrahepatic stones	7
	Gallstones	7
	Common channel stones	4
Hepatic or portal complications (n = 6)	Portal vein thrombosis	2
	Intrahepatic abscess	2
	Secondary biliary cirrhosis	2
Pancreatic abnormalities (n = 8)	Pancreas divisum	1
	Pancreatic cyst	1
	Left pancreatic atrophy	1
	"Curled" main pancreatic duct	4
	Pancreatic adenocarcinoma	1

presented with biliary carcinoma associated with BDC (Table 4). There were three women and two men with a mean age of 39 ± 8.8 years. Three malignancies were located in the cyst wall (2 type IA, 1 type IVA); two occurred 8 and 16 years after a cyst drainage procedure, and one was associated with carcinoma of the head of the pancreas. Another patient (type IVB cyst) had adenocarcinoma of the gallbladder without cholelithiasis, and one patient (type IVA cyst without intrahepatic stone) had carcinoma arising at the junction of the hepatic ducts, 22 years after cystjejunostomy.

In the 31 patients without cancer treated by complete cyst excision, the resected specimens exhibited no mucosal dysplasia or metaplasia. Histologic studies showed gross abnormalities of the bile duct epithelium with scattered areas of cylindrical or columnar epithelium separated by large erosions. Fibrotic, dense connective tissue with smooth muscle bundles and elastic tissue infiltrated by inflammatory polymorphic cells were found in the cyst wall.

Surgical Procedures

Extrahepatic cyst excision with Roux-en-Y hepaticojejunostomy was performed for type I or IV cysts in 29 of the 35 patients as initial treatment, and in 5 of the 7 who had undergone previous cyst drainage procedures. Two patients each underwent cyst excision associated with left hepatic lobectomy (type IVB cysts), transduodenal sphincteroplasty, and Whipple procedure for malignancy. Three patients who presented early in this series had partial cyst excision, closure of the distal stump, and Roux-en-Y end-to-side choledochocystojejunostomy, according to Hepp and Mutricy.[14]

Simple removal of the cyst was performed in the two patients with type II cysts. The patient with choledochocele (type III) underwent surgical transduodenal sphincteroplasty.

Two patients with biliary carcinoma underwent total hepatectomy with hepatic transplantation for cancer of the ductal bifurcation in one, and palliative external biliary drainage for unresectable intracystic carcinoma in the other.

RESULTS AND FOLLOW-UP

One patient with secondary biliary cirrhosis and portal vein thrombosis died of hepatic failure and diffuse abdominal hemorrhage after extrahepatic cyst excision, for an overall postoperative mortality rate of 2.4%. Eight other patients (18%) had postsurgical abdominal complications (biliary fistula in six cases, hemorrhagic gastritis and wound infection in one case each), all responding to conservative medical treatment.

The long-term results of the 41 surviving patients were assessed during a mean follow-up of 6.9 years (range 0.4 to 16 years). Twenty-nine patients (71%) remained free of biliary symptoms or complications (cholangitis or late malignancy) and were graded as having good results. Fair results were observed in three patients (7%) who had occa-

Table 4. BILIARY CARCINOMA ASSOCIATED WITH BDC

Age and Sex	Type of Cyst	ADJBP	Site of Carcinoma/Associated Disease	Treatment	Outcome (Months)
40 yr. F*	IVA	Unknown	Junction of hepatic ducts	Liver transplantation	Dead (4)
29 yr. F	IA	Type II	Cyst wall Carcinoma head of the pancreas	Cyst excision Whipple resection	Dead (12)
32 yr. M†	IA	Type II	Cyst wall Liver metastases and abscesses Cystolithiasis	External biliary and hepatic Abscesses drainage	Dead (4)
43 yr. F‡	IVA	Type II	Cyst wall Cystolithiasis	Cyst excision Whipple resection	Dead (30)
51 yr. M	IVB	Type II	Gallbladder (without cholelithiasis)	Cyst excision	Dead (11)

* Cystojejunostomy at age of 18.
† Cystojejunostomy at age of 24.
‡ Cystoduodenostomy at age of 27.
BDC = bile duct cysts.

Table 5. RESULTS OF EXTRAHEPATIC CYST EXCISION ACCORDING TO CYST TYPE

Type of Cyst	N	Deaths	Lost for Follow-up (Months)	Mean Duration of Follow-up (Years)	Intermittent Cholangitis	Severe Cholangitis/Hepatolithiasis
Type I	14	1*	1(18)	8,8	1	–
Type IVA	5	1*	–	8,6	1	1
Type IVB	9	2*†	–	8	–	2
Type IVC	6	–	1(24)	7,9	1	–
Total	34	4	2	8,4	3	3

* Recurrence of malignancy.
† Postoperative death.

sional (one or two) episodes of ascending cholangitis. Poor results were observed in nine patients (22%): all five patients who underwent surgery for biliary carcinoma died of recurrence 4 to 30 months after surgery (see Table 4), and four other patients had severe bouts of cholangitis, caused by intrahepatic stones in three.

Long-term outcome was evaluated in 34 patients who had undergone extrahepatic cyst excision. One patient died after surgery, three patients died of recurrence of malignancy, and two patients remained clinically well until they were lost to follow-up 18 and 24 months after surgery. The mean follow-up time in the 28 remaining patients was 8.4 years (range 4 to 16 years), with 13 patients having a follow-up >10 years. For 22 patients (78%), cyst excision resulted in complete resolution of abdominal pain and cholestasis. Three patients (11%) had occasional (one or two) episodes of ascending cholangitis and were graded as having fair results. At the time of writing, they are clinically well, with normal results on liver function tests. Poor results were observed in three patients (11%) who developed intrahepatic lithiasis: two, with a patent anastomosis, were treated by transhepatic lithotripsy and left liver lobectomy, 12 and 6 years, respectively, after primary surgery. One patient underwent revision of anastomotic stricture with extraction of intrahepatic stones 6 years after primary cyst excision. These three patients are clinically well, with normal results on liver function tests 2, 2, and 5 years after revisional therapy, respectively.

Results of extrahepatic cyst excision were analyzed according to the type of cyst (Table 5). Twelve of the 14 patients with type I cyst treated by extrahepatic cyst excision had long-term survival and complete follow-up: 11 (92%) remained free of symptoms, and 1 had episodic bouts of cholangitis. In comparison, of 16 patients treated by extrahepatic cyst excision for type IV cyst with long-term survival and complete follow-up, 5 (31%) had fair or poor results with episodic or severe cholangitis caused by hepatolithiasis (see Table 5). The difference between these two groups of patients was not statistically significant.

After partial cyst excision with end-to-side choledochocystojejunostomy (Hepp procedure), one patient remained free of symptoms until he was lost to follow-up 4 years later, one has recurrent bouts of cholangitis but still refuses to undergo another procedure at 12 years after surgery, and the last is asymptomatic 10 years after surgery.

Of the two patients treated by simple removal of type II cyst, one was lost to follow-up after 3 years and the other is clinically well 6 years after surgery. The patient with type III cyst remained free of biliary symptoms until he died of cardiac failure 7 years after undergoing transduodenal sphincteroplasty.

DISCUSSION

BDCs presenting after childhood are uncommon[11]: 25% of all cases are diagnosed either antenatally or within the first year of life, and 60% in patients younger than 10 years.[6,15,16] In series reporting both adult and pediatric cases, 40% to 80% of patients were younger than 12 years.[4,9,17] Although BDCs have been reported in an increasing number of patients in recent studies,[10,18] this multicenter series of 42 patients culled during a 12-year period emphasizes the rarity of this disease in Western Europe.

The Todani classification, proposed in 1977,[12] has become the reference. This classification distinguished three subtypes in type I with extrahepatic bile duct dilatation and identified a type IVA with both extra- and intrahepatic deformities. However, Todani provided no precise criteria of fusiform IC dilatation, leading to possible confusion with large dilatation of the common bile duct secondary to stone obstruction.

Formes frustes of choledochal cysts have been reported in children.[19] In adults, this ambiguous concept must be rejected, and clear patterns are required to delineate the patients who can be defined as having true fusiform IC bile duct cysts. To differentiate these patients from those with biliary dilatation secondary to sphincter of Oddi stenosis, we propose the following criteria, used in the five patients with fusiform dilatation excluded from our study: absence of prior history of gallstones or biliary surgery, a common bile duct diameter >30 mm, and APBDJ demonstrated by ERCP or PTC.

The prevalence and prognostic importance of intrahepatic dilatation were first underscored by Tsuchida et al. in

1971.[20] The 50% incidence reported in this European multicenter study is within the 35% to 83% range reported in recent series[10,20–23] but is higher than the figure of 18.9% reported by Yamaguchi in his collective Japanese review.[4] Improvements in diagnostic techniques, with extensive use of computed tomography, PTC, and surgical cholangiography, and closer attention to the radiologic delineation of bile duct anatomy probably explain this difference.

Using surgical cholangiography, Tsuchida et al.[20] distinguished two types of intrahepatic bile duct dilatations: sacciform or cystic, and fusiform or cylindrical. After cyst excision, Todani et al.[24] in adults, and Joseph[8] and Ohi et al.[22] in two pediatric series, reported complete regression of fusiform dilatations, supporting the hypothesis that this type of intrahepatic dilatation might be secondary, not congenital. In contrast, cystic deformities persisted with further complications, such as recurrent cholangitis or intrahepatic stones.

Consequently, attention must be paid to the configuration of the intrahepatic biliary tree. In large cystic dilatations of extrahepatic ducts, ERCP frequently visualizes the intrahepatic biliary tree incompletely because of dilution of contrast material. PTC and surgical cholangiography, in which contrast material can be injected under pressure, are needed to delineate the anatomy of the intrahepatic biliary tree.

In this respect, Todani's classification should be modified because it does not differentiate segmental, cystic, and fusiform type IVA intrahepatic deformities, and it includes associations of multiple extrahepatic cysts with a normal intrahepatic biliary tree as type IVB.

As a result, we propose modifying Todani's classification. The exceptional Todani's type IVB could be shifted to type ID, and type IV should be exclusively devoted to dilatations of both the extra- and intrahepatic bile ducts, with three subtypes: IVA with cystic, IVB with segmental, and IVC with fusiform intrahepatic bile duct dilatations, as in Todani's type I (see Fig. 1).

Babbitt,[25] Kimura et al.,[13] and others have shown that most patients with BDC had an APBDJ outside the duodenal wall, distant from the ampulla of Vater. The incidence of APBDJ with a common channel >15 mm ranges from 96% to 100% in pediatric series[2,26] and from 68% to 94% in adult series.[11,13,27,28] Our data are consistent with these figures but did not confirm the relation between the type of APBDJ (CP or PC junction; see Fig. 2) and the extent of choledochal dilatation, as reported by Iwai et al.[7] and Kimura et al.[13] Moreover, the constellation of multifaceted pancreatic anomalies in our series (pancreas divisum, left pancreatic atrophy, "curled" main pancreatic duct, as illustrated in Fig. 3) provides additional evidence that BDC may represent a part of a wide spectrum of pancreaticobiliary disorders.[8]

Kasai et al.[1] were the first to report an increased incidence of cancer associated with BDC. Similarly, large series of adult patients have shown that the incidence of hepatobiliary malignancies associated with BDC could be as high as

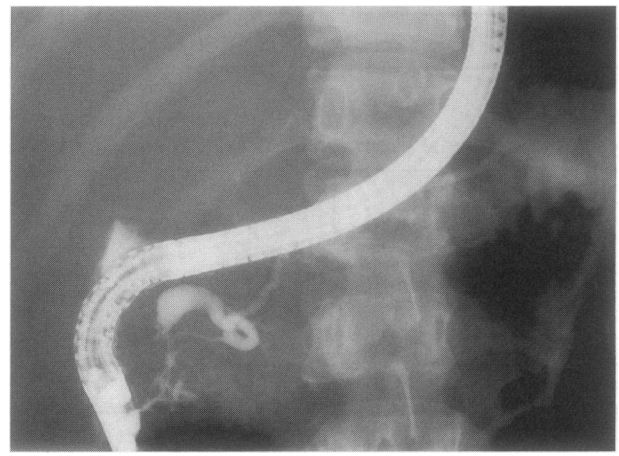

Figure 3. Postoperative ERCP of a "curled." Note the absence of biliary opacification after complete extrahepatic cyst excision.

28%.[11,28–31] The incidence of 12.5% found in our multicenter series is consistent with the figure of 17.5% reported by Todani et al.,[31] who collected data from 73 institutions in Japan, but is higher than the 0.01% to 0.38% incidence noted in large autopsy series in the normal population.[29,32] In patients with BDC, the age-related incidence of cyst-associated cancer has been shown to increase from 0.7% in the first decade of life to 14.3% after age 20 years.[33]

Of the 241 cases of carcinoma collected by Todani et al.,[31] 45 (18.6%) were found in patients with previous cystenteric drainage procedures. Carcinoma arose mainly from the cyst in the drainage group, whereas "primary" cancers developing in the absence of previous cyst drainage procedures were either in the cyst or the gallbladder. In that series, the mean age at which cancer was detected was 35, which is 15 years earlier than that of "primary" carcinoma, and the mean interval between the bypass procedure and the detection of carcinoma was 10 years.[31] Our data are not consistent with these figures (see Table 4), because the mean age of patients with prior cystenterostomy and malignancy was only 1.7 years younger than the age of patients with "primary" carcinoma. Of note, three of five patients with biliary carcinoma had undergone previous cystenteric drainage procedures. These figures support the recommendation of Stain et al.[36] that low-risk patients should undergo elective excision of asymptomatic BDCs previously treated by cystenterostomy.

However, hepatobiliary malignancies are not completely prevented by cyst excision. At least nine cases of cancer developing at the anastomotic site or in the intrahepatic bile duct wall after apparently complete cyst excision have been published.[8,11,35] At least three other cases of carcinoma arising in the distal common bile duct after incomplete retropancreatic cyst excision have been reported.[35]

Treatment of BDC by cystenterostomy is no longer recommended. This procedure often fails to avoid bile stasis, leaving patients at high risk for recurrent cholangitis and liver abscess,[9,11,16,17,28,34] secondary biliary cirrhosis, or

carcinoma arising in the cyst wall.[9,12,18,23,31] Total extrahepatic cyst excision is now the preferred method, as either a primary or a secondary procedure after failure of internal drainage. Even in this clinical setting, this procedure can be performed successfully without increased risk of death or complications,[10,17,18,34] as demonstrated in our experience.

Presurgical (ERCP, PTC) and surgical cholangiographies are essential in planning the surgical strategy because they provide precise information on the anatomy of extra- and intrahepatic bile ducts, the type of the cyst, and the situation and abnormalities of the pancreaticobiliary ductal junction.

Long-lasting cure can be anticipated if three conditions are fulfilled by surgical treatment: restoration of normal bile flow, abolition of pancreatic reflux, and reduction of the risk of malignancy by removing the most common sites of malignant transformation (*e.g.*, the cyst wall or the gallbladder).

In type I cysts, complete extrahepatic cyst excision achieves these goals. Posteriorly, the cyst wall is generally easy to free from the portal vein and hepatic artery. In the present study, the technique described by Lilly[36] was never used, even in patients who had undergone prior surgery.

Distally, the retropancreatic portion of the cyst must be completely excised to prevent malignancy arising in the residual cyst remnant. The use of the Whipple procedure, as proposed by Todani et al.,[21] in patients with severe inflammation of the cyst wall extending to the head of the pancreas is not justified, in our opinion. In this setting, as in large cystic dilatation, the cyst wall can be transected 4 or 5 cm above the head of the pancreas, leaving an "egg-cup bottom," which is then removed by intramural dissection. This technique prevents bleeding and pancreatic duct injury. The outlet biliary tract is generally tiny and easy to suture. Superiorly, the division of the biliary tract must be carried out in healthy biliary tissue. Biliary reconstruction is performed with a long defunctionalized Roux limb anastomosed to the transected common hepatic duct or, more frequently, at the upper biliary convergence after opening the hepatic ducts. In type I cysts, extrahepatic cyst excision, performed according to these technical standards, reduces the risks of postsurgical anastomotic stricture or malignancy and provides gratifying long-term results (see Table 5).[1,10,17,18,28]

In type IV cysts, success with this surgical approach is more uncertain. In recent series focusing on the long-term results according to cyst type, late biliary complications have been demonstrated in 23% to 44% of patients with intrahepatic involvement.[10,35,37] In this setting of intrahepatic bile duct dilatations, the incidence of recurrent cholangitis was 31% in our series (see Table 5).

Recurrent cholangitis can occur as a result of anastomotic stricture, but infection and intrahepatic stones are seen in patients with patent anastomoses as well.[10,37,38] Chijiiwa et al.[37] could not find any correlation between the occurrence of postsurgical delayed biliary complications and the diameter of the anastomosis. As observed in Caroli's disease,[39] specimens of hepatic resection performed for secondary hepatolithiasis have demonstrated that intrahepatic cystic dilatation may be cut off from the biliary tree, leading to bile infection and intracystic stone formation. In the present study, the three patients with severe late postsurgical cholangitis and intrahepatic stones had bilateral or segmental (left-sided) intrahepatic dilatations of the cystic type (see Table 5).

Anastomotic stricture complicating cyst excision can be treated by surgical revision,[3,35,38–40] as for one patient in our series, or by percutaneous transhepatic dilatation.[10,34,37] Treatment of hepatolithiasis by transhepatic endoscopic lithotripsy and percutaneous stone removal[10,37,38] requires repeated hospital admissions and treatment courses. Hepatolithiasis in segmental intrahepatic dilatations is more radically cured by segmental liver resection[34,37,38] or hepatic lobectomy.[10,35,37,38] Hepatic transplantation may be required with complex and severe intrahepatic lesions.[10,17]

The high risk of late biliary complications after extrahepatic cyst excision in patients with intrahepatic dilatations, particularly of the cystic type, justifies preventive procedures. A wide, mucosa-to-mucosa anastomosis, extended to the lateral wall of the hepatic ducts, is known to prevent anastomotic stricture,[28,35,37,40] but the primary management of intrahepatic bile duct dilatations is still controversial. Confection of a jejunal loop attached to the anterior abdominal wall[3] or presurgical insertion of a large-bore transhepatic stent that is left in place after surgery[10] does not prevent late biliary complications but makes their treatment easier by the percutaneous route. Todani et al.[21] proposed a wide hepaticoduodenostomy at the biliary convergence to assess the intrahepatic biliary tree endoscopically during follow-up. These procedures can be recommended in diffuse intrahepatic biliary dilatations.

Primary hepatic resection has been proposed in localized left-sided type IV cysts.[21] In our series, primary left lobectomy of the liver was performed in association with extrahepatic cyst excision in two patients with segmental intrahepatic cystic dilatations and provided excellent long-term results without increasing the postsurgical risk. This "extended" cyst excision procedure, combining complete extrahepatic cyst excision and left hepatic lobectomy, is an attractive therapeutic option, because left-sided intrahepatic bile ducts were involved exclusively in 43% of type IV cysts in our experience (see Table 2). A decreased incidence of late biliary complications could be expected if complete cyst excision is performed in patients with segmental intrahepatic dilatations of the cystic type.

Even after complete extrahepatic cyst excision, however, the patient with residual intrahepatic bile duct dilatations requires close long-term follow-up because of the risk of late biliary complications.

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