

Cystic involvement of the roof of the main biliary convergence in adult patients with congenital bile duct cysts: A difficult surgical challenge

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Background. Complete cyst excision of the extrahepatic disease component with biliary reconstruction on proximal healthy bile ducts is considered to be the treatment of choice in patients with congenital bile duct cysts (BDC). Proximal cystic disease that extends to the roof of the main biliary convergence (MBC) might challenge this standard of surgical care.

Methods. A retrospective multicenter study was conducted in 4 European surgical centers concerning their experience with adult patients suffering from type I and IV BDC according to the Todani classification. Clinical presentation, operative management, and postoperative outcome were compared between patients with or without proximal extrahepatic cystic disease that involved at least the roof of the MBC (defined as being BDC with MBC involvement subgroup).

Results. From an overall series of 49 adult patients suffering from type I or IV BDC according to the Todani classification, 7 patients had BDC with MBC involvement (14%). Patient age, clinical presentation, duration of symptoms, associated major coexistent hepatobiliary and pancreatic diseases, and synchronous cancer were not significantly different in these patients compared with a control group of 42 adult patients with BDC without MBC involvement. Incomplete proximal cyst excision rate was 86% in the cases of BDC with MBC involvement. Early and late postoperative results were similar in BDC with MBC involvement and in the control group of adult patients, but the incidence of subsequent cancer was significantly higher in the BDC with MBC involvement group (29% vs 0%; $P < .02$).

Conclusion. BDC that involves the roof of the MBC is a real surgical challenge to obtain complete proximal cystic disease excision. As suggested in this small study, primary incomplete excision of this particular form of BDC might expose the patient to the risk of subsequent cancer, a feature that must be confirmed in larger series. (*Surgery* 2007;141:187-95.)

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BILE DUCT CYSTS (BDCs) are infrequent congenital anomalies of the biliary tree that are characterized by cystic dilatation of the extra- and/or intrahepatic bile ducts. Disease occurrence after childhood is uncommon. However, approximately 25% of BDCs go undiagnosed into adulthood.^{1,2} BDCs are classified

according to the site, extent, and shape of cystic biliary anomalies. The modification of Todani et al³ of the classification of Alonso-Lej et al⁴ is the most commonly used (Fig 1). Coexistent hepatobiliary and pancreatic (HBP) diseases are associated frequently with BDCs, especially in adults and in patients who have undergone previous cyst-enteric drainage procedures.^{5,6} Moreover, a significant association between BDCs and hepatobiliary malignancy has been reported,^{2,7-9} with an obvious increasing age-related incidence.^{10,11} To obviate the risk of cancer during the natural disease history, complete cyst excision is currently the treatment of choice for the extrahepatic component of the disease, although the optimal treatment of intrahepatic bile duct

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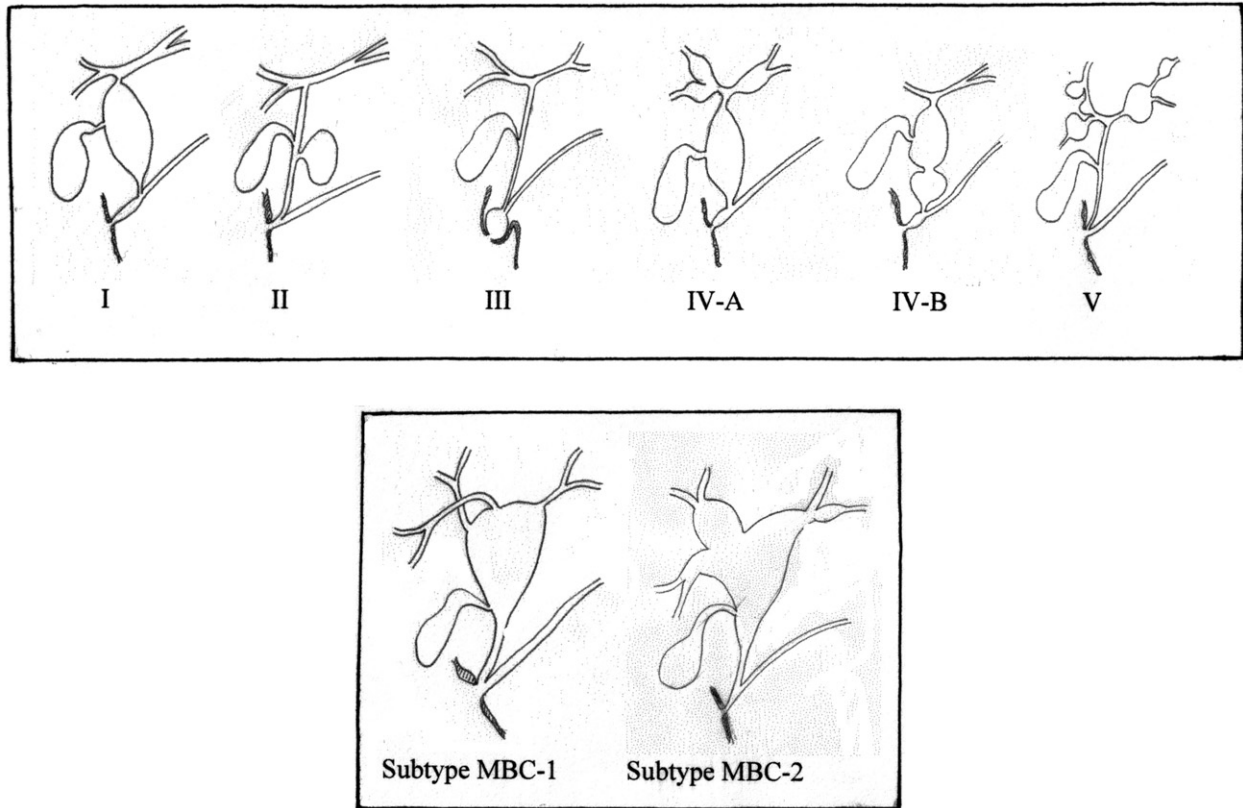


Fig 1. Schematic representation of classification of BDCs (Todani et al³), including types I, II, III, IV-A, IV-B, and V. Additional subtypes according to BDC extent to the roof of the MBC (*subtype MBC-1*) or upward to the secondary divisions (*subtype MBC-2*).

dilatations remains controversial.^{1,5,12} Because 5% to 15% of synchronous biliary malignancies occur outside of the BDC wall and the gallbladder, complete surgical excision of BDCs will not totally prevent the risk of malignancy.^{2,10,13,14} These patients should be candidates for lifelong assessment of late complications or subsequent cancer after BDC resection.¹⁵

When reviewing a multicenter European experience with surgical management of BDCs in adult patients, we identified a subgroup of patients with BDCs with proximal extrahepatic cystic component that involved the roof of the main biliary convergence (MBC) or the secondary biliary divisions. The surgical treatment of these patients might be difficult and may lead to possibly incomplete proximal cyst excision and to biliary reconstruction on nonhealthy bile ducts. Few data have been reported in Western countries concerning these BDCs with MBC involvement. The aim of this study was to assess the difference between patients with BDCs with or without MBC involvement, in terms of clinical presentation, surgical management, and early and long-term results.

PATIENTS AND METHODS

Patients. From 1981 to 2004, 49 consecutive adult patients with congenital type I or IV BDCs, according to the classification of Todani et al,³ were enrolled retrospectively in the present study from 4 European academic surgical centers. The definition of BDCs with MBC involvement was based on 2 combined criteria: (1) the presence of a continuous cystic disease extension from the extrahepatic component of BDCs up to the roof of the MBC or the secondary biliary divisions and (2) a clear demarcation between nondilated peripheral intrahepatic bile ducts (hepatic or sectorial ducts) at their level of entrance into a dilated and diseased main or secondary biliary divisions (Fig 1). This last criterion allowed us to differentiate patients with BDCs with MBC involvement from those defined by Todani et al^{16,17} as type I-C, which has secondary continuous extent of fusiform dilation of the choledochus to peripheral intrahepatic bile ducts, with a return to a normal size of intrahepatic biliary dilatation after surgical excision of the extrahepatic component of BDC (Fig 2). We have defined 2



Fig 2. Intraoperative cholangiography shows a type I-C BDC, by the classification of Todani et al,³ with continuous fusiform extension of the extrahepatic component of BDC to dilated MBC and intrahepatic bile ducts. This type of BDC is different from those reported in Figs 3 and 4.

subtypes of BDC with MBC involvement: cystic dilation that is limited to the roof of the MBC with nondilated hepatic or sectorial bile ducts entering directly into the enlarged roof of the MBC (subtype MBC-1; Figs 1 and 3) and cystic dilation of the MBC that is extended to intrahepatic secondary biliary divisions (left and/or right hepatic ducts; subtype MBC-2), with nondilated peripheral intrahepatic sectorial bile ducts that enter into diseased secondary biliary divisions (Figs 1 and 4). Additionally, intrahepatic bile ducts might be affected more peripherally by the disease in type IV-A BDC (Fig 4). Patients with BDCs without involvement of the roof of the MBC served as the control group.

Complicated clinical presentation was defined as cholangitis, liver abscess, or bile peritonitis from cyst rupture. Major coexistent HBP diseases were defined as the presence of chronic liver disease; biliary or pancreatic ductal strictures; intrahepatic, common channel, or pancreatic ductal stones; and synchronous carcinoma. Radiographic studies included percutaneous ultrasound in 94% of patients, computed tomography in 73% of patients, endoscopic retrograde cholangiopancreatography in 59% of patients, magnetic resonance cholangiopancreatography in 37% of patients, and percutaneous transhepatic cholangiography in 20% of patients.

Methods. During the entire study period, the standard surgical treatment for patients with types I and IV BDCs (Todani et al³) included the attempt of complete excision of the extrahepatic portion of



Fig 3. Intraoperative cholangiography of a BDC with cystic involvement to the roof of the MBC, with nondilated left hepatic and right sectorial bile ducts entering directly into the enlarged roof of the cystic-diseased MBC (white arrows; type I BDC [Todani et al³], subtype MBC-1). (With permission of reproduction by MASSON editions.)

the BDCs. In patients with type IV-A BDCs (Todani et al³), excision of intrahepatic cystic disease was indicated in the case of unilobar-complicated intrahepatic disease. Routine intraoperative examination of the cyst wall by macroscopic and frozen-section examinations was performed to rule out the presence of synchronous cancer. Biliary reconstruction was achieved with a 60-cm Roux-en-Y jejunal limb, in an attempt to perform biliodigestive anastomosis on proximal healthy biliary tissues. Performing biliary reconstruction on nondiseased proximal bile duct was considered to be essential to reduce the risk of late anastomotic stricture. Follow-up information was available for 48 of these patients (98%). The median follow-up period was 67 months (range, 20-178 months) for the 7 patients with BDCs with MBC involvement and 51 months (range, 2-255 months) for the control group of patients with BDCs without MBC involvement. During the postoperative follow-up period, patient condition was assessed by clinical, biologic, and radiologic examinations, which included percutaneous ultrasound in 21 patients (44%), computed tomography in 8 patients (17%), and magnetic resonance cholangiopancreatography in 19 patients (39%).

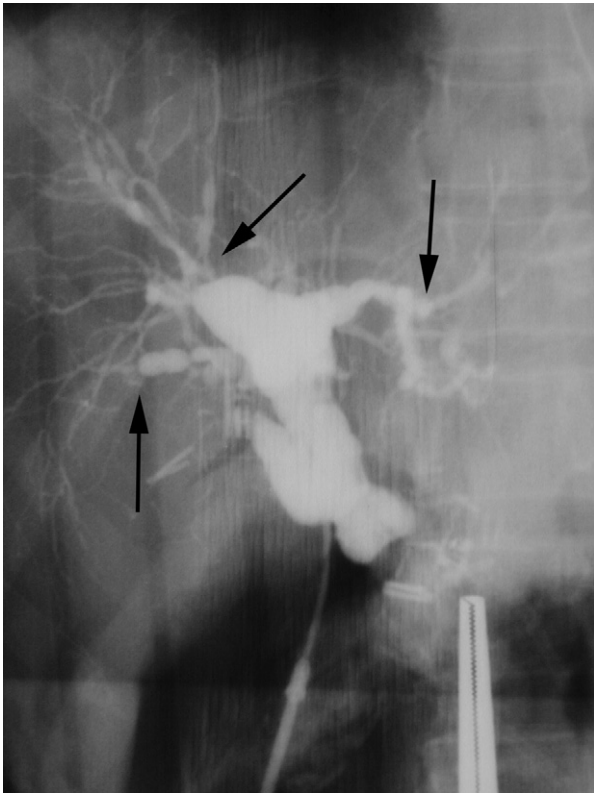


Fig 4. Intraoperative cholangiography of a BDC with fusiform involvement of the MBC and secondary biliary divisions on both sides, with a clear demarcation on the right side between fusiform-dilated diseased main and secondary biliary divisions and nondilated but diseased peripheral intrahepatic bile ducts (black arrows; type IV-A BDC [Todani et al³], subtype MBC-2).

Definition of end points. Evaluation criteria included clinical presentation, coexistent HBP diseases, type and details of the operative procedures, postoperative death, complications and reoperation rates, and late outcome. Long-term results were evaluated according to the previously reported Mayo Clinic score of results evaluation.⁵

Statistical analysis. Statistical analysis included the use of Fisher's exact test and the Mann-Whitney test when appropriate. A probability value of $<.05$ was considered statistically significant.

Results

Patients. BDCs with MBC involvement was observed in 7 of 49 adult patients (14%) (Table I). There were 6 female and 1 male patients, with a median age of 26 years (range, 17-47 years). Two female patients were Asian. All patients were symptomatic, which included abdominal pain in 5 patients, cholangitis in 2 patients, jaundice and hepatomegaly in 1 patient each. Median patient age at examination (26 vs 41 years; $P < .07$), duration

of symptoms (4 vs 10 months; $P =$ not significant), and occurrence of complicated clinical presentation (2 of 7 patients vs 21 of 42 patients; $P =$ not significant) were not significantly different compared with the control group of patients, respectively.

Disease. According to the classification of Todani et al,³ there were 3 patients with type I BDCs, 3 patients with type IV-A BDCs, and 1 patient with type IV-B BDCs, which represented 9% of type I and 23.5% of type IV BDCs. There were 3 patients with BDCs with MBC involvement that was limited to the roof of MBC and 4 patients with BDCs with MBC involvement that extended to the secondary biliary division (Table I).

Major HBP coexistent diseases were present in 2 patients (29%), which included diffuse intrahepatic stones and secondary hepatic fibrosis from repeated attacks of cholangitis in 1 patient each. Minor coexistent HBP diseases included gallbladder and intracystic stones in 1 patient each. No synchronous cancer or intrahepatic bile duct stricture was observed in this group of patients. Previous treatment included cholecystectomy and cyst-enteric drainage procedures in 3 and 2 patients, respectively. Presence of major coexistent HBP disease (2 of 7 patients vs 14 of 42 patients; $P =$ not significant), previous cyst-drainage procedures (2 of 7 patients vs 9 of 42 patients; $P =$ not significant), and incidence of synchronous cancer (0 of 7 patients vs 4 of 42 patients; $P =$ not significant) were not significantly different compared with the control patients.

Type of procedures. Extrahepatic cyst excision was performed in all patients, but complete proximal excision of the diseased MBC was achieved primarily in only 1 of the 3 patients with MBC involvement that was limited to the roof of the MBC and in none of the 4 patients with BDC with MBC involvement that extended to the secondary biliary division, which led to an overall rate of complete proximal cyst excision at primary operation of only 14% (Table I). On the contrary, primary complete proximal cyst excision was achieved in all patients in the control group ($P < .0001$). In patients with incomplete proximal excision, the biliary reconstruction was performed on cystic-diseased MBC or cystic-diseased secondary biliary divisions. In the patient with complete proximal cyst excision at primary operation (patient 2 in Table I), biliary reconstruction was achieved by confection of 3 biliary anastomoses to a Roux-en-Y jejunal loop on thin right sectorial ducts and left hepatic duct (Figs 3, 5, and 6). Associated liver resection was not performed in the 3 patients with type IV-A

Table I. Results of patients with BDC that involved the MBC

<i>Patient</i>	<i>MBC involved up to 2-degree biliary divisions</i>	<i>Type of BDC (Todani et al³)</i>	<i>Complete primary BDC excision</i>	<i>Late results and complications</i>	<i>Secondary complete BDC excision</i>	<i>Late reoperation</i>	<i>Duration of follow-up period (mo)</i>	<i>Subsequent cancer</i>
1	No	I	No	Cholangitis; anastomotic stricture at right side	Yes	Secondary anastomotic biliary revision with right secondary convergence excision at 10 mo	67	Intrahepatic; right liver lobe; death
2	No	I	Yes	1 Episode of cholangitis	—	No	40	No
3	No	IV-B	No	None	No	No	20	No
4	Yes	IV-A	No	Suspicion of intrahepatic stones, but patient free of symptoms	No	No	82	No
5	Yes	I	No	Repeated episodes of cholangitis	No	No	84	Extrahepatic (hilum); death
6	Yes	IV-A	No	Cholangitis; intrahepatic stones; biliary cirrhosis	Yes (orthotopic liver transplantation)	Percutaneous transhepatic biliary drainage; reoperation for access loop formation; orthotopic liver transplantation (postoperative death)	41	No
7	Yes	IV-A	No	Cholangitis; intrahepatic stones; liver abscess	Yes	Left hepatectomy with complete MBC excision at 17 months	178	No

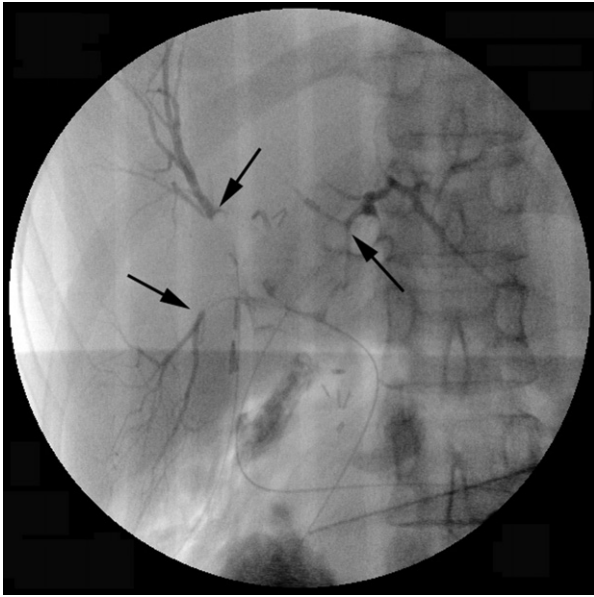


Fig 5. Selective intraoperative cholangiography in the same patient as in Fig 3 after complete resection of BDCs with cystic involvement of the MBC. Thin nondilated and nondiseased intrahepatic bile ducts (*black arrows*) are shown. (With permission of reproduction by MASSON editions.)

(Todani et al³) because of the presence of bilobar disease distribution.

Postoperative complications. 2-month hospital mortality rate was nil in the group of patients with BDCs that involved the MBC and in the control group. Major postoperative complications included intra-abdominal biloma or abscess in 3 patients (43%), whose conditions required percutaneous drainage procedures. The postoperative major complication (3 of 7 patients vs 13 of 42 patients; $P =$ not significant) and reoperation rates (3 of 7 patients vs 8 of 42 patients; $P =$ not significant) were not significantly different compared with the control group.

Late outcome. During a median follow-up period of 67 months (range, 20-178 months), fair or poor results were observed in 4 patients (57%) with BDCs with MBC involvement, a higher but not significantly different rate (38%) from those of the control group. The causes of unsatisfactory results included subsequent cancer in 2 patients (29%), secondary liver cirrhosis and liver abscess from intrahepatic stones in 1 patient each. No subsequent cancer was observed in the control group during a median follow-up period of 51 months (range, 2-255 months; $P < .02$). Late nonsurgical or surgical disease-related re-operative procedures were required in 3 patients (43%; Table I), which was not

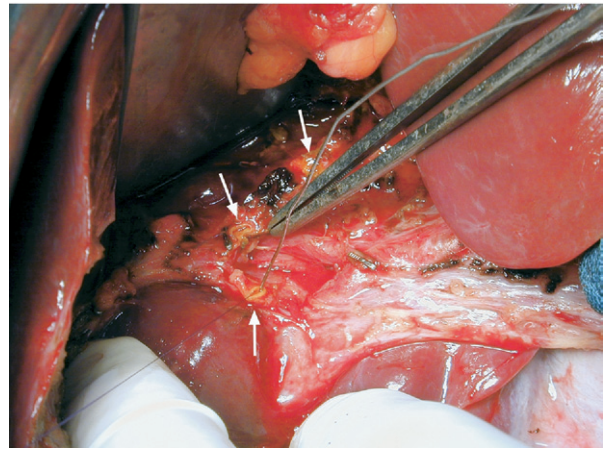


Fig 6. Operative view in the same patient as in Figs 3 and 5 before biliodigestive reconstruction that required multiple anastomoses on thin, nondilated intrahepatic sectorial bile ducts. *White arrows* show biliary stoma of left hepatic duct and right sectorial bile ducts. (With permission of reproduction by MASSON editions.)

significantly different from the control group (13 of 42 patients). One patient with BDC that involved MBC up to the secondary biliary divisions with late repeated attacks of cholangitis had metastatic subsequent hilar cancer 84 months after primary incomplete proximal cyst excision (Fig 7). Percutaneous transhepatic cholangioscopic examination demonstrated the absence of anastomotic stricture and allowed biopsies of a hilar carcinoma that was located at the proximal remnant of cystic MBC. One patient with BDC that involved the roof of MBC underwent anastomotic revision 10 months after primary incomplete proximal cyst excision on the right secondary biliary division (the primary biliodigestive anastomosis had been performed on a small patch of diseased biliary epithelium) and experienced metastatic subsequent carcinoma 57 months after secondary complete proximal cyst excision during secondary anastomotic biliary revision. However, subsequent carcinoma occurred in this patient in the posterior sector of the right liver, far away from the biliodigestive anastomosis. The patient with type IV-A BDC with late secondary liver cirrhosis died of acute graft rejection after liver transplantation. The remaining patient with type IV-A BDC with left liver lobe abscess underwent successful left hemihepatectomy and hilar resection; secondary complete proximal cyst excision was achieved. Finally, if patients with synchronous cancer are excluded, 3 of 7 patients with BDCs with MBC involvement died of disease-related causes (subsequent cancer in 2 patients and death from

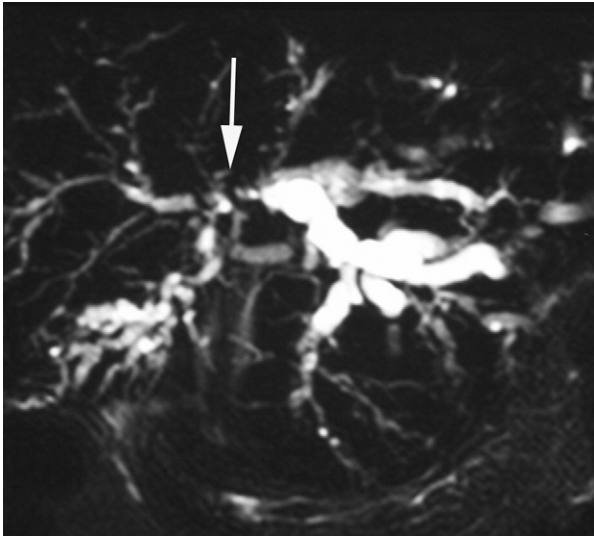


Fig 7. Magnetic resonance cholangiography in a patient with proximal incomplete resection of a complex BDC that involved the MBC; 84 months later, a hilar cholangiocarcinoma at the level of the remnant diseased MBC (*white arrow*) was seen (patient 5 in the Table).

secondary liver transplantation in 1 patient) compared with none of the control group ($P = .0019$).

Discussion This study emphasizes the existence of complex forms of congenital BDCs that involve the MBC, a feature that is not so rare in adult patients (14%). BDC with MBC involvement represents a real surgical challenge to achieve complete proximal cyst excision at the primary operation and may predispose to an increased risk of subsequent cancer.

Factors to consider for the surgical treatment of patients with BDCs include age, symptoms and complications, coexistent HBP diseases, previous cyst-related surgical procedures, cyst type on imaging studies, and age-related risk of synchronous malignancy. When disease extent throughout the whole biliary tree is taken into consideration, the cystic involvement up to the roof of the MBC has not yet been considered to be a major factor that affects the surgical treatment of patients with BDCs. Indeed, the most commonly used classification of Todani et al³ of BDCs mainly considers the site, the extra- and/or intrahepatic extent and the shape (cystic or fusiform) of biliary dilatations. However, Todani et al¹⁶ have emphasized the existence of a secondary continuous extent of fusiform dilatation of the choledochus to the intrahepatic bile ducts, called type I-C, with return to a normal size of intrahepatic biliary dilatation after surgical excision of the extrahepatic component of BDCs (Fig 2), thus not requiring resection. Such regres-

sion of intrahepatic fusiform dilatations soon after extrahepatic cyst excision have been confirmed in 2 pediatric BDC series,^{18,19} which supports the hypothesis that this type of intrahepatic dilatation might be secondary and not congenital. Additionally, Todani et al¹⁷ have also made a special reference to the importance of associated proximal ductal biliary stricture that causes dilatation that extends continuously to the intrahepatic ducts and allows us to distinguish fusiform type I-C BDCs with continuous intrahepatic bile duct dilatation from type IV-A BDCs with true intrahepatic disease extent. The group of BDCs with MBC involvement that are reported in the present series is clearly different from those patients. Indeed, peripheral intrahepatic bile ducts, despite being sometimes more peripherally diseased in type IV-A BDCs (Todani et al³), were not dilated at their level of entrance within the main or secondary dilated biliary divisions. The clear demarcation between nondilated intrahepatic bile ducts and the MBC disease extent leads us to the suspicion that the MBC itself is truly affected by the disease-related dilatation.

We did not find any specific features that were able to predict the presence of BDCs with MBC involvement, even when we considered the type of clinical presentation, coexistent HBP diseases, or a previous surgical biliary history. Only imaging studies were able to demonstrate such MBC disease involvement. Magnetic resonance cholangiopancreatography, which is a noninvasive imaging study that was performed in 37% of the patients in the present series, tends to become the current most accurate preoperative examination to assess cyst anatomy; to identify size, site, and shape of bile duct dilatation; and to detect pancreatobiliary malunion, without the risks of pancreatitis and cholangitis that are associated sometimes with endoscopic retrograde cholangiopancreatography.²⁰ Direct opacification of the biliary tree by endoscopic retrograde cholangiopancreatography, percutaneous transhepatic cholangiography, or intraoperative cholangiography are, however, highly efficient means with which to determine the exact degree of disease extent of the MBC.

Determination of MBC involvement by BDC is not only useful for precise anatomic classification but also is essential when the necessity of complete proximal excision of the cystic dilatation is considered. Indeed, the presence of MBC involvement in such complex BDCs was associated in the present series with a significantly higher rate of incomplete proximal cyst excision, in comparison with the control group without MBC involvement in which com-

plete proximal cyst excision was achieved in all patients with low operative mortality and morbidity rates. To achieve such a goal, we recommend, from a technical point of view, the routine use of extended hilar plate lowering on both sides of the hilum, of meticulous surgical dissection close to the cystic MBC wall, and of isolation of the main or the sectorial hepatic ducts at the roof of the MBC, by liberal use of atraumatic dissection with an ultrasonic dissector. Then, biliary reconstruction requires meticulous confection of multiple biliodigestive anastomoses, which usually is performed on thin, nondilated intrahepatic bile ducts. This approach sometimes represents a difficult technical challenge even for experienced HBP surgeon. We recommend, in such conditions, the use of temporary transanastomotic stenting to be able to assess postoperatively the quality of these fine biliodigestive anastomoses that are exposed to a risk of anastomotic stricture. In 86% of the patients with BDCs with MBC involvement in the present series (especially in patients with BDCs with MBC involvement up to the secondary biliary divisions), complete proximal cyst excision was considered initially not to be technically possible or to be too dangerous because of proximal disease extension to the secondary biliary divisions or in case of type IV-A BDC (Todani et al³) with residual bilobar intrahepatic disease. It should be emphasized that late complications in patients with type IV-A BDCs (Todani et al³) with MBC involvement in the present series were more often related to residual intrahepatic disease rather than to incomplete MBC excision. However, an attempt at complete proximal cyst excision should be recommended strongly in the cases of BDCs with MBC involvement but without intrahepatic disease (type I or IV-B BDC [Todani et al³]) at the time of primary operation, because 2 of 3 such patients in the present series have experienced late complications that have been related to incomplete MBC excision. These late complications included anastomotic stricture on the right side of a multiple biliodigestive anastomosis (the biliary anastomosis was performed on a small patch of diseased biliary tissues during the primary operation) and hilar subsequent carcinoma. Both complications clearly appear to be related closely to retained biliary cyst epithelium. Additionally, the patient whose condition required anastomotic revision that allowed secondary complete proximal cyst excision also experienced the development of metastatic subsequent cancer in the posterior part of the right liver, far away from the biliodigestive anastomosis. The location of subsequent cancer in this patient illustrates that the whole biliary epithelium is at risk of

malignant transformation in patients with BDCs. Indeed, the risk of subsequent carcinoma after the excision of BDCs is well documented in the literature.^{5,7,15,21-25} In a large review series, the incidence of such subsequent cancer in the Japanese population was assumed to be 0.7%.¹⁵ These subsequent cancers can develop throughout the whole biliary tree, including intrahepatic bile ducts,^{6,15,22,23,26-30} the hilum or the biliary anastomosis,^{1,15,21,22,31} the distal intrapancreatic common bile duct,^{7,15,32} and the head of the pancreas.^{24,25} Watanabe et al¹⁵ reported a review of 23 patients who experienced subsequent cancer after BDC resection and found that nearly one half of these patients had carcinoma within the residual proximal or the distal portion of incompletely excised BDCs. They also emphasized that these subsequent carcinomas can occur up to 19 years after the initial procedure. Accordingly, complete cyst resection is recommended strongly, even in the presence of BDCs that involve the MBC. But, on the contrary, Ishibashi et al³³ reported incomplete proximal or distal BDC resection in 28 patients with no further malignancy during a mean follow-up period of 9.1 years. However, as have other investigators,⁷ we consider that, even knowing that subsequent carcinoma can develop outside the cyst, we must not reduce the efforts at total resection that should be made during the primary operation.

In conclusion, BDC that involve the MBC represents a real surgical challenge to achieve complete proximal cystic disease excision. In the case of incomplete resection, this particular form of BDC appears to be associated in this series with a high risk of late complications. In addition to the factors that have been taken into consideration by the classification of Todani et al,³ we suggest the inclusion of MBC involvement by BDCs as a critical factor that affects surgical management. Larger reported experiences with this particular form of BDC are required to confirm the potential risk of subsequent malignancy.

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