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ORIGINAL SCIENTIFIC REPORT WITH VIDEO

Pancreaticobiliary Maljunctions in European Patients with Bile Duct Cysts: Results of the Multicenter Study of the French Surgical Association (AFC)

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Abstract

Background Pancreaticobiliary maljunctions (PBMs) are congenital anomalies of the junction between pancreatic and bile ducts, frequently associated with bile duct cyst (BDC). BDC is congenital biliary tree diseases that are characterized by distinctive dilatation types of the extra- and/or intrahepatic bile ducts. Todani's types I and IVa, in which dilatation involves principally the main bile duct, are the most frequent. PBM induces pancreatic juice reflux into the biliary tract that is supposed to be one of the main factors of biliary cancer degeneration, although the diagnostic criteria of PBM that can be either morphological and/or functional are not well defined especially in Western series.

Objective The aim of this study was to assess the relative prevalence of PBM in BDC in a large European multicenter study, to analyze the characteristics of PBM and try to propose diagnostic criteria of PBMs based on morphological and/or functional criteria and define the positive, negative predictive values, sensibility and specificity of either criteria.

Results From 1975 to 2012, 263 patients with BDC were analyzed. Among them, 190 (72.2 %) were considered to present PBM. Types I and IVa had a similar rate of PBM association. According to the "AFC classification," 57.2 % had a C–P type, 34.5 % a P–C type and 8.3 % a complex type ("anse-de-seau"). The median length of the common channel in patients with PBM was 15.8 ± 6.8 mm (range 5–40 mm). The median intrabiliary amylase and lipase levels were 65,249 and 172,104 UI/L, respectively. For the diagnostic of PBM, a common channel length of more than 8 mm and an intrabiliary amylase level superior to 8000 UI/L were associated with a predictive positive value and a specificity of more than 90 %. Synchronous biliary cancer had an incidence of 8.7 % in all patients with BDC and PBM 11.1 % in adults. Compared to type IV, the type I BDC was associated with statistically more cancer patients in the presence of PBM.

Conclusions Characteristics of PBM associated with BDC in Western population are quite close to reported Eastern series. The results suggest considering both the intrabiliary value of amylase >8000 UI/L and a length of a common channel >8 mm as appropriate values for positive diagnosis of PBM.

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Abbreviations

AFC	Association Française de Chirurgie
ASA	American Society of Anesthesiologists
BDC	Bile duct cyst
C–P	Choledochal to pancreatic
ERCP	Endoscopic retrograde cholangiopancreatography
HBP	Hepato-biliary and pancreatic
MRCP	Magnetic resonance cholangiopancreatography
NPV	Negative predictive value
P–C	Pancreatic to choledochal
PBJ	Pancreaticobiliary junction
PBM	Pancreaticobiliary maljunctions
PPV	Positive predictive value

Introduction

Bile duct cyst (BDC) is congenital anomalies of the biliary tree characterized by distinctive dilatation of the extra- and/or intrahepatic bile ducts in which most frequent types are Todani's types I and IVa (more than 85 % of patients) [1–4]. They are often associated with pancreaticobiliary maljunctions (PBMs) [1–5]. PBMs are congenital anomalies of the junction between pancreatic and bile ducts defined by a union between the pancreatic and the biliary ducts located outside the duodenal wall with a common channel [6]. PBMs induce pancreatic juice reflux into the biliary tract that is supposed to participate in BDC formations and the development of biliary cancer especially in dilated segments [7].

BDC above 100–1000 times lowers incidence in Western series [3, 8, 9]. Most Western series reported adult BDC and described a less association with PBM compared to Eastern series [10]. PBMs are relatively unknown in Western centers. The diagnosis of PBM, at least in Western countries, is not consensual and includes morphologic (MRCP/ERCP) and functional (intrabiliary amylase level) criteria. The aim of this study was to assess the relative prevalence of PBM in BDC in a large European multicenter study, to analyze the characteristics of PBM and try to propose diagnostic criteria of PBMs based on morphological and/or functional criteria and to define the positive, negative predictive values, sensibility and specificity of either criteria.

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Patients and methods

Study population and patients' data collection

A multicenter retrospective study on the surgical management of BDC was performed under the auspices of the French Association of Surgery (AFC). Medical records of 505 patients from 31 different European centers (mostly French), including demographic data, prior surgical interventions including hepato-biliary and pancreatic (HBP) diseases, clinical symptoms, biochemical and imaging studies (MRCP, ERCP or cholangiography), operative data, pathology reports, duration of follow-up and long-term outcomes, were retrospectively collected using an online computerized standardized questionnaires (http://www.chirurgie-viscerale.org). If necessary, additional data were obtained from e-mail, mail exchanges or phone calls with the referral physicians in different participant centers.

All operative, pathological and imaging data were consistently reviewed by 3 senior co-authors (JYM, JFG and RK). If patients' data were unavailable and/or not interpretable, the patient was excluded for further analyses. Diagnosis of PBM was made on two strict criteria: (1) the presence of an evident extraduodenal junction between the choledochus and the main pancreatic duct and/or (2) the presence of an abnormal long common channel >10 mm.

BDC subtypes were classified according to Todani's classification based on imaging studies [2] and PBM subtypes to Komi's and Kimura's classification [6, 10]. Then, to simplify the data analyses and lectures, patients with PBM were classified according to a simplified Komi's classification, so-called AFC classification that included 3 subtypes: type I (C–P type), type II (P–C type) and type III complex type with "anse-de-seau" (Fig. 1).

PBMs were diagnosed either by magnetic resonance cholangiopancreatography (MRCP) in 46.2 % patients or by intraoperative cholangiography in 51.4 % of patients. Endoscopic retrograde cholangiopancreatography (ERCP) made the diagnostic of PBM in 12.7 % of the patients.

The length of the common channel was obtained either by reported data or by revision of available images. As reported recently, there was no associated PBM with type V in the global series; therefore, all patients with BDC type V (Caroli's disease) were excluded from the final analyses of the present series [11]. Finally, 350 patients having Todani's BDC types Ia, Ib, Ic, type II, type III, type IVa and type IVb were studied for morphological and functional analyses. To exclude false-positive values of intrabiliary amylase, patients with previous endoscopic sphincterotomy ($n = 37$) and/or cystenterostomy ($n = 14$) were excluded from the functional data analyses.

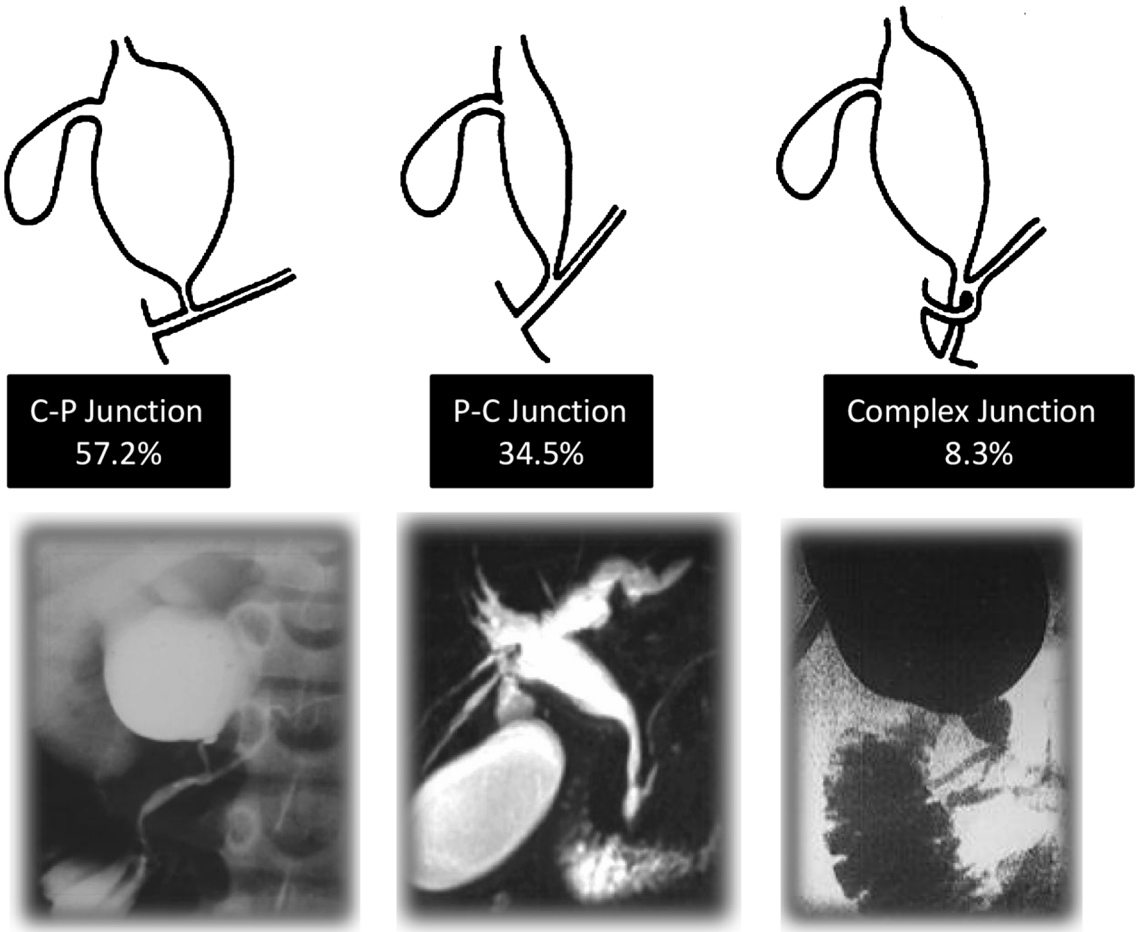

Fig. 1 Simplified AFC classification (derived from Komi's classification). C–P choledochus attempts to pancreatic duct mostly perpendicularly with a relatively large angle, and P–C pancreatic duct attempts to the end of choledochus mainly by a short angle. Type C–P or P–C with the presence of any accessory pancreatic head duct "anse-de-seau"

Statistical analysis

Data were expressed as median (with range). The Chi-squared test was used to compare categorical variables when appropriate (the Fisher's exact test was used when conditions for Chi-square test were not fulfilled). The Student's *t* test was used to compare continuous variables when appropriate (in case of non-normality, the Mann–Whitney nonparametric rank sum test was used). When p was <0.15, the data were entered in a multivariate logistic regression. The log-rank test was used to compare subgroups of patients. Statistical analyses were performed using SAS® version 9.2 (SAS Institute Inc, Cary, North Carolina, USA). A p value <0.05 was considered statistically significant.

Results

Pancreaticobiliary junction (PBJ) was analyzable in 263 patients with BDC (sex ratio W/M: 3.3). Characteristics of the population are presented in Table 1. Among them, 190 (72.2 %) were considered having a PBM and 73 (27.8 %) a normal junction.

In patients with PBM ($n = 190$), the median age was 29 years (range 0.1–91). They were 127 (66.8 %) adults and 63 (33.2 %) children (aged <15 years old).The most frequent symptoms were abdominal pain (64.3 %), jaundice (24.7 %), cholangitis (24 %) and pancreatitis (23.6 %).

According to our classification (Fig. 1), 57.2 % had a C–P type, 34.5 % a P–C type and 8.3 % a complex type "anse-de-seau." The median length of the common channel in patients with PBM was 15.8 ± 6.8 mm (range 5–40 mm). Among them, children had a shorter length of the common channel (13.8 ± 5.3 mm, range 8–20 mm vs. 16.8 ± 5.9 mm range 5–40 mm). In patients with PBM, 97.6 % had a common channel >6 mm and 90.5 % >10 mm. For positive diagnosis of PBM, when considering an abnormal length of common channel >8 mm, the sensibility and specificity rates were 97.6 and 80 %, respectively, while positive predictive and negative predictive values were 99.2 and 57.1 %, respectively (Fig. 2).

Table 1 Characteristic of the studied population

	$N = 263$[a] (%)
Median age	29 [0.1–91]
Sex ratio female/male	3.5
Adult/children ratio	186/77 (2.42)
Symptom	
Abdominal pain	169 (64.3 %)
Jaundice	65 (24.7 %)
Cholangitis	63 (24 %)
Pancreatitis	62 (23.6 %)
BDC Todani's type	
Type I	185 (70.3 %)
Ia	87
Ib	22
Ic	75
Type II	12 (4.6 %)
Type III	9 (3.4 %)
Type IVa	52 (19.8 %)
Type IVb	5 (1.9 %)
Pancreaticobiliary junction	
PBM	190 (72.2 %)
Normal (no PBM)	73 (27.8 %)

[a] Patients in whom pancreaticobiliary junction could be correctly evaluated

In patients with assumed positive diagnostic of PBM on imaging, the median intrabiliary amylase level was 30,932 UI/L (range 5–506,000 UI/L). When considered together, a common channel >8 mm and an intrabiliary amylase level >8000 UI/L were associated with a predictive positive and specificity values >90 % (Table 2, Fig. 3).

The association of PBM and the Todani's BDC types was 78.4 % for types I and IVb, 16.6 % for type II, 33.3 % for type III and 19 % for type IVa (Table 3). P–C type was more frequently associated with Todani's type Ic classification and C–P type with Todani's type Ia (Fig. 4). Patient's characteristics between the two groups are presented in Table 4. Patients without PBM were more aged (40.9 vs. 28.2 years, $p < 0.0001$).

In the whole series, 27 out of 350 patients developed a biliary cancer (8 %), none children and 11.1 % in adults. Previous enterocystic derivation was present in 6/27 patients with cancer patients, but only 3 of them had an analyzable PBJ (lack of reliable documents).

The median age of patients with cancer after cystenterostomy was 43.1 years (range 25–60 years). Among 26 patients with simultaneous cancer, 19 were associated with a BDC type I and 7 with type IVa. Characteristics of the patients with synchronous cancer among types I and IVa are presented in Table 5.

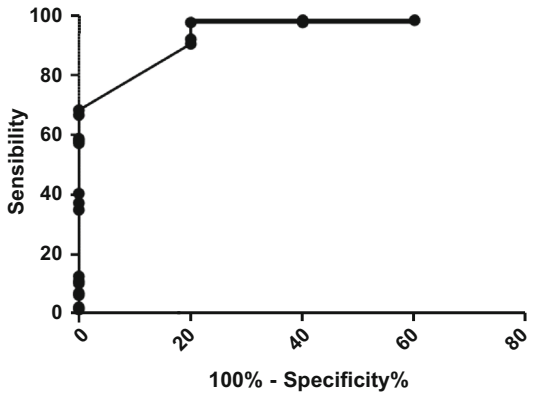

Fig. 2 ROC's curve determining the minimally length of common channel for diagnosis of PBM. ROC's curve/AUC = 0.9476

In cancer patients with analyzable PBJ, 11 had a PBM and 5 had normal PBJ ($p < 0.05$). Patients with PBM who developed biliary cancer were significantly younger (43.3 vs. 54.2 years, $p < 0.05$). The youngest patient with cancer degeneration was a 22-year woman who developed a synchronous biliary cholangiocarcinoma in a BDC type Ia associated with a PBM type C–P. Importantly, the incidence of biliary cancer among patients with BDC types I and IV with PBM was 83.3 and 25 %, respectively (Table 5). Thus, the rate of association between PBM and type I and type IV BDC in adults was not statistically different 73.3 and 63.9 %, respectively.

Discussion

This series is the most important multicenter Western series focused on the diagnostic particularities of PBM among patients with BDC. The present series founded similar characteristics (age, sex ratio, symptoms and complications) as observed in Eastern series concerning BDC [3, 8, 9, 12]. The reported association rate of 72 % between BDC and PBM confirms this resemblance to Asian series [3, 8]. As expected, BDC types I and IV were more frequently associated with PBM (78 and 69 %, respectively) [9], while Todani's types II and III BDC had significantly lower association rates 16.6 and 33.3 %, respectively [3, 6, 8, 10], and none of the type V (Caroli's disease) presented associated PBM [11].

To simplify the morphological classifications of PBM, we deliberately inspired from Kimura's and Komi's classification using only 3 types of PBM: (1) C–P, (2) P–C or (3) complex PBM [3, 6, 9]. C–P type was present in 57.2 % of BDC, while P–C type was observed in 34.5 % of the patients. Patients with complex PBM especially those with the presence of "anse-de-seau" presented 8.3 % of PBM.

Table 2 Sensibility, specificity, positive predictive value (PPV) and negative predictive value (NPV) of different intrabiliary amylase levels and the length of the common channel in patients with PBM

	Intrabiliary amylase >5000 UI/L (%)	Intrabiliary amylase >8000 UI/L (%)	Intrabiliary amylase >10,000 UI/L (%)	Common channel >8 mm (%)	Common channel >8 mm and intrabiliary amylase >8000 UI/L (%)
Sensibility	74.2	69.7	65.2	97.6	87
Specificity	87.5	100	100	80	93
PPV	98	100	100	99.2	99
NPV	29.2	28.6	28.6	57.1	38

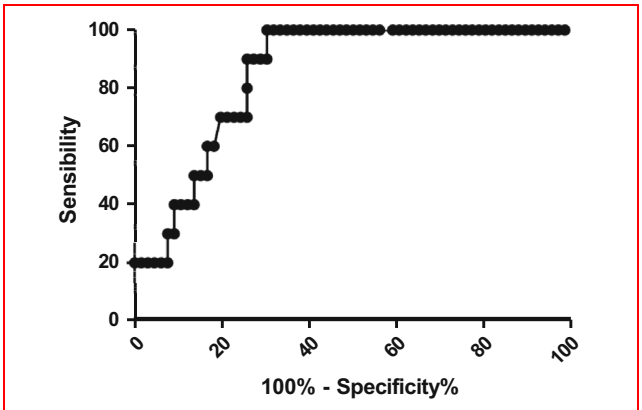

Fig. 3 ROC's curve determining the minimally intrabiliary amylase's level for diagnosis of PBM. ROC's curve/AUC = 0.8523

Thus, C–P type was more frequently observed in the presence of Todani's type Ia BDC, while P–C subtype was more often associated with Todani's fusiform type Ic BDC for which the diagnostic confirmation is usually more difficult especially in previously cholecystectomized patients [13]. Therefore for us, the presence of an evident PBM P–C type in a patient with a slightly large choledochus (close to 20 mm) represents an key argument to retain the diagnosis of Todani's type Ic and thereafter to plan complete resection [14, 15].

Table 3 Association rates between PBM and BDC in patients with analyzable pancreaticobiliary junction

	PBM/evaluable PBJ $N = 190/263$[a]	PBM/evaluable PBJ 72 % (%)
Type I	145/185	78.4
Ia	75/87	86.2
Ib	17/22	77.2
Ic	53/75	70.6
Type II	2/12	16.6
Type III	3/9	33.3
Type IVa	36/52	69.2
Type IVb	4/5	80

[a] None of the type V BDC patients presented PBM (data not reported, see Ref. [5])

In the 10 last years, MRCP was most frequently performed rather than ERCP and intraoperative cholangiography. MRCP was the most useful to detect PBM by showing an extraduodenal wall junction between the choledochus and the main pancreatic duct even in patients with apparently normal common channel. Recent series reported a detection rate of PBM by MRCP ranging from 82 to 100 % [16]. Therefore, rather than ERCP or endoscopic ultrasound, MRCP focused on the head of the pancreas region is actually the first-line method for the detection of PBM in patients with or without BDC [17].

Anatomical studies showed that only 80–85 % of the adults present a short common channel, and when present the mean length of the common channel was 4.6 ± 2.2 mm [18]. In the present series, the mean length of common channel in patients with suspected PBM was 13.8 ± 5 mm significantly greater than 4.6 mm. This confirms Kawisawa et al. [9] proposal to consider the length of the common channel as the main morphological diagnostic tool for PBM. However, there is no consensus among experts for the "cutoff levels" regarding an abnormally long common channel [19]. This length is variable during infancy. The results of the present series confirmed (1) that almost 90 % of the patients (adult and infants) had a common channel length ≥10 mm, and (2) either when taking >8 mm as a cutoff for the "abnormally long" common channel, this presented a high accuracy for the diagnosis of PBM.

Other than morphological tools, functional methods represented by intrabiliary amylase level measurements were easy to perform. In the present series, the mean intrabiliary amylase level in the presence of PBM was significantly higher versus patients without PBM (>50,000 vs. <2000 UI/L, respectively). The literature analysis shows that an intrabiliary amylase level >10,000 UI/L is a significant functional argument to confirm the diagnosis of PBM [20]. Kamisawa et al. [21] demonstrated that a common channel of >5 mm could induce an intrabiliary amylase level >1000 UI/L. As shown in the present series, a high intrabiliary amylase level especially in patients presenting a slightly dilated choledochus, suspicious of presenting a Todani's BDC type Ic (fusiform), can give physicians a strong argument to confirm the diagnosis of BDC with PBM. In such patients, we suggest to measure

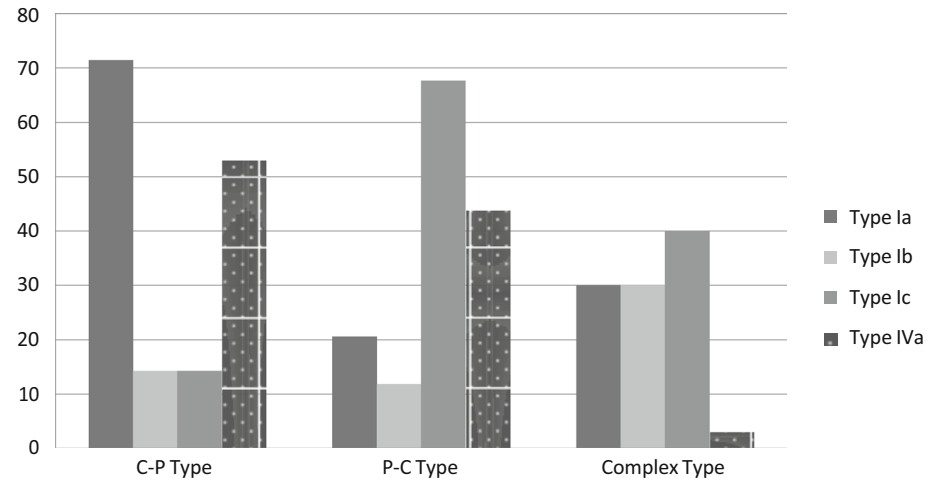

Fig. 4 Association rates between Todani's types I and IVa BDC and different PBM subtypes

Table 4 Characteristics of patients with and without PBM

	PBM $N = 190$	No PBM $N = 73$	p
Median age (ranges)	28.2 [0.1–91]	40.9 [0.5–81]	<0.0001
Sex ratio	3.5	2.8	0.49
Adult/children ratio	127/63 (2.01)	60/13 (4.61)	<0.05
Intrabiliary median values (ranges)			
Amylase UI/L	65,249 [3–506,000]	1787 [1–6600]	0.005
Lipase UI/L	172,104 [4–800,000]	3729 [12–20,400]	0.005
Length of common channel (mm) (range)	15.8 [5[a]–40]	6.5 [5–7]	0.0059

[a] One infant presented an evident extraduodenal pancreaticobiliary junction with a common channel of 5 mm, associated with BDC type I, and high intrabiliary amylase level

Table 5 Incidence of synchronous cancer in types I and IVa with or without PBM in adult patients

	Patients $n = 167$[a]	synchronous cancer $n = 16$[a]	Incidence %
Type I	131	12	9.1
PBM	96 (73.3 %)	10 (83.3 %)	10.5
No PBM	35 (26.7 %)	2 (16.7 %)	5.7
Type IVa	36	4	11.1
PBM	23 (63.9 %)	1 (25 %)	4.3
No PBM	13 (36.1 %)	3 (75 %)	23

[a] Only patients with evaluable PBJ were analyzed

intrabiliary amylase concentration by one of the following techniques: (1) intraoperative sampling of the gall bladder bile during cholecystectomy before any manipulation or cholangiography, (2) preoperatively by fine-needle transhepatic aspiration of the gall bladder or (3) less frequently by protected aspiration of bile during ERCP [18].

Finally, by associating a length of the common channel >8 mm and an intrabiliary amylase level >8000 UI/L, PBM may be accurately diagnosed with sensibility of 87 %, specificity and PPV >90 %. It is notable that the NPV of amylase concentration alone is lower than 30 %. This emphasizes the fact that a normal intrabiliary amylase level alone is not an appropriate way to reject the diagnosis of PBM. In a practical way, when intrabiliary amylase level is low and the length of common channel is less than 8 mm, the NPV is close to 90 % and this is we suppose the correct way to reject the diagnosis of PBM and consequently type Ic BDC.

Likewise, the intrabiliary amylase concentrations were slightly higher in gall bladder rather that in the bile duct

probably by a concentration phenomenon and might strengthen the carcinogenesis assumptions of stagnation theories. These theories accused pancreatic reflux and biliary tract aggression as being the main factor of cancerous degeneration especially in dilated segments [19], while cancerous degeneration into cholangiocarcinoma in the distal part of the biliary tree such as those observed in Caroli's disease (type V) seems not to be directly related to the presence or absence of PBM [8, 11, 20, 22, 23].

The presence of PBM appears to increase the incidence of biliary cancer especially in Todani's type I BDC, with cancerous degeneration into cholangiocarcinoma proximately 10 years earlier than in patients without PBM [2]. In the present series, no cancerous transformation of BDC was observed in children aging less than 15 years, while the first cancer case was 21 years [24]. This is also true for patients who underwent cystenterostomy which are at higher risk of biliary cancer degeneration [24, 25].

The global incidence of cancer was 8.7 % in this study, and this included adults and children. This cancer incidence is close to most Eastern series [3, 8]. Among the 16 patients with synchronous cancer and analyzable BJB, 12 (75 %) had a Todani's type I BDC and 83.3 % an associated PBM, while, among the 4 patients with synchronous cancer in type IVa, only 1 (25 %) presented a PBM. Although the smallness of cancer patients in the present series, these observations might suggest the possible role of PBM in cancer degeneration of Todani's type I BDC, while the impact of the presence of PBM for cancer degeneration of Todani's type IV remained much less clear.

This is a retrospective series with a long period of inclusion. The global incidence of cancer was probably underestimated because of the retrospective data collection methods.

In conclusion, the characteristics of PBM associated with congenital BDC in Western population are quite close to Eastern series. Regarding the results of the present series, we propose to consider both the intrabiliary value of amylase >8000 UI/L and a length of a common channel >8 mm as appropriate values for the positive diagnosis of PBM. Given the similarity of the findings of this study with Eastern series, the therapeutic implications and guidelines of the Japanese Study Group for Pancreaticobiliary Maljunction are applicable to Western patients.

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Compliance with ethical standards

Conflict of interest Authors declare that they have no conflict of interest.

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