Late Postoperative Complications of Congenital Biliary Dilatation in Pediatric Patients: A Single-Center Experience of Managing Complications for Over 20 Years

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Abstract

Purpose: To investigate late complications after surgery for congenital biliary dilatation (CBD).

Methods: We retrospectively reviewed the patients treated for late postoperative complications of extrahepatic bile duct resection with bilioenteric anastomosis for CBD at our hospital between 1999–2019.

Results: Twenty-seven complications, including bile duct stenosis with (n = 19) or without (n = 3) hepatolithiasis, remnant intrapancreatic bile duct (n = 2), intestinal obstruction (n = 2), and refractory cholangitis (n = 1) were treated in 26 patients. The median age at radical surgery and the initial treatment of complications was 3 years, 2 months and 14 years, 5 months, respectively. The median period from radical surgery to initial treatment of complications was 7 years, 1 month. Before 2013, bile duct stenosis was initially treated with bile duct plasty (n = 11) or hepatectomy (n = 3), and 71.4% (n = 10) of patients needed further treatment; after 2013, double-balloon endoscopic retrograde cholangiography (DBERC) was used (n = 8), and 25% (n = 2) of patients needed further treatment. Patients with remnant intrapancreatic bile duct, intestinal obstruction, and refractory cholangitis required surgery.

Conclusion: Long-term follow-up is necessary after surgery for congenital biliary dilatation. DBERC is thus considered to be useful for bile duct stenosis management.

Keywords: Congenital biliary dilatation, late postoperative complication, bile duct stenosis, double-balloon endoscopic retrograde cholangiography, pediatric

1. Introduction

Congenital biliary dilatation (CBD) is a congenital anomaly of the extrahepatic and/or intrahepatic bile duct. It is commonly accompanied by pancreaticobiliary maljunction, which causes biliary carcinoma, cholangitis, pancreatitis, and other disorders of the biliary tract and pancreas owing to the mutual reflux of bile and pancreatic juice. CBD was historically treated by cystenterostomy (an internal drainage procedure), which was associated with a risk of postoperative bile duct inflammation and carcinogenesis. Currently, extrahepatic bile duct resection with bilioenteric anastomosis is the standard radical surgery for separating the mutual countercurrents of bile and pancreatic juice. Although early complete extrahepatic bile duct resection with bilioenteric anastomosis generally provides a good prognosis, late postoperative complications, including cholangitis, hepatolithiasis, pancreatitis, pancreatolithiasis, and biliary carcinoma, have been recognized. These complications are potentially serious and often appear several years after surgery, sometimes decades later. Generally, maintaining a long-term follow-up into adulthood is challenging in patients who undergo surgery during childhood. Therefore, there are few long-term reports on postoperative complications following radical surgery for CBD [1-4], and the optimal therapeutic strategy remains unclear.

The aim of this study was to investigate late postoperative complications after radical surgery for CBD and their treatment course by reviewing our 20-year experience of managing such complications.

2. Materials and Methods

We retrospectively reviewed the medical records of patients who underwent radical surgery between 1978 and 2018, and who were treated for late postoperative complications after

radical surgery for CBD between 1999 and 2019 at Nagoya University Hospital. Patients who underwent extrahepatic bile duct resection with bilioenteric anastomosis for CBD before 16 years of age at any hospital and were subsequently treated for late postoperative complications at our hospital were included. Therefore, this study not only included patients who underwent radical surgery for CBD at our hospital, but also included those who underwent radical surgery for CBD at other hospitals and were treated for late postoperative complications at our hospital. Late postoperative complications were defined as those requiring readmission 30 days after radical surgery. The occurrence and management of complications were evaluated.

For the treatment of postoperative bile duct stenosis after surgery for CBD, double-balloon endoscopic retrograde cholangiography (DBERC) was first introduced in the Department of Pediatric Surgery in 2013. Therefore, we analyzed the management of bile duct stenosis in two distinct periods, before and after 2013, according to the adoption of DBERC. In order to evaluate the therapeutic effects of DBERC on bile duct stenosis, we compared the hepatectomy rate (percentage of cases undergoing hepatectomy at least once) and retreatment rate for bile duct stenosis between the two groups using the Fisher's exact test. The average period from initial treatment to discharge between the two groups was compared using Student's *t*-test. *P*-values ≤ 0.05 were considered to be statistically significant.

This study was approved by the institutional ethics board of Nagoya University Hospital (approval number: 2019-0417) and performed in accordance with the ethical standards laid down in the 1964 Declaration of Helsinki and its later amendments. Since this was a retrospective observational study and the data analyzed were anonymized, informed consent from participants or their parents/guardians was obtained through an opt-out method on our

hospital website in accordance with the Ethical Guidelines for Medical and Health Research Involving Human Subjects in Japan.

3. Results

3.1 Patient characteristics

This study included 26 patients (male/female: 5/21), whose characteristics are summarized in Table 1. There were one type Ia (3.8%), one type Ic (3.8%), and thirteen type IV-A (50.0%) patients according to Todani's classification [5]; one with double extrahepatic bile duct type II according to Saito's classification (3.8%) [6], and ten others with unknown classifications (38.5%). All patients underwent extrahepatic bile duct resection with hepaticojejunostomy; 13 at our hospital and 13 at other hospitals. Three radical operations were performed laparoscopically, and the remaining 23 were performed through an open approach. The median age at radical surgery and the first treatment for late postoperative complications was 3 years and 2 months (range, 1 month to 14 years and 4 months) and 14 years and 5 months (range, 10 months to 39 years and 1 month), respectively. The median period from radical surgery to the first treatment for late postoperative complications was 7 years and 1 month (range, 1 month to 33 years and 3 months). The median observational period since the final treatment for late postoperative complications was 6 years (range, 0 months to 20 years and 2 months).

3.2 Late postoperative complications

A total of 27 late postoperative complications occurred in 26 patients (Table 2), which included bile duct stenosis with hepatolithiasis (n = 19), bile duct stenosis without hepatolithiasis

(n = 3), remnant intrapancreatic bile duct (n = 2), intestinal obstruction (n = 2), and refractory cholangitis (n = 1). One of the two patients with remnant intrapancreatic bile ducts had pancreatolithiasis. One patient developed both bile duct stenosis and intestinal obstruction. No cases of biliary carcinoma were observed.

The complication rates were calculated only from patients who underwent radical surgery at our hospital. Thus, the denominator was the 226 patients who underwent radical surgery for CBD at our hospital between 1978 and 2018, and the numerator was the number of patients who underwent radical surgery at our hospital and were treated for late complications between 1999 and 2018. The complication rates of bile duct stenosis with and without hepatolithiasis, remnant intrapancreatic bile duct, intestinal obstruction, refractory cholangitis, and biliary carcinoma, for which radical surgeries were performed at our hospital, were 4.9%, 0.9%, 0%, 0.9%, 0%, and 0%, respectively.

3.3 Management of bile duct stenosis

The 22 cases of bile duct stenosis occurred at a median of 8 years (range, 8 months to 32 years and 2 months) after radical surgery.

Before 2013, bile duct stenosis occurred in 14 cases, and was initially treated with hepatectomy (n = 3) or bile duct plasty (n = 11), as shown in Figure 1. Of the three patients who initially underwent hepatectomy, one patient who had peripheral bile duct with hepatolithiasis required a repeat hepatectomy — after lateral segmentectomy, the stump biloma remained and hepatolithiasis appeared, so a left lobectomy was performed to additionally excise S4. The other patients required one, and two repeat bile duct plasties after hepatectomy, respectively. One patient, who had hepatolithiasis in the posterior segment branch, underwent posterior

segmentectomy, but septal stenosis and hepatolithiasis were observed in B4b, requiring bile duct plasty. The other patient, who had been considered for hepatectomy or bile duct plasty but refused surgery, had disseminated intravascular coagulation due to cholangitis, underwent hepatectomy, and subsequently required two bile duct plasties due to bile duct stenosis. Of the 11 patients who underwent bile duct plasty as the initial treatment, four improved while seven required additional treatment, of which three underwent a repeat bile duct plasty and four underwent DBERC. Of the three patients who underwent bile duct plasty as the second treatment, one improved, one required another bile duct plasty, and one required DBERC with balloon dilatation of the stenosis. Of the four patients who underwent DBERC as a second treatment, one improved after DBERC with hepatolithectomy, and another improved after two additional DBERCs with both hepatolithectomy and balloon dilatation of the stenosis. The third patient eventually required hepatectomy after failure of bile duct plasty and three DBERCs with both hepatolithectomy and balloon dilatation of the stenotic site. The final patient was scheduled for hepatectomy because DBERC revealed a right hepatic duct obstruction. Therefore, before 2013, the hepatectomy and retreatment rates after the first treatment for bile duct stenosis were 28.6% (4/14 cases) and 71.4% (10/14 cases), respectively. Before 2013, the average period from initial treatment to discharge was 18.1 (range 10–33) days after bile duct plasty or hepatectomy.

In 2013, our hospital began performing DBERC in patients > 10 years old with suspected bile duct stenosis. DBERC was considered for patients < 10 years old at our hospital, with the indication being based on their needs and risks [7]. Since 2013, DBERC has been performed as the initial treatment for bile duct stenosis (n = 8; Figures 2 and 3) [8, 9]. DBERC distinguished between intrahepatic bile duct stenosis (IHBDS) and hepaticojejunal anastomotic stenosis (HJAS). When biliary duct mucosa was observed between the stenotic and anastomotic

segments, the stenosis was diagnosed as IHBDS. If biliary duct mucosa was not observed, then the stenosis was diagnosed as an HJAS. Two of the four patients with IHBDS were successfully treated with initial DBERC with balloon dilatation of the stenosis. Another had additional DBERC with both hepatolithectomy and balloon dilatation of the stenotic area, but further DBERC was planned in the future for persistent hepatolithiasis. The other patient had left hepatic duct obstruction on the initial DBERC and underwent left lobectomy. Two patients with HJAS and two with both IHBDS and HJAS were successfully treated by initial DBERC with balloon dilatation of the stenosed segment. Therefore, after 2013, the hepatectomy rate was 12.5% (1/8 cases), which was not significantly different from the rate before 2013 ($p = 0.613$; Table 3). After 2013, the retreatment rate after the first treatment for bile duct stenosis was 25.0% (2/8 cases), which was not significantly different from the rate before 2013 ($p = 0.074$; Table 3). The average period from initial treatment to discharge was 5.3 days (range, 1–21) after DBERC after 2013, which showed that patients were discharged significantly earlier after DBERC treatment ($p = 0.003$; Table 3). However, one of the patients who underwent DBERC for HJAS developed procedure-related ascending jejunal limb stenosis one month after the procedure. DBERC with balloon dilatation of the stenotic site required approximately 70 minutes. After two unsuccessful DBERCs with insertion of stenting tubes, laparoscopic-assisted resection of the stenosed segment of the Roux-en-Y limb was performed.

3.4 Management of other late postoperative complications

One of the two patients with remnant intrapancreatic bile duct developed pancreatitis three months after radical surgery. Magnetic resonance cholangiopancreatography (MRCP) revealed a 25-mm remnant intrapancreatic bile duct with pancreatolithiasis, which was resected

11 months postoperatively. The other patient had an uneventful postoperative course and was followed up for seven years before the termination of follow-up. However, the patient complained of occasional abdominal pain that started 25 years after radical surgery, and later underwent MRCP 33 years postoperatively. A 22 × 17 mm remnant intrapancreatic bile duct was discovered and resected.

One of the two patients with intestinal obstruction underwent bandlysis one month after radical surgery. The other patient underwent adhesiolysis or revision of Roux-en-Y anastomosis for repeated adhesive intestinal obstruction 4 months, and 16, 17, and 18 years after radical surgery. The patient also underwent DBERC for bile duct stenosis (both IHBDS and HJAS) 25 years after radical surgery.

One patient with refractory cholangitis underwent revision of Roux-en-Y anastomosis 30 years after radical surgery. However, the patient's condition did not improve and a future DBERC has been planned.

4. Discussion

In this study, we retrospectively reviewed the medical records of patients who were treated for late postoperative complications after radical surgery for CBD at our hospital for over 20 years, half of whom underwent radical surgery at other hospitals but were treated for complications at our hospital. Late postoperative complications included bile duct stenosis with or without hepatolithiasis, remnant intrapancreatic bile duct, intestinal obstruction, and refractory cholangitis. The median period from radical surgery to the initial treatment for late postoperative complications was 7 years and 1 month, with the longest interval being over 30 years. No cases of biliary carcinoma were observed in this study, plausibly because all patients who were

included had undergone radical surgery for CBD before 16 years of age. However, there are reports that biliary carcinoma can develop from the residual common bile duct, intrahepatic bile duct, or remnant intrapancreatic bile duct following radical surgery [4, 10-14]. Repeated cholangitis, hepatolithiasis, or cystic remnants due to incomplete intrahepatic bile duct excision appear to be particularly associated with carcinogenesis. The incidence of biliary carcinoma among patients who undergo radical surgery is reported to be approximately 0.7–5.4%, which is 120–200-times higher than that in the general population [11, 12]. Therefore, to detect and treat postoperative complications earlier, long-term follow-up is crucial after radical surgery for CBD.

The most common complication observed was bile duct stenosis. Bile duct stenosis is known to cause cholangitis, hepatolithiasis, and even biliary carcinoma. However, the optimal therapeutic strategy for bile duct stenosis after radical surgery for CBD has not yet been established. In this study, 10/14 patients (71.4%) who initially underwent hepatectomy or bile duct plasty before 2013 required repeated treatment, demonstrating the challenges of treating bile duct stenosis. All three patients who initially underwent hepatectomy required reoperation. Of the 11 patients who initially underwent bile duct plasty, seven required additional treatments, with two undergoing additional bile duct plasties and five undergoing DBERC after additional bile duct plasties. Three of the five patients who underwent DBERC improved with concurrent hepatolithectomy and/or balloon dilatation. Another patient required hepatectomy because bile duct plasty and DBERC had been ineffective. The other patient was scheduled to undergo hepatectomy because hepatic duct obstruction was revealed on DBERC. These indicated that bile duct stenosis was difficult to treat because accurate information about the stenotic area was not known before treatment. Therefore, in 2013, we introduced DBERC as the first line of treatment for bile duct stenosis after radical surgery for CBD in order to detect the precise stenotic area.

DBERC is recognized as a minimally invasive method for the treatment of bile duct complications after Roux-en Y biliary reconstruction, but there are few reports about the outcome of DBERC for bile duct stenosis after radical surgery for CBD [15]. Only 2/8 patients (25%) who initially underwent DBERC since 2013 underwent repeated treatments, though this was not significantly different from the retreatment rate before 2013. One patient required additional DBERC for hepatolithiasis removal, but further DBERC was planned to remove residual disease. The other patient underwent hepatectomy because DBERC revealed hepatic duct obstruction. Although only ultrasonography and MRCP were performed for the diagnosis of bile duct stenosis before the introduction of DBERC, DBERC enabled simultaneous diagnosis and appropriate treatment. Moreover, any findings observed during DBERC could also be addressed by appropriate treatments, such as hepatolithectomy and/or balloon dilatation. Although we found no significant difference in the hepatectomy rates before and after 2013, DBERC can reveal hepatic duct obstruction requiring hepatectomy; accordingly, hepatectomy should be considered only when hepatic duct obstruction is observed during DBERC or when DBERC with hepatolithectomy and/or balloon dilatation is ineffective. In cases of bile stasis in which stenosis of the intrahepatic bile duct is unclear, repeated DBERC procedures can be performed due to their minimal invasiveness. Bile duct stenosis is difficult to treat and not all can be treated with DBERC; however, DBERC can be considered a good initial investigation and/or treatment modality for bile duct stenosis following radical surgery for CBD and can help avoid more invasive surgical procedures.

Gastrointestinal perforation, bleeding, mucosal injury, bile duct injury, cholangitis, and pancreatitis are known complications of DBERC. Yokoyama et al. reported that complications occurred in 5.4% of pediatric patients undergoing double-balloon enteroscopy and DBERC, in

association with concomitant therapeutic procedures, such as endoscopic polypectomy, endoscopic bile duct dilatation, or removal of hepatolithiasis [7]. In this study, DBERC was performed 20 times with only one procedure-related complication (5%): an ascending jejunal limb stenosis one month after DBERC. This may be related to the longer than usual time required to pass the probe through the anastomotic site during DBERC.

Other than bile duct stenosis, the postoperative complications observed in this study were remnant intrapancreatic bile duct and refractory cholangitis. A remnant intrapancreatic bile duct can cause postoperative pancreatitis, pancreatolithiasis, and carcinoma [14]. In this study, two patients with remnant intrapancreatic bile ducts were diagnosed with pancreatitis and recurrent abdominal pain at 3 months and 25 years postoperatively, respectively. This suggests that even if a patient remains asymptomatic for several decades after radical surgery, pancreatitis or pancreatolithiasis associated with remnant intrapancreatic bile duct can occur, and inappropriate treatment may increase the risk of carcinogenesis. Repeated Roux-en-Y anastomosis was not effective in treating refractory cholangitis. Postoperative cholangitis often results from cholestasis due to bile duct stenosis; therefore, DBERC was planned in the future for these patients.

To prevent the aforementioned complications, complete resection of the extrahepatic bile duct, resection of the membrane or septum causing intrahepatic bile duct stenosis, resection of the pancreatic bile duct just above the pancreatobiliary junction using intraoperative cholangiography or cyst endoscopy, and wide hilar hepaticojejunostomy are recommended during the initial surgery [16]. Since these complications can occur long after surgery, even in asymptomatic patients, a long-term follow-up is crucial. Patients should also be educated about the possibility of late complications and the importance of mentioning their history of surgical

CBD treatments when admitted to the hospital for abdominal pain. Blood tests and imaging studies, including MRCP, should be considered in these cases.

There are some limitations associated with this study. This study included patients who were treated for late postoperative complications at our hospital, half of whom underwent radical surgery at other hospitals. Therefore, the overall prognosis after radical surgery for CBD, including the precise incidence of postoperative complications, is unknown. For reference, we calculated the complication rates only from the cases who underwent radical surgery at our hospital. In addition, less than 10 years have passed since DBERC was introduced as the initial treatment for bile duct stenosis after radical surgery for CBD at our hospital and the number of cases is limited. Therefore, long-term follow-up studies are needed in the future.

In conclusion, late postoperative complications of extrahepatic bile duct resection with hepaticojejunostomy for CBD requiring hospitalization can occur after several years or even decades, emphasizing the importance of long-term follow-up. DBERC is useful for both the diagnosis and treatment of bile duct stenosis after radical surgery for CBD.

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Figure legends

Fig. 1 Management of bile duct stenosis after extrahepatic bile duct resection with hepaticojejunostomy for congenital biliary dilatation before 2013

* Three DBERC procedures with hepatolithectomy and balloon dilatation were not effective.

** DBERC revealed a hepatic duct obstruction.

*** Median (interquartile range)

BD plasty, bile duct plasty; DBERC, double-balloon endoscopic retrograde cholangiography; BD, balloon dilatation; HL, hepatolithectomy; Obs, observation; y, years; m, months

Fig. 2 Management of bile duct stenosis after extrahepatic bile duct resection with hepaticojejunostomy for congenital biliary dilatation after 2013

* Residual hepatolithiasis remained

** DBERC revealed left bile duct obstruction.

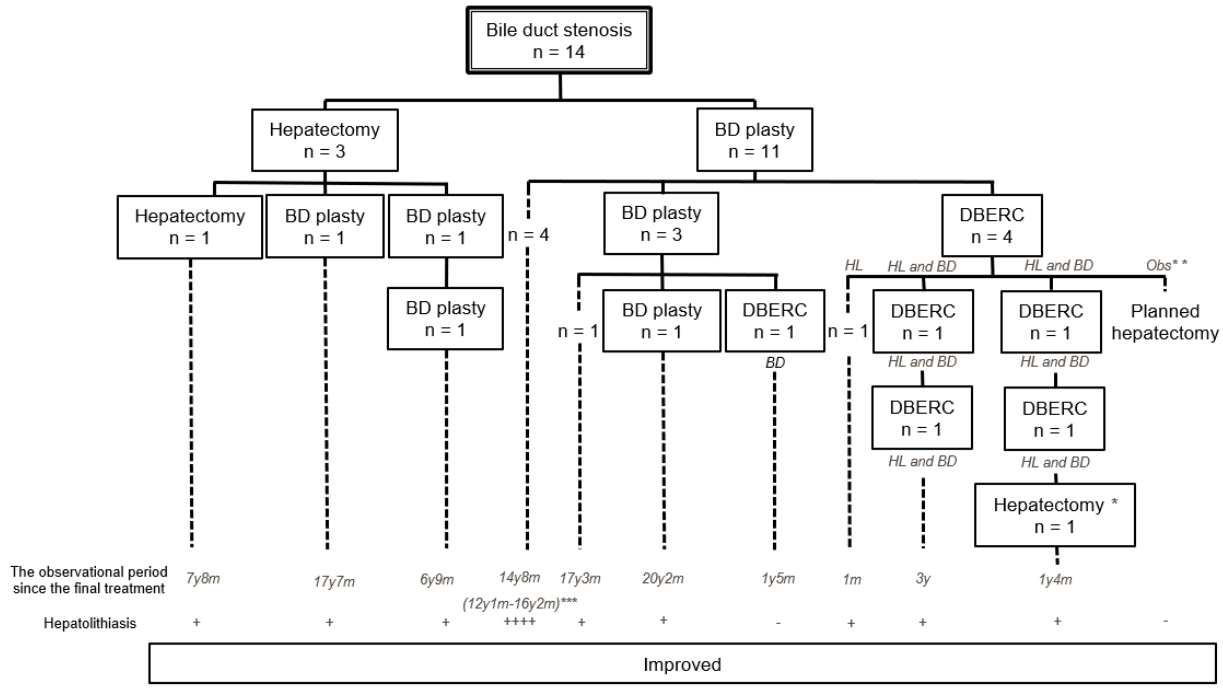
*** DBERC-related complication

IHBDS, intrahepatic bile duct stenosis; HJAS, hepaticojejunal anastomotic stenosis; DBERC, double - balloon endoscopic retrograde cholangiography; BD, balloon dilatation; HL, hepatolithectomy; Obs, observation, y, years; m, months

Fig. 3 Endoscopic balloon dilatation for bile duct stenosis using double-balloon endoscopic retrograde cholangiography

a: cholangiogram of bile duct stenosis (a white arrow); b: endoscopic image of bile duct stenosis; c: endoscopic image of fully dilatated bile duct stenosis after balloon dilatation

Fig. 1


Bile duct stenosis
n = 14
Hepatectomy
n = 3
BD plasty
n = 11
Hepatectomy
n = 1
BD plasty
n = 1
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hepatectomy
HL and BD
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Hepatectomy *
n = 1
The observational period
since the final treatment
7y8m
17y7m
6y9m
14y8m
(12y1m-16y2m)***
17y3m
20y2m
1y5m
1m
3y
1y4m
Hepatolithiasis
+
+
+
++++
+
+
-
+
+
+
-
Improved

Fig. 2

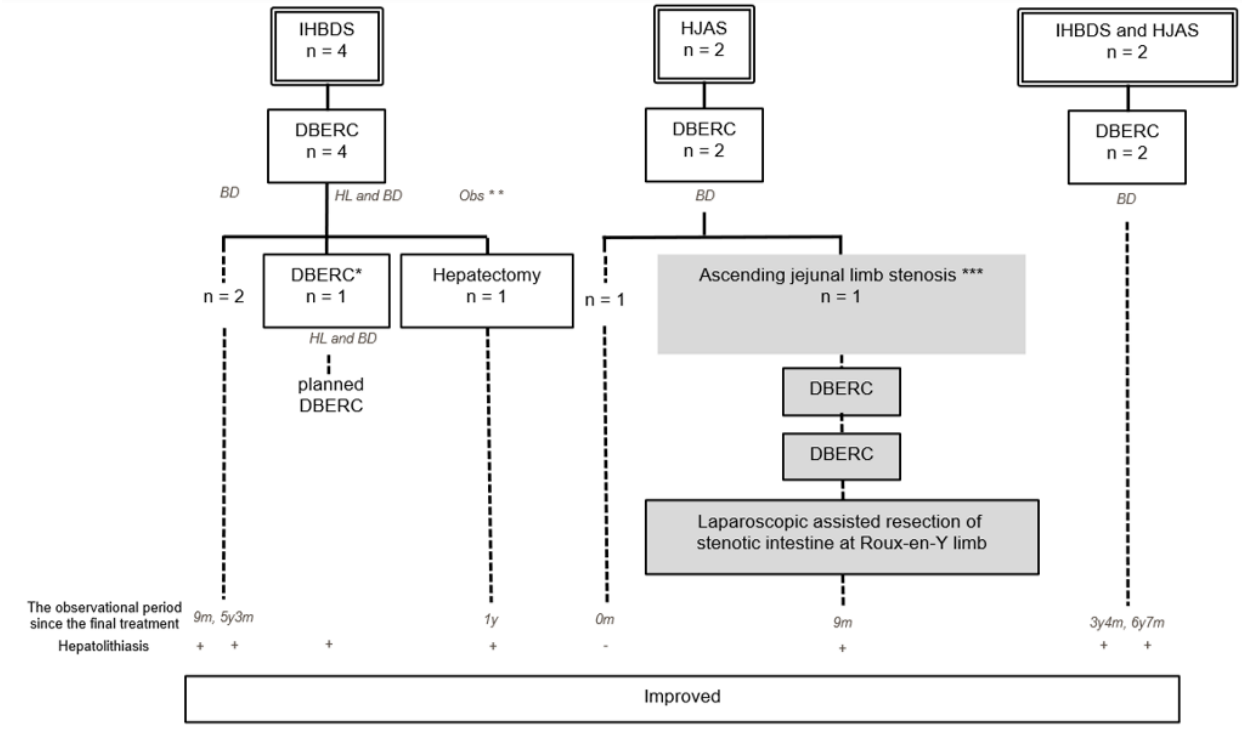
IHBDS
n = 4
DBERC
n = 4
BD
HL and BD
Obs * *
n = 2
DBERC*
n = 1
HL and BD
planned
DBERC
Hepatectomy
n = 1
HJAS
n = 2
DBERC
n = 2
BD
n = 1
Ascending jejunal limb stenosis ***
n = 1
DBERC
DBERC
Laparoscopic assisted resection of
stenotic intestine at Roux-en-Y limb
IHBDS and HJAS
n = 2
DBERC
n = 2
BD
The observational period
since the final treatment
9m, 5y3m
1y
0m
9m
3y4m, 6y7m
Hepatolithiasis
+ +
+
+
-
+
+ +
Improved

Fig. 3

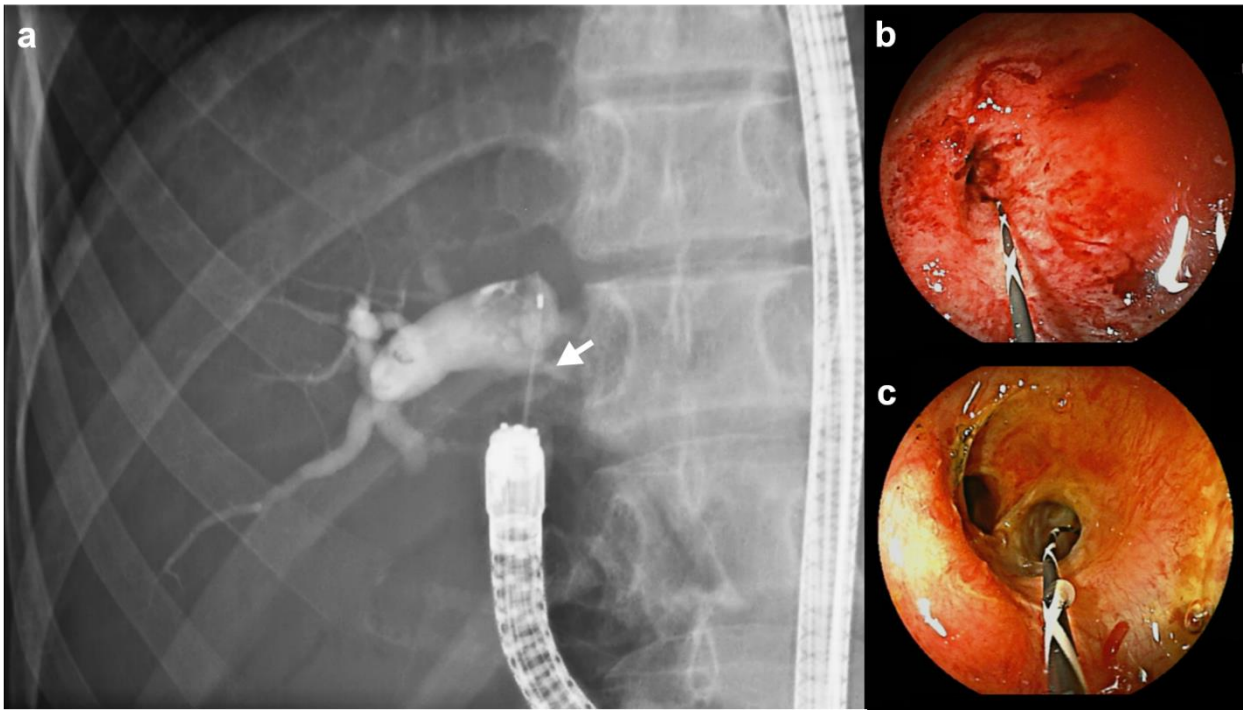

Table 1 Demographic characteristics of the 26 patients

Sex	
Male	5 (19.2%)
Female	21 (80.8%)
Anatomical classification	
Todani type Ia	1 (3.8%)
Todani type Ic	1 (3.8%)
Todani type IVa	13 (50.0%)
Double extrahepatic bile duct, Saito Type II	1 (3.8%)
Unknown	10 (38.5%)
Surgical approach	
Open	23 (88.5%)
Laparoscopic	3 (11.5%)
Median age at radical surgery (range)	3 years and 2 months (1 month to 14 years and 4 months)
Median age at the first treatment for late postoperative complications (range)	14 years and 5 months (10 months to 39 years and 1 month)
Median period from radical surgery to the first treatment for late postoperative complications (range)	7 years and 1 month (1 month to 33 years and 3 months)
Median observational period since the final treatment for late postoperative complications	6 years (0 months to 20 years and 2 months)

Table 2 Late postoperative complications associated with extrahepatic bile duct resection with hepaticojejunostomy for congenital biliary dilatation

Complication	N***	Complication rate**** (%)
Bile duct stenosis with hepatolithiasis	19 (11, 8)	4.9
Bile duct stenosis without hepatolithiasis	3 (2, 1)	0.9
Remnant intrapancreatic bile duct*	2 (0, 2)	0
Intestinal obstruction**	2 (2, 0)	0.9
Refractory cholangitis	1 (0, 1)	0
Biliary carcinoma	0 (0, 0)	0

* Of the two patients with remnant intrapancreatic bile ducts, one had pancreatolithiasis.

**One patient had both bile duct stenosis and intestinal obstruction.

***Total number of complications (the number of complications that arose after radical surgery was performed at Nagoya University Hospital, other hospitals)

**** The complication rates were calculated only from the patients who underwent radical surgery at Nagoya University Hospital; the denominator was the 226 patients who underwent radical surgery for CBD at Nagoya University Hospital between 1978–2018, and the numerator was the number of patients who underwent radical surgery at Nagoya University Hospital and were treated for late complications.

Table 3 Comparison of the therapeutic effects of double-balloon endoscopic retrograde cholangiography (DBERC) on bile duct stenosis between two periods: before and after 2013, when DBERC was adopted

	Bile duct stenosis before the adoption of DBERC (n = 14)	Bile duct stenosis after the adoption of DBERC (n = 8)	*P*-value
The hepatectomy rate (n, %)	4 (28.6)	1 (12.5)	0.613*
The retreatment rate for bile duct stenosis (n, %)	10 (71.4)	2 (25.0)	0.074*
The average period from first treatment to discharge (days)	18.1 (range, 10–33)	5.3 (range, 1–21)	0.003**

*Fisher's exact test
**Student's *t*-test