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Percutaneous Transhepatic Biliary Intervention in Adult Biliary Atresia Patients After Kasai Portoenterostomy

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Abstract: Kasai portoenterostomy (KP) is a standard treatment for patients with biliary atresia (BA). After KP, patients with BA occasionally develop biliary complications, such as recurrent cholangitis, biliary stricture, and cystic dilatation of the intrahepatic bile duct. Percutaneous transhepatic biliary drainage is one of the treatment options for these biliary complications. However, limited information is available on percutaneous transhepatic biliary drainage performed after KP in adult BA patients with native livers. Herein, we describe 8 cases of percutaneous transhepatic biliary interventions performed after KP in 7 adult BA patients with native livers. Cholangiography showed multiple cystic dilatation of the intrahepatic bile ducts. Advancing a guidewire and catheter was difficult due to the multiple dilatations and strictures of the bile duct. Successful biliary drainage tube placement and clinical improvement was achieved in 5 and 3 cases, respectively. Because of its technical difficulty and limited clinical effectiveness, it is not recommended that it be performed easily.

Key Words: cholangiography, percutaneous transhepatic biliary drainage

What Is Known

- Biliary complications occasionally develop in patients with biliary atresia (BA) after a Kasai portoenterostomy (KP), which is performed as a treatment for BA.
- Percutaneous transhepatic biliary drainage is regarded as a treatment option for biliary problems in these patients.

What Is New

- Multiple cystic dilatation is a common cholangiography finding in adult BA patients with native livers who undergo percutaneous transhepatic biliary intervention.
- Because of this characteristic finding, percutaneous transhepatic biliary intervention is technically difficult to perform after KP in adult BA patients with native livers.

INTRODUCTION

Biliary atresia (BA) is a neonatal hepatobiliary disease of unknown etiology in which destructive and obstructive inflammation affects both the intrahepatic and extrahepatic bile ducts (1). Kasai portoenterostomy (KP) is widely accepted as the first-line treatment for BA (2). Majority of patients who live with their native livers after KP experience biliary complications, including recurrent cholangitis, and biliary stricture (3,4). Additionally, cystic dilatation of the intrahepatic bile duct has been known to develop in the native livers after KP (5,6). Percutaneous transhepatic biliary drainage (PTBD) is a treatment option for these complications (2). To date, a small number of case series have described PTBD procedures performed after KP in pediatric patients with BA, with variable clinical effectiveness (7–10). One study recommended liver transplantation in pediatric BA patients with multiple cystic dilatation because of the limited PTBD effectiveness (9). On the contrary, another study reported its usefulness.10 Moreover, studies describing PTBD procedures in adult BA patients are even fewer (11). The objective of this study was to describe the cholangiography findings, and technical detail, clinical results, and complications of percutaneous transhepatic biliary interventions for adult BA patients living with their native livers after KP.

MATERIALS AND METHODS

Patients

This retrospective study was approved by the institutional review board. The institutional review board waived the requirement for written informed consent from patients due to the retrospective design of the study. Between January 2003 and December 2020, KP was performed in 32 BA patients at our institution. Moreover, many patients with BA who had undergone KP at other hospitals were referred to our institution for possible liver transplantations. Accordingly, during this period, 273 liver transplantations were performed for BA patients at our institution. During the same period, 146 percutaneous transhepatic biliary interventions were performed in 85 patients with BA after KP at our institution. Among them, 105 interventions in 57 patients were excluded because the interventions were performed in pediatric patients (younger than 18 years). Further, 33 interventions in 21 patients were excluded because the interventions were performed in patients after liver transplantation. Thus, the study comprised the remaining 8 interventions performed in 7 adult BA patients (18 years old or older) living with their native livers after KP. Among the 7 patients, 4 were women and 3 were men, including 1 man who underwent biliary intervention twice. The median age at the time of biliary intervention in the 8 cases was 24 years (range, 18–35 years). The median age at which KP was performed after birth in the

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7 patients was 71 days (range, 66–92 days). The gross appearance of the liver and biliary tree at KP based on Ohi classification was type I in 3 cases and type III in 4 cases (12). The median of Child-Pugh score was 7 (range, 5–10). The indications of biliary intervention were 1 or 2 of the following: jaundice, elevated liver enzyme levels, and active or recurrent cholangitis. Information on each patient is presented in Table 1.

Percutaneous Transhepatic Biliary Intervention Procedure

Percutaneous transhepatic biliary intervention was performed by 5 board-certified interventional radiologists with 6 to 23 years of experience in percutaneous transhepatic biliary intervention. Cephem antibiotics were administered prophylactically during the peri-interventional period. Under local anesthesia or general anesthesia, the distal or proximal portion of the segmental intrahepatic bile duct was punctured with a 21-G PTBD needle under ultrasound guidance. If bile was aspirated, the contrast medium was slowly injected to opacify the bile ducts. Then, a 0.018-inch guidewire was carefully advanced into the bile duct under fluoroscopic guidance. Using a 2-step kit (Skater Introducer Set; Argon Medical Devices, Texas, USA), a 4-F introducer sheath was placed into the bile duct. A 0.035-inch hydrophilic guidewire (Radifocus; Terumo, Tokyo, Japan) and a 4-F seeking catheter were used to cross the stenosis of the anastomotic site or to place an external drainage tube. If cholangiography showed stenosis of the anastomotic site, dilatation of the anastomotic site was performed with a balloon catheter. If biliary stones were observed, the stones were driven into the jejunum by flushing with saline from a seeking catheter or by pushing them with a compliant balloon. Finally, a 7- or 8.5-F external or internal-external drainage catheter was placed into the bile duct. A few days later, if cholangiography showed good passage of contrast medium to the bowel, the drainage tube was removed. Tract embolization was not performed during tube removal because intra-abdominal adhesions associated with KP were considered to prevent bile leakage into the abdominal cavity.

Findings of Cholangiography

The morphology of the intrahepatic bile ducts was evaluated in patients who underwent successful percutaneous transhepatic cholangiography (PTC). Additionally, the presence of stenosis at the anastomotic site and biliary stones on PTC were evaluated.

Technical Detail and Clinical Result

The technical detail of percutaneous transhepatic biliary intervention was reviewed based on an interventional radiology record. Clinical result was reviewed based on an electronical medical record, referring to the symptoms and laboratory findings. The safety of percutaneous transhepatic biliary intervention was recorded using Society of Interventional Radiology criteria (13). Whether the patients underwent liver transplantation and they were put on the liver transplantation waiting list after the intervention was reviewed using the electronic medical record.

RESULTS

Findings of Cholangiography

PTC was successfully performed in 7 cases, whereas in one case, puncture of the intrahepatic bile duct was unsuccessful because the intrahepatic bile ducts were not dilated. Multiple dilatations and strictures of the bile duct branches were observed in all 7 cases. This morphology was named multiple cystic dilatation according to the classification proposed in 1994 (5). Stenosis at the anastomotic site was observed in 3 cases, and biliary stones were observed in 2 cases. Information on cholangiography findings, technical detail, clinical results, and complications are shown in Table 1.

Technical Detail and Clinical Result

After successful PTC in 7 cases, a 4-F sheath was placed in 5 cases. In 2 cases, a 0.018-inch guidewire could not be advanced into the bile duct deep enough to place a sheath. Among the 5 cases with successful sheath placement, an internal-external drainage catheter was placed in 4 cases (Fig. 1). In 2 cases with anastomotic stenosis, balloon dilatation at the anastomotic site stenosis was performed. In one case with biliary stones, after size-up of the PTBD route to 16-F, cholangioscopy and stone removal were performed by an experienced endoscopist. In one case with biliary stones, stone removal was performed using a balloon catheter. In one case with anastomotic stenosis, an external drainage catheter was successfully placed after sheath placement, but a 0.035-inch hydrophilic guidewire could not be advanced to the anastomotic site, and balloon dilatation of the anastomotic stenosis was abandoned (Fig. 2).

After biliary stone removal and dilatation of anastomotic stenosis, clinical improvement was achieved in 3 cases. In the remaining 5 cases, clinical improvement was not achieved.

One patient underwent liver transplantation 18 years after the intervention. Two patients were placed on the liver transplantation waiting list, and one of them died due to bile duct cancer 8 months after the intervention. The remaining 4 patients live with their native liver and not on the liver transplantation waiting list.

Complications

Three complications occurred, of which 1 was major and 2 were minor. In one case, cholangitis developed after an unsuccessful attempt at biliary drainage tube placement. In this case, cholangitis continued regardless of antibiotic treatment. The second attempt of biliary drainage was successful, and the cholangitis improved. This was regarded as a major complication. In one case, the needle and guidewire were removed after unsuccessful advancement of the guidewire. Subsequently, abdominal pain developed. In another case, a biliary drainage catheter was removed after a successful biliary intervention. Fever and abdominal pain developed after the removal. In both cases, biliary peritonitis due to bile leakage was diagnosed, and the symptoms improved with antibiotic administration. These 2 cases were regarded as minor complications.

DISCUSSION

In this study, most cases showed multiple cystic dilatation of the intrahepatic bile ducts on PTC. Previous studies reported much lower incidence of this finding, ranging from 6% to 28% (5,6,9,14). It is possible that cystic dilatation of the intrahepatic bile ducts in the native livers after KP is more common in adults than in children. Intensive studying is necessary to know the frequency of multiple cystic dilatation in adult BA patients after KP.

This study showed technical difficulty and low clinical efficacy of percutaneous transhepatic biliary interventions in adult BA patients with their native livers after KP. One study reported 3 cases of successful percutaneous transhepatic biliary interventions in adult BA patients with native livers after KP (11). This previous study also reported on the technical difficulty of this technique in adult BA patients, without explicitly specifying the reason for the difficulty (11). In this study, most cases showed multiple cystic dilatation of the intrahepatic bile ducts on PTC. Thus, the technical difficulty for performing percutaneous biliary intervention in these patients was attributed to the difficulty in advancing a guidewire across multiple dilatations and strictures of the intrahepatic bile ducts. To the best of our knowledge, no study has reported this technical difficulty. Clinical improvement was not achieved after technically successful biliary

TABLE 1. Patient characteristics and detailed information on percutaneous transhepatic biliary intervention

Case no.	Patient no.	Age/sex	Ohi classification	Child-Pugh score	Indication of biliary intervention	Cholangiography findings	Technical detail	Clinical result	Complications	LT after intervention
1	1	18F	III-c1-ν	8	Jaundice, liver enzyme elevation	Multiple cystic dilatation	Unsuccessful external drainage tube placement	Jaundice and liver enzyme elevation persisted		Not on the LT waiting list 2 years later.
2	2	18F	III-b1-μ	5	Recurrent cholangitis	Multiple cystic dilatation, anastomotic stenosis	Successful external drainage tube placement. Unsuccessful balloon dilatation of the anastomotic site	Frequency of cholangitis did not decrease	Biliary peritonitis	LT performed 9 years later.
3	3	21F	I	7	Jaundice, liver enzyme elevation	Multiple cystic dilatation, anastomotic stenosis	Successful internal-external drainage tube placement and balloon dilatation of the anastomotic site	Jaundice and liver enzyme elevation persisted		On the LT waiting list 18 years later.
4	4	24F	III-b1-μ	7	Active cholangitis	N.A.	Unsuccessful bile duct puncture	Cholangitis persisted		Not on the LT waiting list 1 year later.
5	5	24M	I-a	5	Recurrent cholangitis	Multiple cystic dilatation, biliary stones	Successful internal-external drainage tube placement and biliary stone removal using a balloon catheter	Cholangitis did not occur for more than 5 years	Biliary peritonitis	
6	5	33M	I-a	5	Recurrent cholangitis	Multiple cystic dilatation, biliary stones	Successful internal-external drainage tube placement and biliary stone removal using a cholangioscopy	Frequency of cholangitis decreased	Cholangitis	Not on the LT waiting list 4 years later.
7	6	32M	N.A.	10	Jaundice	Multiple cystic dilatation	Unsuccessful external drainage tube placement	Jaundice persisted		Not on the LT waiting list 4 years later.
8	7	35M	III-a1-ν	8	Jaundice	Multiple cystic dilatation, Anastomotic stenosis	Successful internal-external drainage tube placement and balloon dilatation of the anastomotic site	Jaundice improved		On the LT waiting list 4 months later and died due to bile duct cancer 8 months after the intervention.

LT, liver transplantation; N.A., not available.

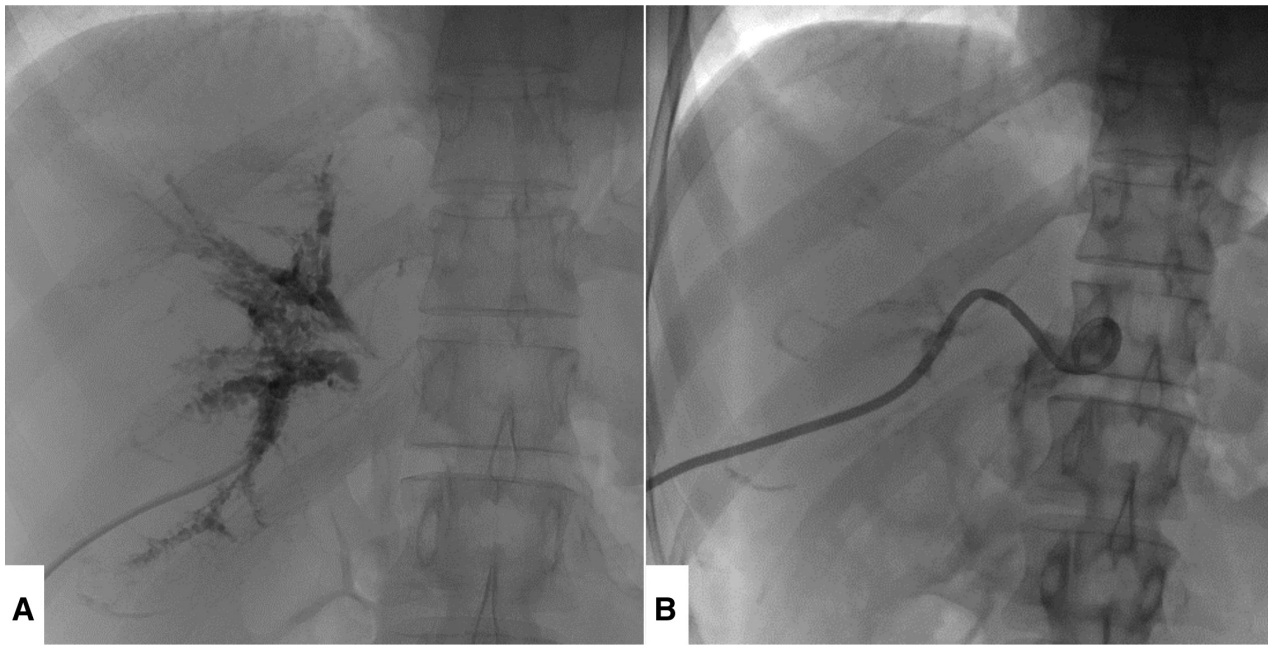

FIGURE 1. Percutaneous transhepatic biliary intervention in a 35-year-old man with biliary atresia after Kasai portoenterostomy (case 8). (A) Cholangiography from an external biliary drainage catheter shows multiple cystic dilatations of the intrahepatic bile ducts. Anastomotic stenosis was diagnosed due to poor opacification of the jejunum. (B) After balloon dilatation of the anastomotic site, an internal-external drainage catheter was placed.

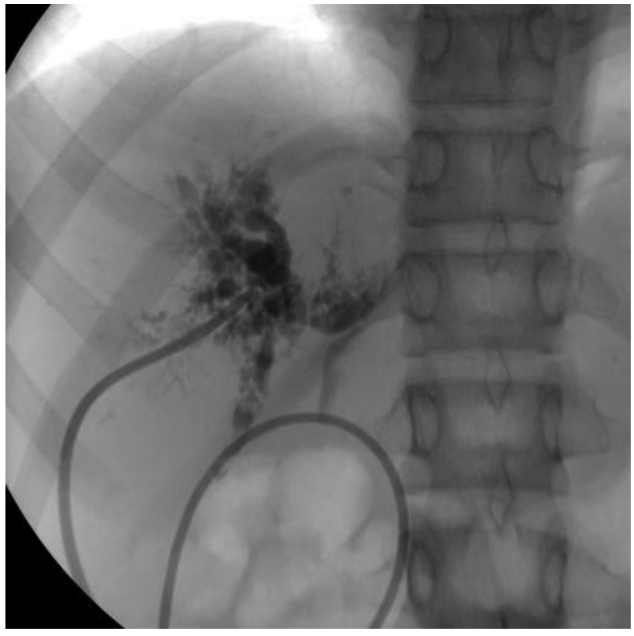

FIGURE 2. Percutaneous transhepatic biliary intervention in an 18-year-old woman with biliary atresia after Kasai portoenterostomy (case 2). Cholangiography from an external biliary drainage catheter shows multiple cystic dilatations of the intrahepatic bile ducts. Anastomotic stenosis was diagnosed due to poor opacification of the jejunum. Advancement of a guidewire and seeking catheter was attempted but unsuccessful because of multiple dilatations and strictures of the bile ducts.

intervention in one case. Multiple cystic dilatation of the intrahepatic bile ducts is thought to cause bile stasis, leading to jaundice and recurrent cholangitis, and the clinical effect of biliary drainage may be limited (9). In all 3 cases with clinical improvement, cholangiography revealed biliary stones or anastomotic stenosis. It can be speculated that clinical improvement of biliary intervention can be expected in patients with biliary stones or anastomotic stenosis, although more studies will be needed to clarify this speculation.

All 7 patients in this study did not show severe biliary problems during childhood, they however manifested in adulthood. These patients were rare previously, and the number of these patients has increased in recent years. Determining the management of these patients is our main issue. We do not consider PTBD as an ideal treatment for biliary problems in adult BA patients after KP because it is technically challenging and the procedure does not have a high-clinical success rate. Nonetheless, the effectiveness of PTBD has been demonstrated in some patients, as mentioned before (11), which is in line with the 3 cases included in this study. The important thing is to recognize that PTBD success rate is not high, and hence, to not perform PTBD easily on these patients.

Three complications, 1 major and 2 minor, were observed in this study. A guideline on PTBD suggested a lower threshold of major complications: 4% (15). In previous studies, a high frequency of complications has not been reported in patients undergoing PTBD with BA after KP. More studies are needed to elucidate the real risk of PTBD in these patients.

This study had some limitations. First, this study was conducted at a single center and the study population was very small. Second, data were collected retrospectively. Third, the procedures were performed by 5 interventional radiologists, and the results were influenced by their individual skills and experiences.

In conclusion, multiple cystic dilatation of the intrahepatic bile ducts was common in adult BA patients with native livers after KP. In these patients, percutaneous biliary intervention may have

high technical difficulty and limited clinical effectiveness. It is recommended not to perform PTBD easily on these patients.

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