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ORIGINAL ARTICLE

Reappraise the effect of redo-Kasai for recurrent jaundice following Kasai operation for biliary atresia in the era of liver transplantation

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Abstract
Purpose This study was conducted to reappraise the efficacy of redo-Kasai (or revision) in the era of liver transplantation as a treatment option in those patients with recurrent jaundice after initially successful Kasai procedure.
Methods We studied ten patients that received redo-Kasai, among a total of 102 patients diagnosed with biliary atresia after receiving Kasai operation from 1986 to 2011.
Results Kasai operation was done at a median age of 55 days and redo-Kasai at 150 days. The bilirubin levels returned to normal in six patients after the procedure. Four of six enjoyed jaundice-free survival with native liver till the time of last follow-up. Three patients died and three received liver transplantation (LT). Only one out of seven patients with three or more episodes of cholangitis survived with native liver, while all the three patients with 1 or 0 episode survived with native liver. The difference was significant ($P = 0.033$). Re-do Kasai did not result in more blood loss or operative time during LT.
Conclusion Redo-Kasai is still valuable in the era of LT and the episodes of cholangitis are the decisive factors affecting the outcome of the procedure.

Keywords Biliary atresia · Kasai operation · Portoenterostomy · Revision

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Introduction

Biliary atresia (BA) is a relatively rare obstructive condition of the bile ducts causing neonatal jaundice, which eventually leads to liver and hepatic failure [1–4]. In 1959, Kasai and Suzuki [5] first reported the surgical technique of portoenterostomy for the treatment of BA. Over the past five decades, it has gradually been clear that early diagnosis, absence of associated congenital malformations, and freedom from postoperative ascending cholangitis are factors predictive of successful bile drainage and survival [6, 7].

Postoperative cholangitis is one of the most common and troublesome complications following Kasai operation for BA, affecting more than 50 % of patients in several reports. The consequent cessation of bile flow frequently leads to failure of the operation unless it can be reversed by appropriate medical or surgical treatment [4, 8]. At this point, redo-Kasai or revision of portoenterostomy may offer hope for clearance of biliary obstruction and drainage of once interrupted bile flow [9–12], but the side effects of adhesion, increasing blood loss, and operative time may make future liver transplantation difficult to perform [13].

The redo-Kasai consists in a procedure to remove fibrotic tissue that prevents the free flow of bile through approach in the front or anterior side of the portoenterostomy, without touching the back of it. Previous reports of

the results of revision of the portoenterostomy varied greatly. Saito et al. [10] reported that only 10 % of their patients survived with adequate biliary drainage after revision of portoenterostomy. In contrast, all five patients in Altman and Anderson's report remained jaundice-free 7 months to 4 years after reoperation [9]. Bondoc et al. [14] reported 40 % success rate in patients subjected to revision and even greater long-term survival in patients undergoing liver transplantation, which is comparable to those receiving liver transplantation primarily after failure of initial Kasai procedure.

Since the impact of the revision and subsequent complications in liver transplantation varies greatly in many centers, we conduct the study to reappraise the effect of redo-Kasai for recurrent jaundice/cholangitis following a successful Kasai operation for BA in a center where liver transplantation is done by a team independent of pediatric surgeons performing redo-Kasai or revision of portoenterostomy.

Materials and methods

Between 1986 and 2011, a total of 102 patients with diagnosis of BA received Kasai operation in this center. During this period, a total of ten patients underwent redo-Kasai operation due to cholangitis and cessation of bile flow after initially appropriate bile drainage following Kasai procedure. The diagnosis of cholangitis was established when the patient had an elevated serum bilirubin level after an episode of fever greater than 38 °C, without an obvious extrahepatic source of infection.

We reviewed the medical records of these patients to evaluate factors that determined success or failure of bile drainage after revision, which includes the age at the moment of Kasai and redo-Kasai procedure, the value of total and direct bilirubin in both preoperative and the postoperative time, number of cholangitis episodes and the outcome of the patients up to the time of last follow-up. Success of revision or re-do Kasai procedure is defined as a total bilirubin level less than 1.5 mg/dl within 3 months after operation. The study has been approved by the IRB of Chang Gung Medical Foundation (# 100-3932B).

Results

All the ten patients included in this study have had an initial successful Kasai procedure. Five of them are males and five females. Only one patient had associated congenital heart anomaly. Nine of the patients were classified as type III and one type I, according to the international classification proposed by Kasai et al. and the Japanese Association of Pediatric Surgeons.

The age of the patients undergoing Kasai procedure ranged from 22 to 87 days, with a median of 55 days. The age at revision ranged from 76 to 300 days, with a median of 150 days. The median total and direct bilirubin level prior to revision were 7.8 and 5.5 mg/dl, respectively. After revision, the bilirubin level returned to normal in six patients, failed to decrease in four.

Of the six patients classified as successful revision, two patients presented new jaundice/cholangitis episodes, one died and one received liver transplantation. Jaundice-free survival with native liver was achieved in four patients. Three were followed up to 9 months, 12 years and 18 years following redo-Kasai. One was lost to follow-up at the age of 12 years. Of the four patients classified as failed redo-Kasai, two received liver transplantation at the age of 2 and 12 years and the other two died 1 and 6 years after revision without further liver transplantation.

The episodes of cholangitis ranged from 1 to 20. Among the five patients with greater than five episodes of cholangitis, two died and three received liver transplantation, while in five with less than five episodes, four survived with native liver and only one died. The difference was significant ($P = 0.048$ by Fisher's exact test). Among the seven patients with three or more episodes of cholangitis following redo-Kasai, only one survived with native liver, while all the three patients with 1 or 0 episode survived with native liver. The difference was also significant ($P = 0.033$). In this limited number of ten patients, age at the time of initial Kasai and of redo-Kasai was not a significant factor determining the success or failure of revision (Table 1).

In the three patients receiving liver transplantation following redo-Kasai, blood loss was 280, 100 and 130 ml, with a mean of 170 ml. The operative time was 708, 430 and 745 min, with a mean of 628 min. There was no mention of increased adhesion in the operative note in these three patients. The results were comparable to those data in a report by Chen et al. [15], in which the mean total blood loss was 176 ml and the mean operative time was 628 min among the 100 patients with BA receiving LT. Most of the patients did not receive revision after failed initial Kasai portoenterostomy prior to LT.

Discussion

Restoration of bile flow in patients with biliary atresia after an initial Kasai procedure does not eliminate the risk of cholangitis which can progress to fibrosis that obstructs the normal bile flow. It also holds true in this study that after revision or redo-Kasai portoenterostomy, the decisive factor

Table 1 Demographic and clinical characteristics

No.	Age Kasai (days)	Age redo-Kasai (days)	Total/direct bilirubin levels prior to redo-Kasai (mg/dl)	Total/direct bilirubin levels after redo-Kasai (mg/dl)	Cholangitis episodes[a]	Outcome	Age at last follow-up
1	63	138	10.8/8.7	1.4/0.7	4 (3)	Survive jaundice free	–[b]
2	30	76	6.8/4.2	0.3/0.1	1 (0)	Survive jaundice free	12 years
3	58	271	8.8/5.4	1.1/0.5	4 (1)	Survive jaundice free	18 years
4	22	185	5.1/4.3	0.9/0.4	1 (0)	Survive jaundice free	9 months
5	56	123	9.4/5.3	1.2/0.5	4 (3)	Died[c]	3 years and 4 months
6	67	108	10.5/8.1	34.9/12.2	13 (12)	Died[d]	1 years and 2 months
7	87	94	6.5/5.4	74/23.7	20 (19)	Died	6 years and 4 months
8	68	101	5.5/3.9	13.4/9.6	10 (9)	Survive with liver transplant	12 years
9	48	300	7.5/5.5	10.2/7.4	6 (4)	Survive with liver transplant	19 years
10	48	104	7.3/4.5	1.5/0.9	6 (3)	Survive with liver transplant	7 years

[a] The number in parenthesis represents the episodes of cholangitis after redo-Kasai

[b] Lost to follow-up at 12-years-old

[c] Grandmother refused liver transplantation

[d] Patient was in the liver transplant program

for success seems to be the episodes of cholangitis. The number of episodes of cholangitis is related directly to the success of revision. The probability of success is significantly higher in the patients with the total episodes of cholangitis less than four or the episode following redo-Kasai is 1 or 0. The findings are novel. In their report, Bondoc et al. [14] took into the account only the episode of cholangitis on the success rate of the initial portoenterostomy.

Our results show that 60 % of patients after revision of portoenterostomy had normal bilirubin levels after the procedure. Moreover, the long-term survival with native liver was possible in at least three patients, with another one enjoying jaundice-free survival 9 months after revision.

A common criticism on redo-Kasai is that an unintended consequence of the revision will increase morbidity and mortality at the time of liver transplantation, which will outweigh the benefit of possibly halting or slowing progressive liver failure [1, 13, 14]. However, in this study, the three patients who required liver transplantation have had long-term survival comparable to those receiving transplantation after initial Kasai procedure failed. The blood loss and operative time was comparable to those patients receiving LT without previous revision of Kasai portoenterostomy [15]. Our results are consistent with the previous report that revision does not affect the final outcome of liver transplantation, regardless it is performed by the same team or not [14–16].

Our results from a single institute in Taiwan indicate that, in selected cases with biliary atresia, redo-Kasai is still valuable in the era of liver transplantation [17], offering patients the chance of survival with the native liver, while at the same time did not increase the risk of liver transplantation.

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