RESULTS OF KASAI OPERATION FOR BILIARY ATRESIA: EXPERIENCE FROM HUE CENTRAL HOSPITAL

Ho Huu Thien

ABSTRACT

Background: *The purpose of this study is to review the short-term outcome of patients with biliary atresia (BA) treated by the Kasai operation at Hue Central Hospital.*

Methods: *Eighty-four BA patients treated by the Kasai operation between March 2006 and March 2018 were reviewed. The diagnosis of BA was confirmed by intraoperative cholangiography. The outcome of treatment was categorized into two groups: jaundice-free (total bilirubin < 2 mg%) and persistent jaundice (≥ 2 mg%). The outcome of Kasai operation was evaluated 1 year after surgery. Data are expressed as mean ± SD.*

Results: *Average age at the time of surgery was 92.3 ± 25.6 days. 76.2% (64/84) of patients had Kasai operation before 90 days of age. Histologically, 43 patients (51.2%) had liver fibrosis at the time of surgery. Sixty-two patients (73.8%) were jaundice-free 1 year after surgery. Age at the time of the Kasai operation did influence early outcome but not liver pathology. The most common complication was ascending cholangitis.*

Conclusion: *Near three fourth of our BA patients who underwent Kasai operation were jaundice-free 1 year after surgery. The lack of impact of age and liver pathology on outcome is presumably due to the briefness of the follow-up. In general, our patients underwent Kasai procedure before 3 months of age. It is therefore important for us to conduct a campaign to highlight the plight of these patients and the urgency of referral for neonates with jaundice.*

Key Words: *biliary atresia, cholangitis, Kasai operation*

I. INTRODUCTION

Biliary atresia (BA), a progressive inflammation and obliteration of both extra- and intrahepatic bile ducts, is a common problem in early infancy with cholestatic jaundice. The outcome is eventual death from liver decompensation or infection when they are left without any treatment, usually by the 2nd year of life.[1] The introduction of hepatic portoenterostomy by Kasai et al to correct obliterated extrahepatic bile ducts has improved patient survival.[2] Most surgeons attribute the successful use of the Kasai operation to early diagnosis, careful surgical procedure and prevention of postoperative cholangitis.[3–5] Nevertheless, liver disease, resulting from antenatal involvement of the intrahepatic bile duct is still progressing in patients who are operated. The progressive disease culminates in

1. Hue Central Hospital

Corresponding author: Ho Huu Thien
Email: thientrangduc@hotmail.com
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the end-state of cirrhosis for these patients who will eventually require liver transplantation.

In recent years, with more knowledgeable about this disease, a number of technical points have been given more attention. The results after Kasai surgery seem to be better.

The purpose of this study
is therefore to review the short-term outcome of patients

with BA treated by the Kasai operation at our institution.

II. MATERIALS AND METHODS

During the period from March 2006 to March 2018, data of all consecutive infants with BA treated by hepatic porto-jejunostomy with Roux-en-Y (original Kasai operation) from the medical records at Hue Central Hospital were reviewed for the following data: gender, age at the time of surgery, pre- and postoperative liver function tests, associated malformations, operative findings and postoperative complications. The majority of BA patients were investigated for diagnosis by repeated ultrasonography. The diagnosis of BA was confirmed by coeliotomy with intraoperative cholangiography before undergoing Kasai operation.

The outcomes of Kasai operation were assessed at 1 year after the procedure and categorized into two groups: jaundice-free (total bilirubin levels < 2 mg%) and persistent jaundice (total bilirubin levels ≥ 2 mg%). None of the patients in this study showed symptoms and signs of fever or ascending cholangitis (fever, elevated serum total bilirubin, changed stool color and leukocytosis) at the time of outcome evaluation. Types of BA were classified as previously described by Nio and Ohi: type I, atresia of common bile duct; type II, atresia of common hepatic duct; type III, atresia at porta hepatis.[6] The association between outcome and age at the time of Kasai operation as well as liver pathology was analysed using x^2 tests. All data are expressed as mean ± SD. For all statistical analyses, SPSS version 10.0 (SPSS Inc., Chicago, IL, USA) was used.

III. RESULTS

Eighty-four BA patients underwent Kasai operation during the review period. There were 38 (45.2%) males and 46 (54.7%) females with a mean age at the time of surgery of 92.3 ± 25.6 days (range, 28–152 days). 76.2% (64/84) of patients had Kasai operation before 90 days of age. Out of the 84 patients, 54 (64.3%) presented with clinical jaundice at birth while the remaining (35.7%) developed recognizable jaundice after the age of 1 month. Associated anomalies consisted of cardiac anomalies in two patients (tetralogy of Fallot, patent ductus arteriosus), Down's syndrome in two patient, duodenal obstruction by annular pancreas in one patient. There was no polysplenia syndrome detected in this series.

Among the 84 patients investigated by abdominal ultrasonography, 72.6% (61/84) presented triagle cord signs and 5% (37/72) had visualized gall- bladder reported by radiologists. Mean preoperative liver function levels were as follows: total bilirubin, 210 ± 93.2 μmol/L; direct bilirubin, 150 ± 54.3 μmol/L; serum glutamic oxaloacetic transaminase, 262.27 ± 150.01 IU/mL; serum glutamic pyruvic transaminase, 174.08 ± 109.79 IU/mL.

As a result of intraoperative cholangiography, 100% of patients had BA type III There was no BA type I and II detected in our series. Mean operative time was 184 ± 30.6 minutes (range, 96–220 minutes). Pathological report of liver biopsy revealed that 51.2% (43/84) already had liver fibrosis at the time of operation.

Perioperative complications occurred in six patients (7.1%). The complications were prolonged bile leakage in three patients, wound infection in

three patients. There was no death. Postoperative complications comprised cholangitis in 32 patients (38.1%), gastrointestinal bleeding from varices in seven patients whose livers were cirrhotic at the time of Kasai operation (8.3%). Among 32 patients with cholangitis, 16 (50%) had a single episode only and the remaining 16 (50%) had recurrent episodes.

1-year post-surgery, 62 (73.8%) were jaundice-free, and 22 (26.2%) had persistent jaundice. In the good outcome group, the mean duration between the operation and jaundice-free onset was 3.6 ± 2.8 months (range, 1–6 months). By using x^2 tests, there was no association between outcome and either age or liver pathology at the time of the Kasai operation, as shown in Table 1.

Table 1. Outcome of Kasai operation and patient age and liver histology at the time of surgery

	Jaundice free (n%)	Jaundice remaining (n%)	p
Age at operation			0.01
≥ 90 (n 62)	46	16	
≤ 90 (n 22)	10	12	
Liver pathology			0.19
No liver fibrosis (n 41)	24	17	
Liver fibrosis (n 43)	19	24	

IV. DISCUSSION

At present, the Kasai operation remains the most common initial management for BA patients at our institution. In addition, the timing of surgical intervention for BA patients is a crucial factor. Most of the major series have confirmed that the best results from Kasai operation are achieved when patients have the surgery before the age of 90 days.[7–12] An analysis of the BA patients treated by the Kasai operation at our institution showed that most of them (76.2%) had the surgery before the age of 90 days. . Mass screening for BA patients by examining all neonatal stools with stool colour cards, which was previously proposed by Maki et al,[13] may help patients with quicker diagnosis and surgery as a promising offensive policy. Alternatively, a campaign towards primary healthcare providers regarding the importance of early treatment needs to be urgently conducted.

A possible concern, 73.8 % of our patients were jaundice-free at the 1-year follow-up. The jaundice-free rates reported by various authors range from 15.5% to 54.9%.[4,5,15–18] A large series reported by Kasai et al demonstrated that 84 of 245 BA patients (34.3%) achieved jaundice-free status after surgery (Table 2).[5]. The results indicate that there has been an improvement in surgical outcome at our institution. This may be attributed to our improved surgical techniques (large dissection at portal region, isolation of hepatic arteries and portal vein, enough resection of fibrosis tissue) and more advanced knowledge in prevention of postoperative cholangitis (use of long-term oral antibiotics and Acide ursodesoxycholique). However, the outcome and the mean follow-up duration of BA patients in each institution were different; therefore, it is not appropriate to compare the results from different studies. The outcome of Kasai operation in this study was analysed at 1 year after surgery, which is quite short, and long-term follow-up has to be further evaluated.

Table 2. Jaundice-free rate in biliary atresia patients after Kasai operation

Reference	Period	n	Jaundice free rate %	Mean follow-up time
Kasai *et al [5]*	1953-1987	245	34.3	1-34 yr
Lilly *et al [15]*	1973-1988	215	28.0	86 mo(1-15yr)
Lin *et al [16]*	1976-1989	60	38.3	7.2 yr(1-14 yr)
Wildhaber *et al[29]*	1974-2001	81	38.0	92 mo (1-27 yr)
Carceller *et al [30]*	1974-1998	63	43.0	1-28 yr
Paiboon *et al [31]*	1996-2002	75	50.6	1 yr

Approximately half of the BA patients (54.4%) had liver fibrosis at the time of surgery. Hays and Kimura[19] reported a relationship between liver fibrosis and progno- sis. In their series, no infant whose liver biopsy showed severe fibrosis survived. Nevertheless, our data showed no association between outcome and the presence of liver fibrosis during surgery ($p = 0.19$).

Ascending cholangitis is the most common postoperative complication, which was noted in 38.1% (32/84) of our patients after Kasai operation in addition to the com- mon perioperative complication of abdominal surgery. Cholangitis frequently results in the cessation of bile flow, and repeated attacks cause a progressive deterioration in hepatic function. Prevention of cholangitis is therefore an essential factor for maintaining bile drainage. At our department, only hepatic portojejunostomy with Roux-en-Y has been carried out since 2006 with the special attention to the length of more than 20 cm of ascending jejunal limb. The incidence of ascending cholangitis in our series is comparable to the 40–60% rate of others.[3,24,25] Cholangitis is reported to be a risk factor associated with portal hypertension.[26] Because portal hypertension could develop even in BA patients who are jaundice-free, examination of oesophageal varices should be included in routine surveillance.

Despite the successful correction of extrahepatic biliary obliteration by Kasai operation, the on-going inflammation of intrahepatic involvement still exists. The on-going process elucidates why ultimate liver fibrosis is inevitable in a number of patients, who eventually require liver trans-plantation. Some authors have also improved survival rates via liver transplantation as a primary therapy for BA patients.[27,28] However, we think that liver transplantation should not be the first line of treatment in our country because of the shortage of liver donors and the very high cost of the operation. Therefore, liver transplantation in BA patients at our hospital was performed only in selected patients, who experienced end-stage liver cirrhosis and can comply with strict long-term follow-up.

V. CONCLUSION

Near three fourth of our BA patients, who underwent Kasai operation, were jaundice-free at 1 year after surgery. The age at the operation did influence jaundice free rate but not the liver pathology at the time of surgery. It is therefore important for us to conduct a campaign to highlight the plight of these patients and the urgency of referral for neonates with jaundice.

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