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Problems in Survivors of Biliary Atresia -5 or More Year After surgery-†

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=Abstract=**Hepatic porto-enterostomy or Kasai's original Roux-en Y porto-jejunostomy was done on 56 patients during the 1980-1985 period. Seventeen patients or 30 per cent had survived more than 5 years at the end of 1990, and are presently being followed up. Average survival age is 7.5 years after surgery. No survivor has clinical or laboratory jaundice. Esophageal varices were demonstrated in 8 patients (47%), and 4 had bleeding episodes. Five patients or 29 per cent had more than one episode of hospitalization for cholangitis. Growth was above 75 per centile in height in 53 per cent. Nine patients(53%) have normal liver function, showed neither varix and nor ascites. Good prognostic factors for 5 year survival were an operative age of 60 days or less, mild fibrosis of the liver, and a big fibrotic mass at the porta hepatis at the time of surgery.**

Key Words: *Biliary atresia, Kasai operation, Hepatic-portoenterostomy, Long-term survivors in biliary atresia*

INTRODUCTION

Long term survival after surgery of so-called uncorrectable biliary atresia in infants is possible after the Kasai operation (Ohi *et al.* 1986). Kim (1987) reported a 39 per cent two year survival rate among Korean patients. But very little is known about the long-term results after successful Kasai operations, especially outside of Japan, because of the small numbers involved. The purpose of this study is to show the problems and current status of 17 non-icteric survivors who were followed for more than 5 years.

MATERIALS AND METHODS

During the 1980-85 period, a classical Kasai operation or Roux-en Y hepatic porto enterostomy was performed on 56 patients with biliary atresia at the Seoul National University Hospital in Seoul, Korea. Death followed in 39 patients within two years after surgery, mainly from unsuccessful maintenance of bile flow. The remaining 17 patients(30%) have been living without jaundice for more than 5 years. The presence of portal hypertension, variceal bleeding, postoperative cholangitis, ascites, growth in

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height, and liver function are being followed regularly, and these problems are investigated. Also statistical analysis of possible good prognostic signs for the longer term survivors compared to the failed cases was carried out.

RESULTS

Seventeen out of 56 patients with biliary atresia survived more than 5 years after Kasai operation(Table 1). The average age of patients is 7.5 years of age(range 5-10). There had been no deaths among these patient at the end of 1992. Most of the death occured within 2 years of age. Several factors regarding prognostic significance were compared between the survived and failed groups(Table 2). Statistically significant prognostic factors were age at time of operation, degree of fibrosis of liver, size of fibrotic mass and ductuli at porta hepatis and the degree of clearing of jaundice. No patient with jaundice survived more than 2 years of age.

The status of 17 survivors is summarized in Table 3 and 4. Nine patients (53% of survivors) do not have either esophageal varices or evidence of abnormal liver function. Physical growth is within normal range in all nine normal patients. Two patients required repeated hospitalization for the treatment of postoperative cholangitis. One of them had persistant ascites, which finally cleared after the age of 8.However, the remaining 8 patients (47%) had developed portal hypertension. Varices were confirmed by either radiologic study or endoscopic examination. Half of the patients with varices eventually had bleeding episodes. Bleedings were controlled by conventional,conservative treatment. Endoscopic injection of sclerosing agent into the varices was considered seriously in one patient. It is assumed that this procedure will be required eventually in the near future. Abnormal liver function tests were evident in all except one during both the bleeding and post bleeding period. In that patient, the only abnormal sign was bleeding varices from portal hypertension. Surgical intervention of bleeding control,transection of the esophagus,is considered in this particular patient if bleeding continues. All patients with varices are on medical treatment. In four patients who had no bleeding episodes despite the presence of varices, spontaneous reduction of the severity of the varices was shown on either radiologic/endoscopic examination. These patients are also being closely followed. Platelet count of less than 100,000/cc was observed in all patients with bleeding varices and splenomegaly. It was also noted in one patient who does not have esophageal varices or splenomegaly.

Table 1. Survival without jaundice after Kasai operation

Year	No. of patients	>5 year survival
1980	11	3
1981	11	4
1982	11	2
1983	5	2
1984	8	3
1985	10	3
Total	56	17 (30%)

Most of these patients are living a very active life. Even with portal hypertension, their daily activity is not restricted except during the bleeding episodes. All patients are regularly attending nursery or primary school. One patient who was operated on in 1980 deteriorated after the age of 8. He developed a severe degree of cirrhotic liver and portal hypertension. He began to have severe variceal bleeding, requiring on average 4 hospitalization per year. Although he is not icteric, he is a candidate for liver transplantation. Partial liver transplantation was carried out in an earlier failure case(age 11 months) in our institution in 1992. For the long-term survivors, uncontrolled portal hypertension and a failing liver will be our criteria for the liver transplantation.

DISCUSSION

The survival after hepatoportoenterostomy for biliary atresia has gradually improved due to

Table 2. Good prognostic factors in >5 year survival without jaundice

	Dead (39)	Alive (17)
Operative age of 60 dats or less *	20	14
Mild fibrosis of liver *	5	7
Big fibrotic mass at porta hepatis *	10	11
Size of ductuli at porta hepatis * (>200 micron)	7	8
Clearing of jaundice within 6 mos.*	3	15

*: p value <0.05

Table 3. Status of 17 survivors without jaundice

Varices on x-ray/endoscopy, big spleen:	8/17 (47%)
Episodes of varix bleeding:	4/17 (24%)
Hospitalization for cholangitis after 5 yr.:	5/17 (29%)
Ascites (occasional):	5/17 (29%)
Growth (> 75 per centile height) :	9/17 (53%)
Normal liver function,no varix, no ascites:	9/17 (53%)

Table 4. Result of 56 Kasai operation after >5 years

Dead	39(70%)
Alive without jaundice	17(30%)
Normal liver function, no portal hypertension	9
Cholangitis(2), occasional ascites(1)	
Normal, no cholangitis, no ascites(7)	
Esophageal varices	8
Bleeding varices (4)	
Cholangitis(3) severe hepatic dysfunction(1)	
Non-bleeding varices (4)	

early operation, refinements in surgical techniques, and improvements in postoperative care. At present, about 30% of biliary atresia without jaundice are able to survive for a long time, and a majority of them have either portal hypertension or hepatic dysfunctions. Ohi *et al.*(1990) have reported a 10 year survival of 8-48%,an early to recent result, averaging 36%.Other institution well known for surgery for biliary atresia has reported 28% long term survival (Ohya *et al.* 1990) In the United States, a larger survey of 1976-89 period with 904 patients in over 100 institutions revealed a five year survival of 30% or a little more. (Karrer *et al.*1990) After the report of a 39% two year survival in 1987, the 5 year survival rate became 30% in this study. This indicates an acceptable range of survival rate.

The importance of early diagnosis and early operation for surgical treatment of this disease is strongly emphasized (Ohi et *al.* 1990). It has been demonstrated that early treatment is essential not only for immediate results, but also for long-term results. In our study, prognostic factors such as degree of hepatic fibrosis, size of fibrotic remnant of the bile duct at the porta hepatis (so-called fibrotic mass), and size of ductuli at the porta hepatis were actually dependant on the age at operation. The earlier the surgery, the more favorable all these time-related factors. Thus, operative age of 60 days or less is the most important prognostic factor for five year survival.

In 1989, Kimura and Akiyama reported the results of a survey collected from 49 institutions in Japan. There were 325 patients surviving more than 10 years in that survey. Among them, 48.3% are healthy, 16.9% are moderately healthy (but without signs of cirrhosis or jaundice). Fifty

eight or 17.8 per cent have portal hypertension. Ohi et al.(1986) reported an incidence of 33% in long term survivors, while Saeki *et al.* (1986) reported higher incidence of 69% by endoscopic examination. Detection of varices using both radiologic and endoscopic examinations in our study was 47%. Portal hypertension subsequent to the biliary cirrhosis of the liver have become major problems in long term survivors (Saeki et al. 1986, Ohi *et al.* 1990, Ohya *et al.* 1990). But esophageal varices and hypersplenism are not always fatal, because in many patients these sequelae can be managed medically. Endoscopic injection of sclerosing agents has become the standard treatment of pediatric esophageal varices.(Ohi *et al.* 1986) Also spontaneous improvement or disappearance of varices are not uncommon, as is the case in our study.

Daily activities, such as attending school, participating in sports were not impaired in most cases except end stage liver failure or in active variceal bleeding. Growth in weight and height compared to normal Korean age were not significantly different. No mental retardation or learning difficulties were noted in our series. Similar results have been reported in many papers. (Saeki *et al.* 1986, Ohi et al. 1990, Ohya et al. 1990)

For end stage hepatic dysfuction, liver transplantation is the only chance of survival. For early failure, an 11 month old post-Kasai patient received a left lobe from a living donor for a liver graft. For the long term survivor, a boy who was operated on in 1980 requires a liver graft. There is much controversy as to whether Kasai operation is ever indicated in the era of liver transplantation.(Stewart et al. 1988, Kasai *et al.* 1989). The success rate is definitely higher in liver transplantation, however, only one third of candidates actually can have a liver given from a donor. Thus, the scarcity of livers versus the one third success rate of Kasai should be weighed very seriously. The present study indicated that the Kasai operation can served either as a primary therapeutic as well as temporary, interim measure before transplantation is widely available.

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