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Recent Results of Surgical Treatment for Biliary Atresia: Experience at the Seoul National University

Woo-Ki Kim

Department of Pediatric Surgery, Seoul National University Children's Hospital and Department of Surgery, College of Medicine, Seoul National University, Seoul 110, Korea

= **Abstact** =Biliary atresia is a progressive, inflammatory, obstructive lesion of the bile duct, especially common bile duct, of the newborn period. It is universally fatal if either the Kasai operation or liver transplantation is not performed in due time. The first successful long term survivor of biliary atresia at S.N.U. Hospital was achieved in 1980 by the Kasai operation (hepatoportoenterostomy) and this 6 year old boy is now living normally and attending school. Since then, 17 more patients survived for more than two years of age without jaundice in 1978-1984 period (18/56 or 32 %).

Key Words: ***Biliary atresia, Jaundice, Neonatal jaundice, Cholangitis, Portal hypertension, Hepatic portoenterostomy, Conduit in Kasai, Kasai operation***

INTRODUCTION

The first successful long term survivor of biliary atresia at the Seoul National University in Seoul, Korea, was achieved in 1980, although ten previous hepatoportoenterostomies attempted in 1978-79 period all failed. Since then, 18 of 46 patients (39 %) with biliary atresia are living without jaundice for more than two years of age. (Table 1)

MATERIALS AND METHODS

1. Mode of Surgery: Success vs. Failure

Two major changes in surgical procedure were made for the first and subsequent successful cases: (1) the dissection and transection of the portal fibrous cone, and (2) the biliary-intestinal conduit.

DISSECTION AND TRANSECTION OF THE PORTAL FIBROUS CONE: Faulty level of transection of the portal fibrous cone and dissection in our 1978-1979 series was recognized by Kimura and Kasai through personal communications (Kimura *et al.* 1979)

Strict adherence to Kasai's portal dissection and transection method (Kasai *et al.* 1979) brought the first success. Freeing of the fibrous cone after the ligation of small venules from the portal vein to the cone is essential for the proper dissection. To avoid either supraportal (or intrahepatic) or infraportal transection of the portal fibrous cone, both portal vein and its major branches were used as a landmark for the transection (Ohi *et al.* 1980). Better bile excretions were achieved by this change of surgical technique.

BILIARY-INTESTINAL CONDUIT: The author had seen many failed cases after attempted hepatoportoenterostomy during pediatric surgical training in the United States. Experiences of caring of the massive G-I bleeding from the systemic portal-shunt at the enterostomy due to the portal hypertension guided us for the use of original Roux-en-Y hepatic portojejunostomy.

A 40 cm length of proximal jejunum was used for that purpose. Unfortunately, the first ten cases failed. Most of them did show excretion of bile after surgery for some limited period. Faulty level of transection of the portal fibrous cone seemed to be the main reason, since supraportal dissection (or presence of liver tissue at the resected specimen)

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Table 1. Survival of biliary atrelsia after hepatoportoenterostomy

Year	no. of patients	Survivor without jaundice	Operative procedure
1978	4	0	Kasai(?) I, 40 cm R-Y loop
1979	6	0	(0/10 survived)
1980	11	3	Kasai I, 60 cm R-Y loop
1981	11	6 *,**	(11/33 survived)
1982	11	2	*:varix bleeding, **:Cholangitis
1983	5	2	Kasai I, 40 cm R-Y loop
1984	8	5	(7/13 survived)
1978-84	56	18(32%)	(*:one has varix bleeding, the other
1980-84	46	18(39%)	has episodes of cholangitis)

Table 2. Mode of surgery and survival rate

Year	Survival rate	Mode of transection/Conduit
1978-79	0/10 (0 %)	Supraportal / 40 cm Roux-Y loop or Infraportal / 40 cm Roux-Y loop
1980-82	11/33 (33 %)	Portal / 60 cm Roux-Y loop
1983-84	7/13 (54 %)	Portal / 40 cm Roux-Y loop

or infraportal dissection were attempted in all. Practically all of these cases had suffered from frequent attacks of severe cholangitis. Thus, the evaluation of this conduit is practically impossible.

For the first successful case, a 60 cm loop was employed with the hope of reducing the chance of cholangitis. A total of 33 patients received this type of intestinal conduit. But a study by us (Kim and Park 1983) fail to demonstrate any advantage on using this variation. Since then, 40 cm loop Roux-en-Y hepatoportojejunostomy is being used (Table 2). There is no significant statistical difference in survival rate between the 60 cm loop group and 40 cm loop group ($p[z] > 0.10$).

2. Postoperative Measures:

Followings are our routine postoperative measures.

a. Choleretics: urso deoxycholic acid, p.o.
b. Oxygen therapy, immediate postoperative period.
c. Steroid, prednisone or prednisolone, tapering in 2 weeks.
d. I.V. antibiotics, for a 4-8 week period and oral antibiotics for a year or two.
e. Nutritional support, vitamin E., iron preparation.

RESULT

1. Long term survivor without jaundice:

Table 2 is the result of the modification. Survivors have no jaundice and are alive for more than two years.

2. Incidence of cholangitis:

Patients who showed good bile excretion after surgery usually did not have postoperative cholangits. In our eighteen successful patients, only three had episodes of severe cholangitis which required prolonged hospitalization (Table 3). For the patients who did not show good bile excretion, cholangitis was commonly seen after discharge from the hospital. Table 3 (Kimura *et al.* 1983) indicates an incidence of cholangitis in various operative procedures. But it is unfair to compare the incidence of cholangitis with various modification procedures.

3. Early diagnosis and surgery:

In our series, the mean age at operation is 77.1 day with standard devation of 27.8 day. However, there is no significant difference of age at the time of surgery between the failed or successful cases (Table 4). This indicates still our patients are too old when we get them.

4. Portal hypertension and variceal bleeding:

In our 18 long term survivors, three patients had episodes of cholangitis which required hospitalization. One five year old boy still has episodes of occasional mild cholangitis which respond well to

Table 3. Operative procedures and cholangitis (1977-82)

Procedure	No. of pts.	Cholangitis	Survivors
Tanaka: jejunal interposition with jejunal valve	20	2	18
Akiyama: Roux-en-Y	32	5	13
Yura: Double tract hepatic portojejunostomy	30	23	18
Suruga: Suruga II	59	14	40
Kasai: Double Roux-en-Y	39	18	30
Kim: Roux-en-Y (1980-84)	46	3/18*	18

* long-term survivor only

Table 4. Mean age at the time of surgery in biliary atresia

Mean age of surgery for the survivor:	72.9 day (S.D. 16.3) Range of 39-97 days
Mean age of surgery for the failure :	84.1 day (S.D. 30.9) Range of 40-146 days
T-value: 1.625 $p > 0.10$	

Table 5. Portal hypertension and B.A.

Pts age	Frequent Cholangitis	Varices on x-ray	Bleeding
6 years	episodes within 1 yr	yes	no
5 years	milder attacks	yes	no
5 years	milder attacks	yes	no
3 years	?	yes	yes

oral antibiotics. But he has an enlarged spleen and radiologic evidence of varices which have not yet bled.

The other, age 6, had several episodes of cholangitis within the first year after surgery, but he is now doing well. He has x-ray evidence of esophageal varices but has not yet experienced bleeding. The other five year old boy also shows x-ray evidence of varix. One patient, age three, who did not have episodes of cholangitis which required hospitalization also did have several episodes of varix bleeding. Thus, postoperative cholangitis seems to be related to the development of portal hypertension. (Table 5)

DISCUSSION

This operation or hepatic portoenterostomy does not belong to the type of surgery that is easily reproduceable. Still many surgeons all over the world have their "own" concepts about the portal dissection and transection. Also there still remains confusion about the intestinal conduit portion of this surgery among surgeons other than so-called "biliary atresia experts" who did accumulate a large number of patients. It is very hard to compare the result of various modifications against cholangitis since there are too many variables. The author have basically used the same intestinal conduit in all patients. In author's experience, poor bile drainage is not directly related to the intestinal conduit portion of the surgical procedure.

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= 국문초록 =

신생아 담도폐색증(Biliary Atresia)의 치료성적 : 서울의대 소아외과의 56예 성적

서울대학교병원 소아외과 및 서울의대 외과학교실

김 우 기

신생아 담도폐색증 환자에 대한 Kasai 수술후 서울대학교 병원 소아외과에서의 첫번째 장기 생존예는 1980년도에 이루어졌다. 현재 만 6세가 된 이 남자아이는 황달없는 건강한 상태로 국민학교 1학년을 다니고있다. 이후 17명의 환자들이 황달없이 2년이상 생존중이다. 그 요약은 아래와 같다.

Year	No. of patients	Survivor without jaundice	Operative procedure
1978	4	0	Kasai(?) 1, 40 cm R-Y loop
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1980-84	46	18 (39%)	the other has cholangitis)

1978-1979년 기간중의 수술수기상의 오류가 발견되고, 교정된 다음부터 생존예 들이 나오기 시작했다. 다시 말하면 Kasai 교수의 수술 방식을 철저히 따르는 것이 수술 수기중 가장 중요한 것이라고 단정할 수 있다.