

LONG TERM FOLLOW-UP OF KASAI OPERATION FOR BILIARY ATRESIA: A SINGLE CENTER EXPERIENCE

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Abstract- Children who are operated for biliary atresia by the Kasai procedure have approximately 30% chance of survival for 5 years. In an attempt to define the role of this operation for biliary atresia, the surgery records of the past 15 years were reviewed. The aim of this study was to assess the benefits achieved from this operation in infants with biliary atresia. This study was conducted in the department of pediatric surgery Taleghanei Medical Center from 1986 to 2000. A total of 36 cases, 15 boys and 21 girls were reviewed retrospectively. All the operations were performed uniformly by Kasai procedure by three investigators. Data regarding patient history, clinical presentation, laboratory and radiological features, operative finding, complication and mortality were collected and retrospectively studied. In these series 36 cases were classified as three groups. Group A, represented the successful category after the Kasai operation (11 patients, 30.5%) characterized by survival of more than 3 years and no jaundice. Group B (2 patients, 5.5%) was defined as survival of more than 3 years, but with jaundice, and group C (23 patients 63.8%) was defined by survival of less than 3 years (this group was further divided to subgroups). It seems that jaundice is the main prognostic factor after operation.

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Key words: Biliary atresia, Kasai operation, hepaticoportointerostomy, liver transplantation

INTRODUCTION

Extra hepatic biliary atresia is the single most common entity responsible for prolonged neonatal cholestasis. The prognosis was uniformly fatal within first 12 to 18 months of life before the use of hepaticoportointerostomy (HPE), first described by Kasai from Sendai, Japan, in 1959 (1, 2).

Overall, children who are operated for biliary atresia by the Kasai procedure have approximately 30% chance of survival for 5 years (3, 4). In recent years liver transplantation in biliary atresia patients has led to a success rate of more than 80%, and so

this procedure has become the first line therapy (4, 5). Unfortunately, liver transplantation is not performed routinely in this country. In an attempt to define the role of Kasai procedure, this report evaluates the efficacy of HPE in 36 infants with biliary atresia in the past 15 years who were treated in our hospital.

MATERIALS AND METHODS

Thirty six infants with biliary atresia were operated in the Taleghanei Medical Center from 1986 to 2000, which included 15 boys (41.6%) and 21 girls (58.3%).

All the operations were performed uniformly by Kasai procedure and were performed by the three investigators. During a 15 year period at this pediatric ward, 30 patients (83.3%) within 90 days of life, 4 patients (11.1%) within 90 to 120 days and 2 patients (5.5%) beyond 120 days underwent

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hepatico- portojejunosomy (HPJ) and were followed up for 1 to 14 years (mean 7.2 years).

First the gallbladder, which usually was rudimentary, was detached from the liver bed. The fibrous cord, including the remnant of the hepatic duct, was dissected denuding the portal vein and the hepatic arteries just below the portal bifurcation. The posterior surface of the portal fibrous mass became visible by downward retraction of the portal vein's bifurcation. Fibrous cord was then transected at the closest point to the liver with a sharp knife; meticulous care was observed not to enter to the liver parenchyma. HPJ was then performed using interrupted 5-0 Vicryl and anastomosis was done in end-to-side fashion. The Roux-en-y limb of the enterobiliary conduit was about 45 cm in length. If operative cholangiogram showed patent gall bladder, cystic duct, and distal common bile duct, hepaticoportocholecystostomy was then be performed.

Antibiotics were routinely given postoperatively for approximately 3 months. If the patient developed cholangitis, the prescription of antibiotics would be extended. In the post operation follow-up, these 36 patients were divided into 3 groups. Group A was characterized by survival > 3 years and without jaundice. Group B was defined as survival > 3 years but with jaundice. Group C was the group of patients who died at time of review. Group A was interpreted as success after Kasai operation.

RESULTS

Eleven patients (30.5%) were categorized in group A. Two patients (5.5%) belonged to group B, and 23 patients (63.8%) were categorized as group C. This group was further divided to group C₁ who died 1 year after operation (16 patients), group C₂ who died between 1 and 2 years (4 patients) and group C₃ (3 patients) who died between 2 and 3 years.

In the jaundice free group, no mortality was recorded during the 3 year follow up. The only major complication in this group was variceal bleeding due to portal hypertension in one of the patients which was managed by repeated transfusion and endoscopic sclerotherapy. Episodic cholangitis was noted in some patients of this group but easily controlled by antibiotic (2).

DISCUSSION

In this series of thirty six cases, 11 (30.5%) were classified as group A and represented the group of patients with a successful Kasai operation. In other words, this group represents the potential success rate in these series. Ohkohchi *et al.* made a useful contribution by highlighting the fact that 90% of 92 children without jaundice after portoenterostomy survived for more than 10 years, while 70% of the 39 children with persistent jaundice after the operation were dead by 6 months age (4). They further emphasized that only the total bilirubin was a reliable marker for hepatic failure in the end stage of biliary atresia (6, 7).

On the other hand, some patients who improved after Kasai operation were not completely cured from their jaundice and might not achieve long term survival if liver transplantation was not done for them. This certainly will avoid the risk of higher complication rate of transplantation at a very young age (8, 9).

Twenty three patients (63.8%) died within 3 years after Kasai operation, this group of patients were considered as unsuccessful (10). All of these patients never achieved a jaundice free status, the liver functions went downhill consistently and they died of hepatic decompensation (11). This group of patients should be referred for liver transplantation (7, 5). Patients with biliary atresia usually die within 12 to 18 months of life, so the operation definitely provided a better chance of survival (12).

Improved surgical techniques and immuno-suppressant agents and antibiotic have increased the success rate of liver transplantation (8, 10). In children, unfortunately, because of the limited number of suitable donor organs available, a significant number of children die while they are on the waiting list, the so-called hidden mortality of liver transplantation (13). Furthermore, the expertise and financial expenditure of a transplantation team is not always accessible in all parts of the world. These findings suggest that liver transplantation is not always a feasible alternative to the Kasai operation at present time. The jaundice free long-term survival rate after Kasai operation reported by various authors ranged from 15.5% to 45%. Kasai *et al.*

reported the largest series of 245 patients with the longest follow-up, 84 of whom (34.3%) achieved a jaundice-free survival (9) and the current report suggests that 30.5% of patients who underwent Kasai procedure will probably not require liver transplantation and group B patients (5.5%) will have extended survival and will reach an age with fewer transplantation complication and increased chances for obtaining a suitable size of liver (14).

Conflict of interests

We have no conflict of interests.

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