The Kasai Portoenterostomy: When Is It Too Late?

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Background/Purpose: Kasai portoenterostomy is recommended as the primary initial therapy for extrahepatic biliary atresia if the procedure can be performed within 10 to 12 weeks of life. The optimal management for infants with delayed presentation of biliary atresia remains controversial. The purpose of this study was to determine the success rate and outcome for patients who underwent a "late" Kasai portoenterostomy.

Methods: The authors conducted a retrospective review of the medical records of all patients with biliary atresia who underwent a Kasai portoenterostomy at their institution from 1986 to 1999 (n = 31). The authors analyzed success rates compared with age at the time of the Kasai procedure and the association with patient demographics. Surgical success was defined as achievement of a total serum bilirubin ≤ 2 mg/dL. Long-term follow-up assessments included the need for liver transplantation and patient survival rate.

Results: The demographics of this study cohort showed a predominance of African-Americans, 19 of 31 (61%), and girls, 23 of 31 (74%). Assessment of success compared with subject age at the time of the initial portoenterostomy showed that 52% (13 of 25) had successful Kasai procedure at 0 to 75 days, compared with 83% success rate (5 of 6) at age 76 days or older (P = .359). Liver transplantation was performed in 16 of 31 patients (45%). Overall survival rate for the entire cohort is 23 of 31 (74%), whereas 12 of 31 (39%) are currently alive without a liver transplant.

Conclusion: These data suggest that there is no contraindication to performing a Kasai portoenterostomy for biliary atresia in children over 75 days of age.

INDEX WORDS: Biliary atresia, Kasai portoenterostomy, surgical outcome.

BILIARY ATRESIA is defined as a progressive obliteration of the extrahepatic bile ducts in early infancy. When untreated, the outcome is death, usually by 2 years of age. In 1959, Kasai and Suzuki[1] described hepatic portoenterostomy to reestablish bile flow in the "noncorrectable" type of biliary atresia.

Kasai and other investigators have shown that surgical success and survival rate correlate with the age of infants at the time of surgery. To date, the majority of published studies show that the best surgical results are obtained in infants who are 60 to 80 days of age at the time of operation.[2-9] Clinical practice guidelines urge practitioners to refer infants with cholestatic jaundice for early evaluation, but significant numbers of infants present for surgical referral beyond 60 days.[10] The management approach to these infants is controversial, with some being treated with portoenterostomy and others being referred directly for primary liver transplantation.

Biliary atresia currently is the leading indication for liver transplantation in the pediatric population. Untreated infants with biliary atresia require transplantation early in life at an age at which suitable donors remain scarce. Donor livers from infant cadavers may have worse functions than those of older donors, and transplantation results are worse in small children because of technical difficulty and an increased complication rate.[11-13]

This study aims to assess the success of "late" surgery for biliary atresia in infants presenting after 75 days of age to determine whether it is a worthwhile alternative to early referral for liver transplantation. We also evaluate which preoperative factors may be predictive of surgical success in infants undergoing portoenterostomy after 75 days of age.

MATERIALS AND METHODS

Records were reviewed of all infants (n = 31) with biliary atresia who underwent portoenterostomy between February of 1986 and March of 1999 at Children's Healthcare of Atlanta at Egleston, Atlanta, Georgia. The following demographic, clinical, and laboratory variables were assessed: gender, race, age at time of surgery, preoperative transaminases, bilirubin level, alkaline phosphatase level, gamma glutamyl transpeptidase level, and postoperative bilirubin level at 1 week, 1 month, 6 months, 1 year, and 2 years. Complications of portoenterostomy and chronic liver disease as well as long-term outcome were studied. The definition of surgical success was clearance of jaundice (bilirubin level less than 2 mg/dL).[14]

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Analyses of demographic and clinical variables associated with surgical outcome were conducted using the SPSS (SPSS, Inc, Chicago, IL) and Computer Programs for Epidemiologic Analysis (USD, Inc, Stone Mountain, GA) software programs. In the comparison of children undergoing portoenterostomy (≤75 days *v* >75 days), for categorical predictors the exact mid *P* value was calculated, whereas for continuously measured variables, an independent *t* test was performed if the data were normally distributed, or else the Wilcoxon rank sum test was used.

RESULTS

Thirty-one infants underwent portoenterostomy during the study period. Twenty-three (74%) were girls. Nineteen (61%) were African-American. Twenty-five infants were operated on before or at 75 days of age with a success rate of 52%. Six infants had their portoenterostomies performed after 75 days of age with a success rate of 83%. There was no significant difference in success rates between the children undergoing portoenterostomy at ≤75 days versus more than 75 days ($P = .359$). Four infants had portoenterostomies between 90 and 120 days of age, and all had successful outcomes. All of these were girls. Three of these were African-American and one was white.

No demographic or preoperative laboratory variable was found to predict success in this population. The age at presentation was not significantly different between white and black infants ($P = .70$). In addition, there was no difference in overall surgical outcome based on race ($P = .489$). Follow-up studies of living patients ranged from 1 month to 167 months (median, 24 months). One patient was lost to follow-up. Survival data show 12 of 31 (39%) alive with portoenterostomy alone and 11 of 16 (69%) alive after portoenterostomy and transplant. Total survival rate is 23 of 31 (74%).

DISCUSSION

Contrary to previous reports, our study shows that infants undergoing portoenterostomy between 75 and 120 days of life have an equal (or better) chance for surgical success than infants undergoing this surgery before or at 75 days of age. The demographics of our patient population were different from those reported in previous studies. The predominance of African-American patients in our study (61%) is similar to that described by Yoon et al[15] in their population-based study in Atlanta. In contrast, Karrer et al[16] in their biliary atresia registry consisting of 816 patients undergoing portoenterostomy reported only 20% African-American patients. In that study, the investigators reported white race as a predictor of poor surgical outcome.[16] We did not find this in our population but question the influence of demographics on our patients' improved surgical success when operated on at older ages. Tagge et al[17] and Suruga et al[18] have shown results of late surgery similar to ours. Tagge et al[17] reported 77% of patients operated on after 12 weeks of age became anicteric. Suruga et al[18] reported 40% of children with biliary atresia operated on after 90 days of age had good outcomes.

Our clinical findings may be supported by the histologic review of Tan et al,[19] which found no correlation between the age at time of surgery and portal duct patency or the degree of portal inflammation in 205 cases studied. They state "it appears probable that the timing of onset, rate of progression, and severity of the disease process in biliary atresia varies from case to case and argues against definitive assumptions based purely on postnatal age."[19] The demographics were not mentioned in this study.

Several arguments may be made for performing a Kasai portoenterostomy in late-presenting infants. Clinically, these patients have the potential to do very well. All 4 of our patients who underwent portoenterostomy between 90 and 120 days of age are growing normally and have no significant medical problems. All 4 have evidence of portal hypertension, but only one patient has had complications (undergoing successful sclerotherapy for bleeding varices in her first year of life—she is now 10 years old).

Extrahepatic biliary atresia is the most common indication for liver transplantation in the pediatric population. Successful portoenterostomies in infants up to 120 days of age would decrease the number of infants awaiting donors at an age when organ scarcity is most prevalent. Recent UNOS statistics show more than twice as many patients awaiting organs between 0 and 5 years of age as between 6 and 10 years of age.[20]

Finally, several investigators have described no adverse effect of previous portoenterostomy on transplant outcome. Sandler et al[21] found no difference in survival rate between patients with a previous portoenterostomy compared with those with primary transplantation. Wood et al[22] found no difference in perioperative complications between biliary atresia patients and those patients undergoing liver transplant for other indications. Moreover, patient survival rate was greater in the biliary atresia group (all but 2 of whom had undergone portoenterostomy). Millis et al[23] found no difference in blood loss, perioperative complications, or survival rate between those patients with previous portoenterostomy and those without.

Based on our data, we believe Kasai portoenterostomy may be performed successfully in all infants presenting up to 120 days of age. These patients have a reasonable chance for a successful outcome, and if they do not, the prior portoenterostomy will not adversely affect the results of a liver transplantation procedure. Patients with successful portoenterostomies grow and develop well, so that they may ultimately be better liver transplant candidates.

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Discussion

E. Tagge (Charleston, SC): This is not the first time this has been presented. At the University of Michigan we reviewed our experience some 10 or 12 years ago in a report on which my wife was actually the primary author published in *Annals of Surgery*, and we found the exact same thing, that age at presentation, whether it be 2 months or less, 2 to 4 months, or greater than 4 months, there was no statistical difference in the survival rate or the success rate of the Kasai procedure. I really do not have a question, but just to comment and support your presentation.

H. Lee (response): Thank you.