Comparison of Drainage Techniques for Biliary Atresia

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Purpose: Traditional Kasai portoenterostomy and porto-appendiceal duodenostomy have been utilized for biliary atresia. Differences in outcome between patients who underwent either Kasai portoenterostomy or porto-appendiceal duodenostomy were compared.

Methods: A review of all children who underwent a drainage procedure for biliary atresia from 1986 to 2000 (n = 30) was performed. Age at drainage procedure, subsequent liver transplantation, and outcomes were evaluated. Outcome variables included success rates (total bilirubin < 2.0 mg/dL) and survival rate. Statistical analysis was done with χ^2 and Student's *t* test.

Results: Long-term follow-up was available on 28 of 30 patients. Age at biliary drainage was insignificant. Success rates between porto-appendiceal duodenostomy (31%) and Kasai portoenterostomy (82%) were statistically significant. Survival rate for patients who underwent a Kasai portoenterostomy was 10 of 11 patients. Survival rate for patients who underwent porto-appendiceal duodenostomy was 14 of 16 patients. Overall survival rate was comparable between porto-appendiceal duodenostomy (88%) and Kasai portoenterostomy (91%).

Conclusions: Although overall survival rate was comparable, patients who underwent porto-appendiceal duodenostomy were less successful in alleviating hyperbilirubinemia compared with Kasai portoenterostomy. This is shown further by the greater incidence of subsequent liver transplantation in infants with prior porto-appendiceal duodenostomy. Although the appendix may serve as an alternative biliary conduit, traditional Kasai portoenterostomy appears to achieve better biliary drainage.

INDEX WORDS: Biliary atresia, Kasai portoenterostomy, appendix.

BILIARY ATRESIA (BA) is a disorder of infancy characterized by the progressive obliteration of the extra- and intrahepatic biliary duct system leading to the obstruction of bile flow. Persistent cholestasis results in progressive conjugated hyperbilirubinemia, cirrhosis, hepatic failure, and death. In the 1950s, Kasai introduced the hepaticoportoenterostomy, which improved long-term prognosis for children with BA.[1] Despite good surgical outcomes, recurrent cholangitis, progressive jaundice, complications of portal hypertension, and end-stage liver disease persist. Multiple variations of intestinal conduits for biliary reconstruction to prevent these complications have been described.[2-5]

In 1971, Grosfeld et al[6] first reported the use of an appendiceal graft for biliary reconstruction. Since then, various techniques utilizing the appendix as a biliary conduit have been introduced with mixed results.[7-9] Crombleholme et al,[8] from this institution, described the use of biliary appendico-duodenostomy as a nonrefluxing conduit for biliary reconstruction in 2 cases for extrahepatic biliary atresia and type I choledochal cyst.

The current study reviews the experience with the standard Kasai portoenerostomy (KPE) and porto-appendiceal duodenostomy (PAD) as primary treatment of BA with regard to adequate biliary drainage, subsequent need for liver transplantation, and overall survival rate.

MATERIALS AND METHODS

From 1986 to 2000, all infants (n = 30) with BA and treated primarily by a biliary drainage technique at the University of California, San Francisco were identified by reviewing the pediatric surgery database. Biliary drainage technique was either standard KPE or PAD.[8]

The medical records of this cohort were reviewed retrospectively. Data points included age at drainage procedure, subsequent liver transplantation, rate of successful biliary drainage, and survival rate. Successful biliary drainage was defined as total bilirubin level less than 2.0 mg/dL at any point postoperatively but before liver transplantation.

Data are presented as mean and median values. Statistical comparisons between groups were done with Student's *t* test and χ^2 with Yate's correction. This study was performed with approval from the Committee on Human Research.

RESULTS

Long-term follow-up was available in 28 of 30 patients. Twelve of 28 infants (43%) underwent standard

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KPE, whereas 16 of 28 infants (57%) underwent PAD as their primary surgical treatment for BA. The 2 infants lost to follow-up underwent PAD. Within the KPE group, 6 infants (50%) were boys, and 6 infants (50%) were girls, whereas the PAD group consisted of 10 boys (63%) and 6 girls (37%). The median follow-up for the entire series was 6.8 years (range, 1.0 to 18.9 years) with 5.6 years (range, 1.0 to 18.9 years) for KPE and 8.2 years (range, 1.8 to 14.8 years) for PAD.

Preoperative laboratory values between the 2 groups were analyzed. Serum total bilirubin, direct bilirubin, aspartate aminotransferase (AST), alanine aminotransferase (ALT), alkaline phosphatase, and gamma-glutamyl transpeptidase levels (GGTP) were compared. No significant difference in preoperative chemical profiles existed between infants that underwent KPE and PAD.

Kasai Portoenterostomy

KPE was utilized as primary treatment for BA in 12 infants. Four infants had associated congenital anomalies including 3 with situs inversus, one with intestinal malrotation, and 2 with complex heart disease. The median age at surgery was 54.0 days (range, 12 to 97 days). Successful biliary drainage was achieved in 9 infants (75%) at some time after KPE. Six infants (50%) that had initially undergone KPE required subsequent liver transplantation, including one patient who had undergone previously successful biliary drainage.

There was no perioperative mortality in the KPE group. Survival without liver transplantation was 66% (4 of 6 infants), whereas the overall survival rate was 83% (10 of 12 infants). In the 2 nonsurvivors, infants died secondary to their congenital heart disease. Neither patient had received liver transplantation after KPE. Both died approximately one year after initial biliary reconstruction, in which one had successful biliary drainage.

Porto-Appendiceal Duodenostomy

PAD was utilized as the primary treatment for BA in 16 infants with a median age of 65.5 days (range, 9 to 224 days). Successful biliary drainage was achieved in 5 infants (31%). One infant underwent conversion of PAD to standard KPE and eventual liver transplantation. Fourteen patients (88%) eventually required liver transplantation after PAD. Of the 2 infants without liver transplantation, both had successful biliary drainage.

Overall survival rate in infants that underwent PAD as their primary procedure was 88% (14 of 16 infants). This includes both infants that have not required subsequent liver transplantation. There were no perioperative deaths. Both deaths had occurred in infants who underwent liver transplantation with subsequent lymphoma and chronic rejection.

Summary of Drainage Techniques

To date, 24 of 28 infants (86%) with BA treated primarily with biliary reconstruction are alive, and 4 of 28 (14%) have died. Only 2 deaths appear to be related to their biliary disease and the other 2 from cardiac causes. Overall successful drainage was achieved in 14 infants (50%). Subsequent liver transplantation was necessary in 20 of 28 infants (71%). Six of 28 (21%) patients are alive and have not needed liver transplantation.

Outcome variables were analyzed and compared between KPE and PAD patients (summarized in Table 1). Overall survival rate and survival rate without liver transplantation were not statistically significant. However, rate of successful initial biliary drainage was significantly greater in infants who underwent KPE compared with PAD. (75% KPE *v* 31% PAD). This translated into a higher rate of liver transplantation in the PAD group (88% PAD *v* 50% KPE).

DISCUSSION

In 1978, Adelman[10] described the natural history of BA in 89 infants without surgical intervention. The "rate of apparent cure" was 1.1% with the average age of death at 12 months. Approximately 25 to 35% of infants who undergo KPE will survive more than 10 years without liver transplantation. Approximately one third will drain bile but develop complications of cirrhosis and eventual require liver transplantation. For the remaining one third of infants, bile flow is inadequate after KPE, there is raid progression to cirrhosis, and these infants usually die by 2 years of age without liver transplantation.[11] Nearly all children with untreated BA will die by 2 years of age.

Several groups have introduced various reconstructive

Table 1. Comparison of Biliary Drainage Techniques

	Porto-Appendiceal Duodenostomy (n = 16)	Kasai (n = 11)	*P* Value
Median age at biliary drainage	65.5 days	54 days	>.5
Patients receiving liver transplantation	14	4	<.05*
Successful biliary drainage	5	9	<.05*
Survival without liver transplantation	2	6	>.5
Overall survival rate	14	10	>.5

*Statistically significant ($P < .05$).

conduits to achieve better biliary drainage and reduce biliary reflux.[3-5,7-9] The use of an appendiceal graft has been attractive because of its small-caliber and isoperistaltic contraction. These features hypothetically could contribute to preventing reflux and preventing ascending cholangitis.[8,12] However, the long-term impact of appendiceal conduits as biliary reconstruction for adequate biliary drainage, subsequent need for liver transplantation, and survival is unknown.

In comparing biliary drainage techniques, there is no difference in overall survival rate or survival rate without liver transplantation. Patients who underwent PAD had less successful biliary drainage compared with standard KPE. This failure to adequately drain bile translated into a higher rate of liver transplantation in the group who underwent PAD. Although the overall survival rate does not appear to be affected, the appendix does not appear to be an adequate alternative to standard KPE in achieving long-term adequacy of biliary drainage in infants with BA.

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